Investigation and management of the neck lump

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Abstract

A neck lump is a common presenting sign in both the paediatric and adult population and may represent a broad range of benign or malignant diagnoses. The appropriate initial assessment, investigation and management is key to delivering appropriate treatments and to avoid missing potentially serious diagnoses. There is a range of imaging modalities available to the treating clinician and huge variability in the appropriate surgical or non-surgical management of disease. In this article we discuss the approach to the assessment of patients with a neck lump, including the history and examinations which should take place. We discuss the imaging modalities which are most appropriate for each condition and the range of management options available. Both common and rarer diagnoses are discussed through the course of the review.

Keywords Head and neck cancer; lymph node enlargement; neck lump; thyroid

Introduction

A neck lump is a common presenting sign in the adult and paediatric population and a wide variety of conditions require consideration. The initial priority in the assessment of a neck lump, particularly in the adult population, is to identify malignancy in order to expedite treatment. In this article we discuss the assessment of the patient presenting with a neck lump and then discuss the conditions which cause neck lumps, their investigation and management.

Assessment

History

All patients should have a comprehensive ear nose and throat history. The key points specific to the head and neck to include in the history are included in Box 1. It is vital that a full and comprehensive history is taken from every patient, including past medical and surgical history, drug history (including allergies) and social history, including details of living arrangements, employment and support networks. Accurate documentation of smoking and alcohol consumption is imperative.

History and examination of the patient presenting with a neck lump

- History
  - Onset
  - Duration
  - Change/fluctuation in size
  - Pain
  - Skin changes
  - Related symptoms
    - Fever/rigors
    - Weight loss
    - Anorexia
    - Night sweats
    - Symptoms of hyper/hypothyroidism
    - Hoarse voice
    - Dysphagia
    - Odynophagia
    - Otalgia
    - Haemoptysis/bleeding
- Examination of mass
  - Size
  - Tenderness
  - Overlying skin changes
  - Mobility
  - Pulsation
  - Temperature
  - Translucency
  - Movement on swallowing/tongue protrusion

Box 1

Examination

All patients should have a complete ear, nose and throat examination together with an assessment with a flexible laryngoscope. Neck examination should include all levels of the neck, including the parotid and submandibular glands and an examination of the skin, face and scalp if it is clinically necessary. The clinical assessment of the lump itself is detailed in Box 1.

Investigation

The most appropriate investigation depends on the age of the patient and the possible differential diagnoses; these will be discussed later in this article. Venous blood sampling may point towards and infective cause. Ultrasound (US) provides good characterization of lymph nodes and thyroid nodules; it also allows concurrent needle sampling of the mass if required. Computed tomography (CT) provides detail of the bony anatomy; its use for soft tissue of the neck depends largely on local availability and expertise. Magnetic resonance imaging (MRI) provides excellent visualization of the soft tissues and requires no exposure to X-ray. Positron-emission tomography helps in the detection of a primary source in carcinoma of unknown primary and can also be used in the investigation of the patient with advanced cancer of the head and neck (see Figure 1).

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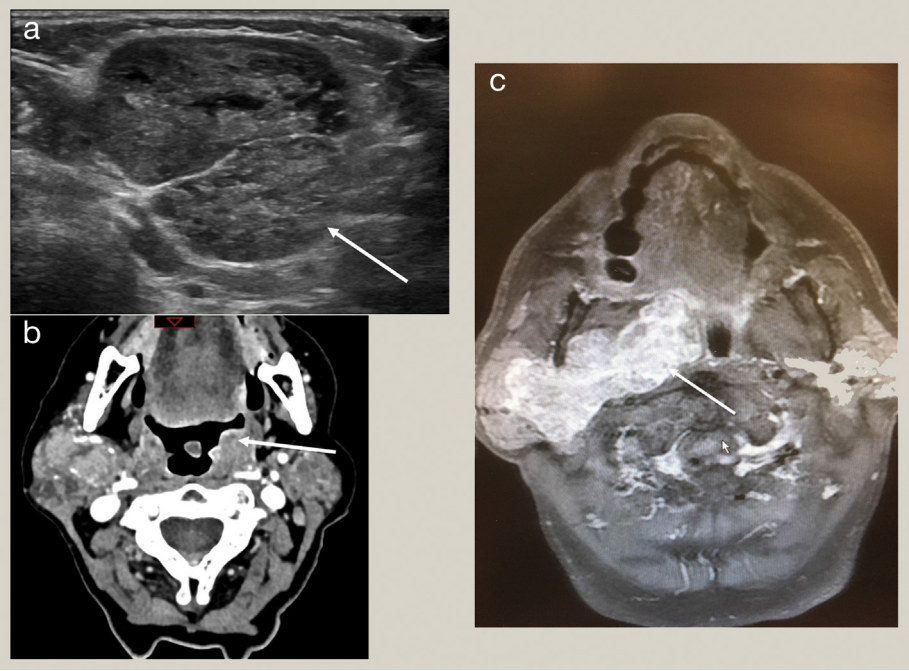

Figure 1 Ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) have various benefits depending on the requirement for tissue sampling, soft tissue or bony detail and visualization of deeper structures. The use of each modality often relies on local availability and expertise. Arrow points to abnormal finding in each scan. (**a**) US showing loss of normal hilar structure in lymph node. (**b**) CT scan showing unilaterally enlarged left tonsil. (**c**) MRI scan showing large mass in right parotid involving deep lobe.

Management of a neck lump

It is important to emphasize that not all neck lumps need any other management than reassurance. Many patients present with masses, which are a normal part of their anatomy (submandibular gland, hyoid bone etc), or with benign lymphadenopathy which will settle with time. If initial investigations are reassuring, the clinician must bear in mind that surgical management in order to gain a diagnosis and relieve anxiety may hold more risks to the patient than conservative management; of course, patients should have appropriate 'safety-netting' information if this plan is followed. If pathology is suspected then further investigation (and resultant management) should be performed based on the information gathered in the history and examination. In the majority of cases, ultrasound with possible fine needle aspiration is a common, and wholly appropriate, starting point.

Congenital neck lumps

Thyroglossal duct cyst

During foetal development, the thyroid gland forms from an endodermal thickening at the foramen caecum and then descends to lie in the neck at the 8th week of gestation. Failure of this tract to obliterate can then lead to the formation of a thyroglossal duct cyst. They are the most common midline neck swelling and can be found in up to 7% of the population. Thyroglossal duct cysts typically present as a midline neck lump which moves on protrusion of the tongue. US confirms the diagnosis and confirms the presence of normal thyroid tissue. Surgery forms the mainstay of treatment and should include excision of the middle portion of the hyoid bone (Sistrunk's procedure).

Dermoid cyst

Dermoid cysts can present anywhere along the lines of embryonic closure and are the second most prevalent midline neck mass. They are formed through the inclusion of epithelial cells and contain both ectodermal and mesodermal elements which can lead to the presence of hair follicles and sebaceous glands. Just under half are present at birth, with the remaining normally presenting by the age of 5 years. The diagnosis is usually clinical but if there is any doubt, US should be performed. Dermoid cysts do not move on swallowing or tongue protrusion and are treated with surgical excision as they can become repeatedly infected.

Branchial abnormalities

First branchial arch anomalies are very rare but can present with a mass in the lower pole of the parotid gland and can therefore be easily misdiagnosed as parotitis or a parotid tumour. Investigations include US and MRI to assess the relationship to the facial nerve. Treatment is surgical and its close proximity to the facial nerve dictates a parotidectomy incision and facial nerve monitoring.

Second branchial arch anomalies are the most frequent branchial abnormality and tend to present with a pit or swelling in the mid or lower neck at the anterior border of the sternocleidomastoid muscle. They classically run over the carotid sheath and hypoglossal nerve and beneath the posterior belly of the digastric muscle between the internal and external carotid arteries. Diagnosis is usually clinical though a sinogram which may help delineate the extent of the tract. Surgery is the treatment of choice in symptomatic cases and can be performed with a step-

ladder incision; in small children excision may be possible through a single horizontal incision. It is important to assess the child for other pits (particularly pre-auricular) and any concurrent ear abnormalities as in these cases renal US is needed to exclude branchio-oto-renal syndrome, a rare autosomal dominant syndrome.

Third and fourth branchial arch anomalies: These are exceedingly rare and tend to present with recurrent cervical abscesses or thyroiditis with nearly all occurring on the left side. On examination with a flexible nasendoscope it may be possible to see an opening in the piriform fossa. A barium swallow is indicated to look for a tract; ideally this should be performed after any acute infection has settled down. Treatment involves surgical excision of the entire tract with closure of the opening into the piriform fossa. It may be necessary to perform an ipsilateral hemithyroidectomy.

Branchial cysts form due to a failure of obliteration of one of the branchial clefts. Most branchial cysts are from the 2nd arch and present anterior to the sternocleidomastoid muscle; they can have fistulae leading to the tonsillar fossa. They most commonly present as a soft and trans-illuminatable lump and there may be a history of enlargement and tenderness following an upper respiratory tract infection. One needs to be mindful as these can be clinically indistinguishable from a cystic metastatic node and therefore a full head and neck examination and fine needle aspiration (FNA) cytology needs to be carried out. In clinical practice, patients over 40 years of age should be treated as malignancy until proven otherwise. The mainstay of treatment is surgical excision.

Vascular tumours

Vascular tumours are broadly separated into haemangiomas and vascular malformations.

Haemangiomas are common, classically present after birth (around 6 weeks) with rapid endothelial proliferation; the natural history is for involution and complete resolution by the age of 10. The treatment for most haemangiomas is conservative provided there is no airway compromise or evidence of skin breakdown. In these cases the use of propranolol may be appropriate which would need to be done in conjunction with the paediatric team. Oral prednisolone, intralesional triamcinolone or interferon therapy can be also used.

Vascular malformations differ from haemangiomas in that they are present at birth, grow with the child and do not involute. Lymphatic malformations ('cystic hygroma') will most commonly be seen in the ENT clinic presenting as a soft, trans-illuminating neck mass. Treatment depends on the location of the malformation and consists of either surgery or sclerotherapy.

Benign lymph node enlargement

Cervical lymphadenopathy is a common presentation in the paediatric ENT clinic and the concern is missing serious pathology. In the paediatric population reactive lymphadenopathy is common with more than a third of children receiving the diagnosis of cervical lymphadenitis every year.[1] In general, lymph nodes which are larger (greater than 2 cm) or those in the supraclavicular fossa are more likely to be concerning and therefore merit further investigation. The unreliability of FNA cytology in lymphoma together with the preponderance of this malignancy in children argues against the use of this investigation in this population; either US, if the radiologist has the expertise to distinguish between benign or malignant pathologies, or an excision biopsy should be undertaken. It is reasonable to send bloods for full blood count and CMV, toxoplasma, bartonella and EBV serology as sampling has low morbidity and can identify an infectious cause in 10% of patients.

Kawasaki disease (mucocutaneous lymph node syndrome)

Kawasaki disease is a vasculitis of unknown aetiology. Diagnosis relies upon the clinical presentation including a fever of a least 5 days duration and classically four out of the five following signs: cervical lymphadenopathy (>1.5 cm and usually unilateral), erythema and cracking of the lips, bilateral bulbar conjunctival injection, rash, erythema of the hands and feet which may lead to desquamation.[2] The mainstay of treatment is with intravenous immunoglobulins and should be managed urgently with the paediatric team to reduce long-term cardiac complications.

Kikuchi-Fujimoto disease

Kikuchi-Fujimoto is a rare, benign and self-limiting disease. It is characterized by cervical lymphadenopathy and sweats. A biopsy is usually required to rule out an underlying malignancy and the histology classically shows necrotizing lymphadenitis. Treatment is supportive and the majority of patients get complete resolution within 4 months.

Infective lymph node enlargement

Non-tuberculous mycobacteria

Non-tuberculous mycobacteria (NTM) are a frequent cause of cervical lymphadenitis. The most common organisms are: *Mycobacterium avium, M. haemophilum, M. kansasii* and *M. scrofulaceum*.[3] This condition classically occurs in immunocompetent children aged 1–5 years and normally presents as a slowly enlarging neck mass in the submandibular or parotid region. There is a wide variety of progression ranging from spontaneous resolution to increasing growth, violet discolouration and fistula formation. Positive culture of NTM is only gained in about 40–50% of patients and therefore one needs to be mindful of other potential diagnoses. In the case of a negative culture, one is guided by the clinical picture (slowly enlarging mass in the parotid or submandibular area with purple discolouration), the US (showing liquefaction and microcalcification) and the histology (necrotizing granuloma). The choice of first-line management is controversial. Surgery should involve removal of the affected node as incision and drainage may lead to a chronic discharging fistula. If the position of the node presents an unacceptable risk of complications, long-term antibiotics may be beneficial; rates of control of the disease and cosmetic outcomes is however considered to be worse than if surgery were used.

Infectious mononucleosis

Primary infection with the Epstein–Barr virus (EBV) is usually seen in childhood as a mild self-limiting disease. The severity of the infection tends to increase with age with a typical presentation including malaise, fever, cervical lymphadenopathy, sore throat and fatigue. Diagnosis is confirmed using the monospot test and/or EBV serology. Liver function tests are abnormal in up to 80% of patients and the spleen is commonly enlarged precluding contact sports for 6–8 weeks following diagnosis because of the risk of possible rupture. The treatment is usually conservative, but patients may need antibiotics (benzylpenicillin ± metronidazole) if there is a superimposed bacterial component.

Cat scratch fever

Cat-scratch disease typically presents with a papule at the site of infection and then regional lymphadenopathy lasting weeks to months.[4] It is caused by the bacterium *Bartonella henselae* or the newly recognized *Bartonella clarridgeiae*. Lymphadenopathy associated with this is generally self-limiting and the outcome is not improved with the use of antibiotics in immunocompetent patients.

Tuberculous lymphadenitis

Patients often present with hilar and bilateral cervical lymphadenopathy with the cervical nodes initially being non-tender and firm but which may subsequently develop into a fluctuant abscess. It is caused by *M. tuberculosis*, an air-borne pathogen which more commonly affects those with recent history of foreign travel, the immunocompromized and the elderly. Incision and drainage of these nodes is contraindicated as that can lead to chronic fistulation: the primary treatment is isolation of the patient, identification of any other high-risk family members and multidrug antibiotic therapy. This should be done in conjunction with the local microbiology or infectious diseases teams.

Toxoplasmosis

Toxoplasmosis is caused by the parasite *Toxoplasma gondii* and usually present in the posterior triangle of the neck. The diagnosis is made through serological testing and the treatment is usually conservative unless the patient has underlying health concerns. Women of child bearing age should be advised to avoid pregnancy until clear of the disease.

Brucellosis

Brucellosis is a highly contagious bacterial disease transmitted from animal to human either by the ingestion of contaminated food, unpasteurized milk or by direct contact with infected animals. Diagnosis is made using serological testing and a history suggesting likely exposure to the pathogen. A variety of antibiotic regimes are available such that microbiology advice is indicated.

Actinomycosis

Actinomycosis is a rare progressive granulomatous infection that can present at any age but the features that often occur in children are dental caries with lymphadenopathy in the submandibular region or at the angle of the mandible (lumpy jaw). Multiple abscesses can form with fistulation and a purulent discharge containing 'sulphur granules' in a sizeable number of cases. Diagnosis is made on culture of aspirate and if found in children, they should be investigated for an underlying granulomatous condition. Long-term penicillin is the mainstay of treatment but surgical debridement may have a role.

Salivary gland disease

Many masses in the head and neck originate from the salivary glands. The majority of these originate in the parotid gland and most are benign.

Benign

Salivary duct stones are more common in the submandibular gland than the parotid gland. They lead to an enlarged and painful gland, which may fluctuate in size, sometimes related to eating. Palpation of the floor of the mouth should reveal the presence of the stone. Acutely, stones may pass spontaneously; however, removal of the stone may be required. US can be used to identify intraglandular stones, stones in the duct or the presence of infection or abscess. If submandibular duct stones are recurrent or problematic, removal of the gland may be required.

Sialoadenitis: Thorough clinical assessment with history and examination is often all that is required for diagnosis of an acute infection of the parotid or submandibular gland but US scan can rule out an abscess if there is clinical suspicion. It is more common in those who are elderly, dehydrated or immunocompromized. Treatment consists of amoxicillin, co-amoxiclav or clindamycin together with adequate hydration and measures to increase saliva production.

Benign tumours: The majority of tumours (80%) presenting in the salivary glands occur in the parotid gland, and the majority of these (80%) are benign. Pleomorphic adenoma is the most commonly presenting benign tumour (80%) and is usually unilateral. It presents as a slow-growing, painless mass with no over-lying skin changes. Warthin's tumours are the second most common benign lesion and may be bilateral (see Figure 2). The

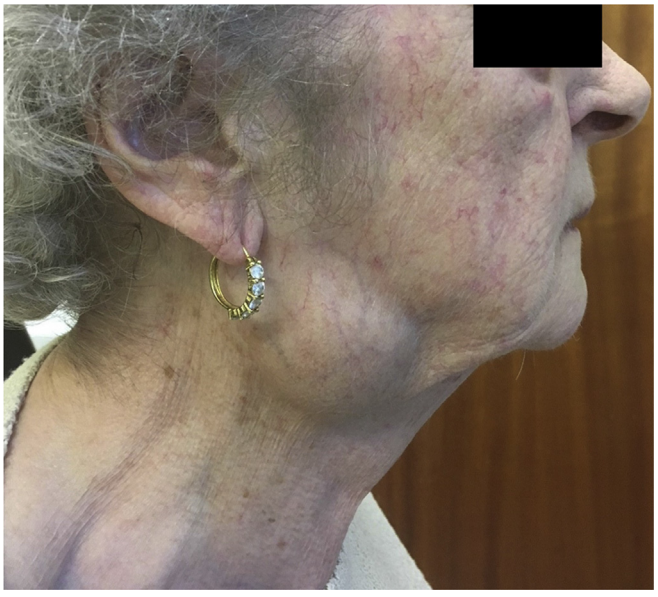

Figure 2 Warthin's tumours are often bilateral and often in the tail of the parotid. Note how parotid tail lesions often appear to be quite low in the neck and may be misdiagnosed as a result.

latter are more common in older males who smoke. All tumours require FNA assessment, usually obtained using US guidance. US scans will give information about the size and position of the tumour, and some information about deep lobe involvement. MRI allows a more detailed assessment of the tumour position and size, especially if there is deep lobe involvement. Pleomorphic adenoma have around a 5% rate of malignant transformation and surgical excision is usually recommended. There is a significantly lower rate of malignant transformation of Warthin's tumours (0.3%). Parotidectomy carries a small risk of temporary or permanent facial nerve dysfunction.

Paediatric salivary masses: Less than 5% of all salivary tumours occur in the under 16 age group and so diagnosis can often be missed. Up to 60% of solid tumours are likely to be malignant although these are usually low grade. As with adults, a rapidly increasing mass or facial nerve weakness normally indicates malignancy. Mucoepidermoid carcinoma accounts for 50% of paediatric salivary malignancies with acinic cell tumours comprising about 20%. Pleomorphic adenomas make-up about 30% of all paediatric salivary tumours and mainly occur in the parotid gland. Treatment is the same as for adults and treatment should take place in a MDT setting including a paediatric oncologist.

Malignant

Salivary gland malignancies are rare and although, in adults, the parotid gland has the highest frequency of tumours, the incidence of malignant salivary disease is relatively higher in the submandibular and sublingual glands. All salivary gland lesions should be seen in a rapid access head and neck clinic. They tend to present as a single, discrete and mobile lesion. Accompanying pain, fixation or facial nerve involvement is suggestive of malignancy. It is important to examine the patient for, and ask about, any current or past skin lesions as a parotid lump may represent a lymph node metastasis from a skin squamous cell carcinoma (SCC). US with FNA is the primary investigation of choice and this can distinguish benign from malignant disease in 90% of cases. Surgery is the mainstay of treatment with accompanying neck dissection. In patients who are not fit for surgery or where the tumour is inoperable, primary radiotherapy can have a role. Postoperative radiotherapy is indicated in larger tumours (>4 cm), residual neck disease, extracapsular spread from lymph nodes, recurrent disease and adenoid cystic carcinoma.

Mucoepidermoid tumours: Mucoepidermoid tumours are the most common major salivary gland tumour (4–9%) and more than 90% occur in the parotid gland. They tend to present in patients between 30 and 50 years old and are divided into low, intermediate and high grade. Survival depends on the grade and is around 86% for low-grade and 22% for high-grade tumours.

Acinic cell carcinomas: These most commonly occur in the parotid gland and have a peak incidence in the fifth decade. Lymph node metastases are present in approximately 10% of tumours and the 5-year survival rate is 90% at 5 years and 55% at 20 years. The majority of acinic cell carcinomas are low-grade. Post-operative radiotherapy is given less readily as it adds no survival advantage.

Adenoid cystic carcinoma are characterized by slow growth and classically a high incidence of peri-neural infiltration and often patients present with intractable pain.

Carcinoma ex-pleomorphic adenoma: These tumours occur in patients who have been treated for multiple recurrences of a pleomorphic adenoma. They are better described as malignant mixed tumours as there are a certain proportion that occur in patients who have had no pre-existing lesion. The prognosis is poor with a 5-, 10- and 15-year survival of 40%, 24% and 19%, respectively.

Thyroid masses

Thyroid masses are not only a common presenting complaint, but also a common incidental finding on scans performed for other reasons. Only 10–15% of thyroid nodules are malignant. Clinical assessment should include a detailed history, including detail about symptoms of thyroid over/under-activity, previous neck surgery or radiation therapy and a family history of thyroid disease or multiple endocrine neoplasia (MEN). Clinical examination should include palpation of the neck; thyroid nodules characteristically move with swallowing (see Figure 3). Assess whether there is a discrete lump within the gland or whether the whole gland is enlarged (which would suggest a thyroiditis as a cause). Fibreoptic examination of the larynx confirms normal vocal cord movement which provides information about the likelihood of malignancy. Thyroid function tests should routinely be performed. The first line of investigation should be an US, which will provide information about whether it is diffuse

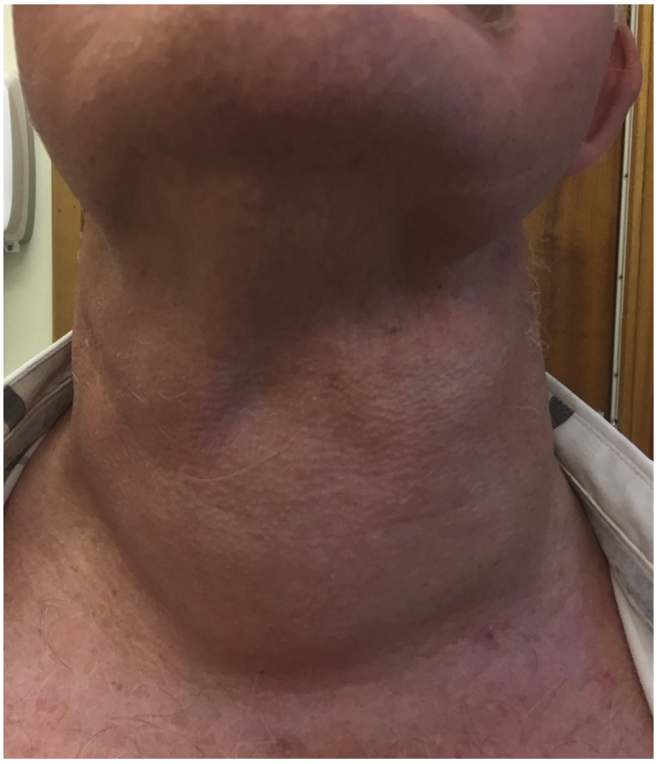

Figure 3 Thyroid goitre. These are often longstanding. Note how the larynx is displaced to the right side. This mass moves when the patient swallows.

enlargement of the thyroid or nodular disease and will also provide information about the likelihood of malignancy.

Simple thyroid goitre

This condition can be due to diffuse enlargement (often due to iodine deficiency), multiple nodules (multinodular goitre) or autoimmune conditions (discussed later). If thyroid function tests are normal and US confirms the benign nature of the disease, management can be conservative. If the patient is experiencing symptoms from pressure effects (often difficulty with swallowing, breathing or cosmetic changes), surgical removal can be offered. CT scanning is often performed preoperatively to confirm the size and extent of the goitre, retrosternal extension and effect on the airway: this is of particular interest when planning peri-operative airway management.

Thyroid tumours

Thyroid cancer is a rare cancer affecting 5 in 100,000 women and 2 in 100,000 men; it is the most common endocrine cancer but makes up only 1% of all malignancies.[5]

Assessment

As with most other thyroid presentations, assessment of the patient who presents with a single thyroid nodule should be with a US assessment. The grading system detailed in Table 1 then classifies lesions according to the number of adverse features identified. When indicated, any patient with adverse radiological features should go on to have a FNA. Again, the findings are graded according to the presence or absence of abnormal features and graded according to the thyroid FNA diagnostic categories (detailed in Table 1). Further investigations and treatment are based on these two core investigations, making USS ± FNA the bedrock of the management of suspected thyroid cancer. The British Thyroid Association 2014 Guidelines provide a detailed account of the assessment, investigation and management of thyroid cancer.[6]

Differentiated thyroid cancer: This is the most common type of thyroid cancer and has an excellent prognosis. The two most common variants are papillary and follicular. Most patients are diagnosed following an indeterminate US and FNA finding (often thy 3 and thy 4) which has led to a diagnostic hemithyroidectomy. Surgical excision, in some cases followed by radioactive iodine therapy or even external beam radiotherapy, forms the basis of the management of these cancers. If the tumour is smaller than 1 cm then a diagnostic hemithyroidectomy provides histological information and may be the only treatment required if there are no adverse tumour or patient factors. Patients with tumour of more than 4 cm or those with multifocal disease, extrathyroidal spread, involved nodes or

Ultrasound and cytological grading for thyroid nodules

U grading	**U1**	**U2**	**U3**		**U4**	**U5**
Ultrasound features	Normal thyroid tissue	Halo Iso-echoic or mildly hyper-echoic Cystic change ± ring down sign Microcystic.spongiform Peripheral egg shell calcification Peripheral vascularity	Homogenous Hyper-echoic Solid, halo Equivocal echogenic foci Cystic change, mixed/central vascularity		Solid Hypoechoic or very hypoechoic Disrupted peripheral calcification Lobulated outline	Solid hypoechoic Lobulated or irregular outline Microcalcification Globular calcification Intranodular vascularity Shape (taller>wide) Characteristic associated lymphadenopathy
Action	No follow up	No follow up: no FNA needed unless high clinical suspicion of thyroid cancer	FNA		FNA	FNA
Thy grading	**Thy 1**	**Thy 2**	**Thy 3F**	**Thy 3A**	**Thy 4**	**Thy 5**
Cytological features	Non-diagnostic	Non-neoplastic	Follicular lesion	Atypia present	Suspicious of thyroid cancer	Diagnostic of thyroid cancer
Action	Repeat FNA cytology	No follow up if no suspicious US eatures and no clinical suspicion of thyroid cancer	Diagnostic hemithyroidectomy. Consider total thyroidectomy in lesions >4 cm where incidence of malignancy is higher	Repeat US and FNA. If second Thy 3A, discuss at MDT and consider diagnostic hemithyroidectomy	Diagnostic hemithyroidecomy	Appropriate further investigations for staging. Total thyroidectomy and central node clearance in appropriate high-risk patients

Reproduced from Mitchell, A.L., Gandhi, A., Scott-Coombes, D. and Perros, P. (2016) 'Management of thyroid cancer: United Kingdom National multidisciplinary Guidelines', *Journal of Laryngology and Otology*, 130, pp. S150-S160.

Table 1

metastatic spread should have a completion thyroidectomy in preparation for postoperative treatment with radioactive iodine. Patient with tumours measuring between 1 and 4 cm and no adverse features can safely be managed by hemithyroidectomy alone, but practice varies according to patient and surgeon preferences. Radioactive labelled I^{131} is recommended in those patients with adverse features such as larger tumour size (>4 cm), extrathyroidal extension, lymph node involvement or adverse histological features. Patients are given a low iodine diet 2 weeks before treatment and pre-treatment thyroid hormone withdrawal has been largely replaced with recombinant TSH therapy. External bean radiotherapy is reserved for those with unresectable tumours or those with residual disease following surgical excision. Following radioactive iodine therapy, a regime of TSH suppression is followed; 9–12 months following treatment, the patient undergoes dynamic risk stratification to determine their response to treatment and to guide their ongoing follow up requirements. Measurement of serum thyroglobulin is central to the follow up of these patients.

Medullary thyroid cancer: This is a rare form of thyroid cancer; more than a quarter are familial and associated with multiple endocrine neoplasia (MEN). The initial assessment includes thyroid US and FNA: when medullary thyroid cancer is diagnosed, patients should undergo a range of other investigations to rule out MEN such as calcitonin levels, urinary catecholamine and RET proto-oncogene. All patients should have a total thyroidectomy and clearance of the central neck nodes. Patients often require ipsilateral or bilateral lateral neck dissections. Following genetic screening, if any family members are shown to be positive for RET proto-oncogene they should have a prophylactic total thyroidectomy. Lifelong follow up is required for all patients and regular measurement of serum calcitonin is of vital importance.

Anaplastic thyroid cancer: This is a highly aggressive, fast growing tumour which is very rarely curable. Often adequate palliation is the main aim of treatment. Total thyroidectomy may be curative for only very small tumours. External beam radiotherapy with chemotherapy may be of limited use for disease control. The main aim of treatment is airway management, particularly at the end of life.

Paediatric thyroid nodules: Thyroid nodules are much less common in children as compared to the adult population. If present, they are much more likely to be malignant and so investigation should be undertaken in all. A full history including family history of thyroid disease (familial non-medullary thyroid cancer, MEN 2, Cowden syndrome, familial adenomatous polyposis and Gardner syndrome), radiation exposure and any iodine deficiency should be noted. All children should undergo a complete head and neck examination, thyroid function tests and an US. If the US shows abnormal or indeterminate features, further investigation is necessary including FNA cytology. The majority of paediatric thyroid cancers are papillary and there is a higher incidence of neck metastases in children. Due to the aggressive nature of the disease, treatment usually involves total thyroidectomy, central or lateral neck dissection and postoperative radio-iodine ablation. Despite the aggressive nature, the prognosis is good with excellent 10 and 20-year survival rates.

Malignant disease of the neck

Paediatric population

There is a stark difference in the pathologies seen within paediatric and adult head and neck malignancies. In the adult population, squamous cell carcinoma is the predominant pathology whereas this is vanishingly rare in the paediatric population. In children, the majority of malignancies are lymphomas followed by rhabdomyosarcoma, thyroid carcinoma, nasopharyngeal carcinoma, salivary malignancy, neuroblastoma and teratomas.

Lymphoma: This can be grossly divided into Hodgkin's (including moderate sclerosing, mixed cellularity, lymphocyte predominant and lymphocyte depleted) and non-Hodgkin's lymphoma. The difference histologically is the presence of Reed-Sternberg cells in the former. They can both present with progressive cervical lymphadenopathy with extra-nodal disease being more common in non-Hodgkin's lymphoma and normally presenting in a younger age group. Other symptoms can include weight loss, night sweats and fever. Lymph node biopsy is the gold standard for histological diagnosis as FNA may produce insufficient material for accurate analysis. Many centres now provide core biopsy service which may provide sufficient histological information for diagnosis. Radiological techniques including CT scanning, PET-CT and MRI will be required for staging prior to treatment which will take the form of chemotherapy with or without radiotherapy in the first instance.

Rhabdomyosarcoma is the second most common malignancy in the paediatric population, with 35% occurring in the head and neck. Staging involves CT, MRI and US scans and the treatment is usually multimodal involving surgery, chemotherapy and radiotherapy.

Adult population

Head and neck cancer is a large group of cancers originating from sites including the voice box (larynx), throat (oropharynx, hypopharynx and upper oesophagus), mouth (oral cavity), nose (nasal cavity, sinus and nasopharynx) and salivary glands. Over 90% of head and neck cancer (HNC) are squamous cell carcinomas (SCC). There are around 7000 new cases in the UK each year, with the most common primary site being the larynx. It is more common in males than females and incidence increases with increasing age. There has been a recent rise in the incidence of head and neck cancer secondary to human papilloma virus (HPV) infection, which has led to HNC being diagnosed more commonly in younger patients who do not smoke. HNC is more commonly diagnosed in patients with characteristics associated with lower socioeconomic status such as unemployment or lower educational attainment, but much of this difference is due to increased rates of smoking.

A neck lump is an increasingly common presenting symptom of squamous cell carcinoma of the head and neck. Indeed, any patient over the age of 35 presenting with a new presentation of neck lump should be considered to have head and neck cancer until proven otherwise. In the same way as any neck lump, a full history and examination should take place, but a particular

emphasis must be placed on identifying symptoms of head and neck cancer and patients at high risk of the disease. Symptoms of hoarse voice, dysphagia, odynophagia, otalgia (especially if unilateral), haemoptysis or bleeding, pain, weight loss and anorexia may point towards the site of a primary lesion. Detailed smoking and alcohol history must be taken as well as information about comorbidity status and social circumstances which may be central to the treatment decision. Examination should include all sites of the head and neck and a fibreoptic examination of the upper aerodigestive tract is mandatory. This assessment may find a likely primary source of disease which leads to a very different investigation trajectory than if no primary site is found.

Carcinoma with unknown primary

In a small number of cases, the initial assessment of the patient does not identify the primary site of disease. In this situation, the patient should undergo core biopsy of the affected node; this allows us to ascertain the HPV status as this information is not available from FNA alone. The patient should also undergo cross sectional scanning of the head and neck. CT scanning may identify a primary site, but is arguably inferior to MRI for identifying small volume disease. If no primary is found, the patient goes on to have a positron emission tomography-computed tomography (PET-CT) which uses the fluorodeoxyglucose (FDG) tracer to highlight cells with a more rapid turnover (see Figure 4). PET-CT may be useful in identifying a possible primary site or at least provide guidance for targeted biopsies. The patient then undergoes an examination under anaesthetic of the upper aerodigestive tract and, depending of the outcome of the investigations thus far, a bilateral tonsillectomy. If this does not yield a primary, and the core biopsy is HPV positive, the patient may then undergo a mucusectomy of the base of tongue. Tongue base mucusectomy involves the transoral robotic or laser removal of the lymphoid tissue at the base of the tongue before sending it for histological analysis. If these investigations identify the primary site of disease, treatment can proceed as normal. If no primary site is found, the patient undergoes either surgical management of the neck with postoperative (chemo)radiotherapy or primary (chemo)radiotherapy to the neck. The role of total mucosal irradiation to the head and neck is uncertain, but excellent overall control rates can be expected for this patient group.[7]

Carcinoma with known primary

When a patient presents to the head and neck clinic with a new onset of neck mass and it is thought to be SCC of the head and neck, examination may identify a likely primary site (such as a unilateral bulkiness to the base of tongue or an exophytic lesion on the vocal cords). In this case, investigations should be performed expeditiously and focus on tissue sampling of the likely primary tumour. FNA is preferable under US control. CT or MRI of the neck should be performed, and the modality used varies depending on local availability and expertise. If the imaging confirms the suspicions for a primary site, the patient goes on to have to full examination of the upper aerodigestive tract. The first priority of this procedure is to assess the size and extent of disease, the involvement of the structures of the head and neck, the suitability for surgical resection, the presence of second primaries and to obtain a tissue diagnosis. If the biopsies confirm the presence of SCC, the chest should be imaged with CT to complete the staging of the patient before a treatment decision is made. All patients are staged using the tumour, node, metastasis (TNM) staging system.[8] The tumour (T) stage concerns the size, position and local invasion of the tumour, and is graded between T1 (small tumours) and T4 (large, invasive tumours). The nodal (N) stage depends on the number, position and size of the lymph nodes that the cancer has spread to and is graded between N0 (delineates no detectable spread to lymph nodes) and N3. Patients with HNC rarely have a distant metastasis at first presentation. If no metastasis is present, the tumour is graded M0, if the

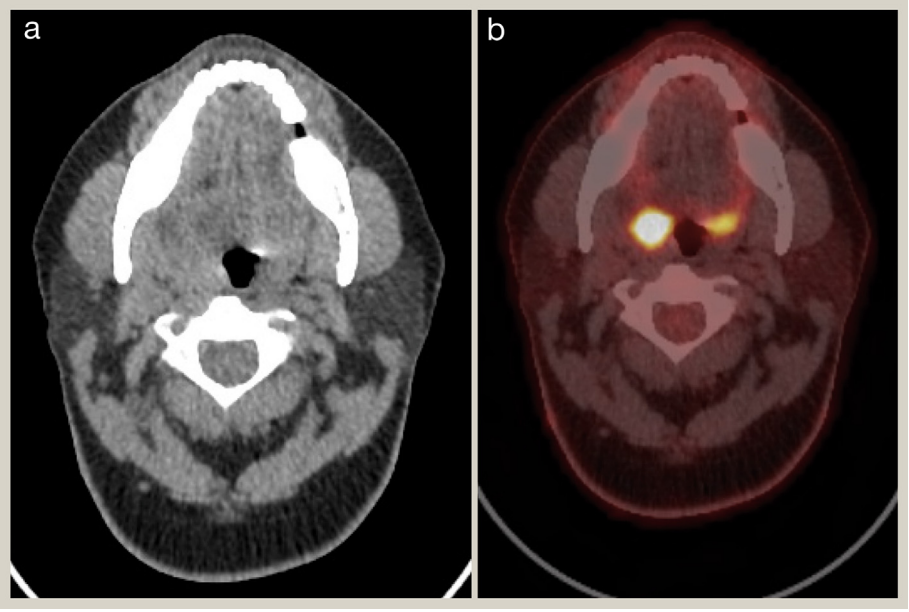

Figure 4 CT and PET CT scan showing a right tonsillar tumour. The tumour is not obvious on the CT scan, but lights up with clear asymmetry on the PET-CT scan.

cancer has spread to distant structures (most often the lung) it is graded M1.

Branchial cyst

A branchial cyst is a benign head and neck lesion; however, clinicians should maintain a high index of suspicion for malignant disease. If the patient is more than 35 years old, has risk factors or if the cyst is not solitary it should be treated as a cancer until proven beyond all reasonable doubt. This is particularly true if there are solid areas on imaging. All branchial cysts should have a core biopsy together with an MRI of the neck. If there is high clinical concern, one may consider a PET CT even in the presence of a benign core biopsy. Often in these situations the branchial cyst is found to be malignant however if not the patient should be offered treatment. Surgical excision will provide diagnostic certainty but has risks; nevertheless, it should be routinely offered, especially in the setting of any clinical suspicion. Surgery for branchial sists may require removal of some of the surrounding lymph nodes.

Treatment of head and neck cancer

Following identification of the site and stage, it is mandatory in the UK for patients to be discussed as part of the head and neck cancer multidisciplinary team (MDT).[9] Treatment options for the patient consist of surgery, radiotherapy (with or without chemotherapy) or palliation/best supportive care. If a patient has presented to the clinic with a neck lump, then it is likely that their head and neck cancer is already at an advanced stage. Overall, patients with advanced disease should be treated with a combination of treatments, either chemotherapy and radiotherapy or surgery with postoperative radiotherapy. The treatment decision rests on a variety of clinical and non-clinical factors, but of primary importance is the site and stage of disease, the effect on swallow and dietary function, the general performance status of the patient, airway issues and the personal values and preferences of the patient.

Non-surgical management of head and neck cancer

Radiotherapy to the head and neck allows preservation of the structures therein, which has obvious cosmetic and functional benefits. Survival is increased if chemotherapy is added to radiotherapy as a primary treatment or in the postoperative setting with adverse pathological features.[10,11] However, radiotherapy has significant side effects, which are dose dependent and worsened when combined with chemotherapy.[12] The main short-term side effects include skin reactions and mucositis; in the long term, pharyngeal stenosis leads to difficulty swallowing and laryngeal scarring causes hoarse voice. In the most severe cases, this may lead to a requirement for tracheostomy. Dysphagia often leads to the need for supplemental feeding through the course of treatment in the form of a nasogastric or gastrostomy tube; in some patients these feeding tubes are required in the longer term.

Surgical management of head and neck cancer

There is a range of surgical options available to the patient with head and neck cancer. Large open resections used to form the mainstay of treatment of head and neck cancer; although this has largely been replaced with chemoradiotherapy in many tumours, there has been a more recent surge in the popularity of transoral techniques using either laser or robotic surgery. These techniques allow removal of the tumour with less functional side effects of treatment. When combined with the increasing recognition of the short and long-term side effects on non-surgical management, the benefits of surgery, even large open resections, have gained favour: with effective postoperative rehabilitation, good postoperative swallow and voice outcomes can be obtained for patients. Surgical management of the neck may allow accurate pathological staging of disease and may therefore limit the field required for postoperative radiotherapy.

With the recent advances in transoral techniques the modern management of the patient with head and neck cancer includes a host of surgical and non-surgical techniques with varying effects on survival, swallow and voice. Ongoing trials compare these treatment and assess their role in the de-escalation of treatment, especially those with HPV positive disease. There is an increasing recognition of the importance of postoperative function and quality of life. This makes the decision for whether and how to treat the patient with head and neck cancer fraught with difficulty.

Conclusion

A presentation of neck lump in the ENT clinic can be a sign of a myriad of benign and malignant pathologies. The treating clinicians should maintain a high index of suspicion in order not only to identify those at high risk of serious disease, but to expedite its accurate tissue diagnosis and staging so treatment can be commenced. ◆

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