The role of radiology in head and neck tumours in children

Claire Lloyd and Kieran McHugh

Department of Radiology, Imperial College Healthcare NHS Trust, Charing Cross Hospital Campus, Fulham Palace Road, London, SW6 8RF, UK; Department of Radiology, Great Ormond Street Hospital for Children, Great Ormond Street, London, WC1N 3JH, UK

Corresponding address: Dr Claire Lloyd, Department of Radiology, Imperial College Healthcare NHS Trust, Charing Cross Hospital Campus, Fulham Palace Road, London, W6 8RF, UK.

Email: clairelloyd@doctors.org.uk

Date accepted for publication 30 December 2009

Abstract

Head and neck malignancy is rare in children. However, distinguishing malignant tumours from the more common and numerous benign causes of neck masses in childhood is crucial as many malignant conditions have an excellent prognosis with appropriate oncological management. Ultrasound, computed tomography and magnetic resonance imaging all have crucial roles in the diagnosis of head and neck malignancy in children and there is an emerging role for positron emission tomography, particularly in the management and follow-up of lymphoma. We describe the imaging appearances of the common malignant tumours arising in the extracranial head and neck in children, focusing on lymphoma, rhabdomyosarcoma and nasopharyngeal carcinoma. The clinical presentation and radiological appearances of benign tumours in the head and neck in children may overlap with those seen in malignant disease. We describe the imaging appearances of juvenile angiofibroma, vascular abnormalities involving the extracranial head and neck and cervical teratomas. Advances in both imaging techniques and cancer staging systems, many of the latter aimed at avoiding overtreatment and treatment-related complications, will lead to an increasingly central role for imaging in childhood head and neck cancer.

Keywords: CT; MRI; lymphoma; nasopharyngeal carcinoma; rhabdomyosarcoma; juvenile angiofibroma; teratoma.

Introduction

Neck masses in children commonly have an inflammatory, infective or congenital cause. Neoplastic lesions are less common with only 5% of childhood cancers arising in the head and neck region\(^1\). However, a number of benign and malignant tumours may occur in the extracranial head and neck and an awareness of their occurrence and imaging appearances is important for any radiologist involved in imaging children. Early diagnosis of head and neck malignancy is paramount as many of these cancers are readily treatable, and often curable, with current oncological management strategies. Imaging has an important role in characterizing and assessing the extent of head and neck mass lesions and, in the case of malignant disease, in detecting the presence of metastatic spread and in following response to treatment. In addition, imaging may be used to guide fine-needle aspiration (FNA) or needle biopsy. In this review, we describe the imaging techniques used to assess neoplastic lesions of the head and neck in children and then review the common malignant and benign tumours and their imaging findings.

Imaging techniques

Ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) provide the mainstay of imaging head and neck tumours in children. Ultrasound is particularly useful in examining superficial masses whereas CT and MRI are used to delineate deeper lesions, particularly those involving the skull base and central nervous system (CNS). Plain radiographs have
a very limited role although a soft tissue mass or bony involvement may be demonstrated. Barium swallow examination may be indicated if symptoms related to swallowing are present.

**Ultrasound**

US is widely used as the initial imaging technique in the assessment of extracranial head and neck masses in children, largely due to its ready availability and avoidance of ionizing radiation. The frequent superficial location of head and neck tumours make them readily accessible to US examination. If necessary, the examination can be performed at the bedside and sedation is rarely necessary.

Technical developments in high-resolution gray-scale (B-mode) ultrasound have improved the ability of US to characterize the internal architecture of masses, beyond simply distinguishing cystic from solid lesions. The child should be examined, when possible, in the supine position with the neck hyper-extended. A 5–8 MHz sector probe is useful in examining deeper regions and in attaining an overview of the anatomy. A higher frequency (7–12 MHz) linear probe is useful in providing more detailed views of superficial structures. The additional use of colour Doppler ultrasound (CDUS) and power Doppler allow assessment of vascularity within the lesion, particularly helpful in the assessment of haemangioma and vascular malformations and also contributory in the assessment of enlarged lymph nodes.

**Computed tomography**

Current multidetector computed tomography (MDCT) technology allows rapid and detailed examination of the entire neck with the ability to produce multiplanar reformatted images. However, as with CT examination of any other region of the child’s body, the radiologist should be mindful of minimizing radiation dose and adhering to the ALARA (As Low As Reasonably Achievable) principle. Assuming the airway is not compromised, the child should be scanned supine with the neck in the neutral position. If the likelihood of airway compression is high, anaesthetic opinion should be sought regarding the need for intubation. Younger children will usually require sedation to minimize movement artefacts. The region scanned extends from the top of the sphenoid sinus to the sternoclavicular joints. A bone algorithm, in addition to a standard soft tissue algorithm, is particularly important for tumours that may involve the skull base. The presence of calcification or fat within the lesion is well demonstrated on CT and helps with lesion characterization. Intravenous contrast administration (1–2 ml/kg) is mandatory to delineate the mass, or lymphadenopathy, from adjacent normal structures. Enhancement patterns maybe helpful in characterizing some masses, such as vascular tumours.

**Magnetic resonance imaging**

The superior soft tissue resolution of MRI makes it an excellent modality in imaging head and neck masses. It is particularly useful in delineating intracranial extension of disease. The examination is performed with the child supine in quiet respiration. Infants can often be fed prior to examination and swaddled to minimize movement artefact. Slightly older children will often need sedation or anaesthesia, particularly when there is any suspicion of airway compromise. With explanation and gentle persuasion, children over the age of 6 years can often remain still for the examination without recourse to sedation. A head or neck coil should be chosen, depending on the location of the mass. Standard examination should include a T2-weighted fast spin echo (FSE) sequence in axial and coronal planes, a T2-weighted fat suppression or inversion recovery sequence and a plain T1-weighted FSE or spin echo (SE) sequence. In evaluating mass lesions, a further fat-saturated T1-weighted SE sequence following gadolinium administration will often improve characterization of the mass. Additional diffusion-weighted imaging appears to also have a role in characterizing head and neck masses. A recent study found a significant difference in apparent diffusion coefficient (ADC) values between malignant tumours and benign lesions with a sensitivity and specificity for differentiating benign from malignant lesions of 94% and 91% respectively when a threshold ADC value was used.

**Positron emission tomography**

Positron emission tomography (PET) provides whole-body functional imaging with good spatial resolution. [18F]Fluorodeoxyglucose ([18F]FDG) PET has a wide range of applications in staging oncological disease and monitoring response to treatment. Moreover, the increasing availability of PET-CT allows improved localization and definition of disease activity. The role of PET and PET-CT in childhood malignancies continues to be defined, but it is widely used in the staging and follow-up of lymphoma and may also have a role in soft tissue sarcomas.

**Malignant tumours**

**Lymphoma**

Lymphoma is the commonest malignancy involving the extracranial head and neck in children. Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL) together account for 10–15% of all childhood malignancies in developed countries, with only leukaemia and brain tumours occurring more commonly. Most children with head and neck lymphoma present with cervical lymph node involvement. NHL may present with extranodal disease involving the Waldeyer ring (the lymphatic tissue found in the nasopharynx, base of tongue, tonsils...
and soft palate) (Fig. 1) or the jaw. However, extranodal disease in the head and neck in childhood NHL is less common than in other parts of the body. Extranodal involvement of the head and neck in children with HL is very rare.

Enlarged cervical lymph nodes in children are benign and reactive in the vast majority of cases, usually a consequence of viral or bacterial infection of the upper respiratory tract. However, persistent (>6 weeks duration) nodal enlargement, particularly if painless and involving the posterior triangle or supraclavicular region is of clinical concern[7]. The superficial cervical lymph nodes are readily accessible to examination by US and in general US has good sensitivity and specificity for distinguishing benign, reactive lymph nodes from those infiltrated with malignant disease. Sonographic features of nodal malignancy include increased size, a round shape, decrease in the internal echogenicity and loss of the normal echogenic hilum[8]. Although the risk of malignancy increases in nodes measuring more than 3.0 cm in longest diameter[9,10], size alone cannot be used as an absolute criterion for distinguishing benign from reactive nodes.

Historically lymphomatous nodes were described as pseudocystic, hypoechoic with posterior enhancement[11]. However, modern high-resolution, high-frequency transducers reveal internal reticulation and the pseudocystic appearance is less likely to be seen[12]. Assessment of intranodal vascularity with CDUS improves detection of malignant infiltration in superficial nodes. The presence of peripheral, subcapsular vessels, distortion or displacement of the intranodal vessels and focal areas of absent perfusion are suspicious for malignant involvement[13]. The presence of these gray-scale and CDUS features, particularly in combination, should alert the sonographer and prompt consideration of FNA or biopsy.

In children with lymphoma involving the cervical lymph nodes, disease is usually staged with intravenous contrast-enhanced CT of the neck (to include the Waldeyer ring), chest, abdomen and pelvis. Lymph nodes measuring less than 10 mm in short axis diameter are considered normal. In the current European study of childhood and adolescent classic HL (EuroNet-PHL-C1), lymph nodes measuring more than 20 mm in short axis are considered to be involved, and the results of FDG-PET are taken into account for intermediate-sized (i.e. 10–20 mm in diameter) lymph nodes, with PET-positivity indicating involvement[14]. The soft tissue resolution of MRI produces excellent depiction of the cervical lymph nodes and there is a trend towards more MRI lymphoma staging but lung assessment at first diagnosis must be with CT. Consequently, CT is generally the modality used at initial diagnosis and MRI is more specifically used in those cases in which there is concern about disease extension into the CNS or primary CNS involvement[15]. MRI is also sensitive for the detection of bone marrow involvement.

Figure 1  NHL involving the Waldeyer ring. Contrast-enhanced axial CT section showing circumferential soft tissue thickening of the nasopharynx (arrows). This proved to be extranodal NHL of Waldeyer ring.

FDG-PET and PET-CT are increasingly important in staging, evaluating treatment response and monitoring for relapse of childhood lymphoma, replacing gallium scans for the assessment of metabolically active disease[16]. FDG-PET can better distinguish between active disease and fibrotic/necrotic residual masses and therefore has better specificity than CT/MRI. Both FDG-PET and PET-CT have been shown to alter disease staging in paediatric lymphoma, the latter modality in up to one-third of patients[17]. A negative PET-CT examination in the routine follow-up of lymphoma has a very high negative predictive value, strongly suggesting the absence of recurrence[17,18].

**Hodgkin lymphoma**

HL is the form of lymphoma more commonly seen in adolescents. Histologically it is characterized by the presence of Reed–Sternberg cells. The World Health Organization classification divides HL into the rare nodular lymphocyte predominant (LP) form and the more common form, classic HL. Classic HL is further subdivided into nodular sclerosis (NS), lymphocyte rich (LR), mixed cellularity (MC) and lymphocyte depletion (LD) subtypes. NS is the most common subtype of HL with the incidence of each subtype varying with age and sex.

HL typically presents with enlarged, painless cervical lymph nodes (Fig. 2). As classic HL involves contiguous lymph node groups, coexistent mediastinal involvement is common[19]. Staging of disease is performed according to the Cotswold modification of the Ann Arbor staging system (Table 1). Disease staging aims to define risk groups, which in turn determine treatment. ‘Favourable’ disease comprises localized (stages I and II) asymptomatic nodal disease; advanced (stages IIIB and IV) nodal disease is considered unfavourable.
In EuroNet-PHL-C1, FDG-PET is formally integrated into staging and response assessment\(^{14}\). The study aims to further optimize first- and second-line therapy for childhood HL to avoid over-treatment and decrease long-term complications. All patients undergo initial FDG-PET along with conventional staging with CT/MRI at diagnosis except in oncological emergencies. Following two cycles of chemotherapy, treatment response is assessed with an early follow-up PET examination. Patients judged to have achieved an ‘adequate response’ (either those in complete remission on conventional imaging or those in partial remission but with a negative PET study) receive no further therapy. Patients with an inadequate response (those not in complete remission and with at least one initially involved region that remains PET positive) after two cycles of chemotherapy receive further treatment (either radiotherapy, chemotherapy or both depending on initial risk group).

**Non-Hodgkin lymphoma**

NHL is more common than HL in children under 10 years of age. The WHO classification divides paediatric NHL into four subtypes: Burkitt lymphoma (BL), diffuse large B-cell lymphoma (DLBCL), anaplastic large cell lymphoma and lymphoblastic lymphoma. NHL presenting with cervical lymph node involvement is frequently disseminated. This contrasts with HL in which disease is frequently confined to the neck and chest. Staging systems are designed to assess tumour load and divide patients into those with ‘limited’ disease (stages I and II) and those with ‘extensive’ disease (stages III and IV). B symptoms do not form part of the staging system. The St Jude staging system is commonly used (Table 2).

### Table 2 St Jude staging classification for NHL

| Stage    | Definition                                                                 |
|----------|---------------------------------------------------------------------------|
| Stage I  | Involvement of a single lymph node region or lymph node structure          |
| Stage II | Involvement of two or more lymph node regions on the same side of the diaphragm |
| Stage III| Involvement of lymph nodes regions or lymph nodes structures on both sides of the diaphragm |
| Stage IV | Involvement of extranodal sites beyond E sites                            |

**Annotations to stage definitions**

| Annotation | Definition                                                                                   |
|------------|---------------------------------------------------------------------------------------------|
| A          | No B symptoms                                                                               |
| B          | At least one of the following systemic symptoms:                                             |
|            | - Inexplicable weight loss of more than 10% within the last 6 months                        |
|            | - Unexplained persisting or recurrent temperature above 38°C                                |
|            | - Drenching night sweats                                                                    |
| E          | Involvement of a single extranodal site contiguous or proximal to a known nodal site        |

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BL occurs in endemic, sporadic and immunodeficiency-associated forms. It is a B-cell lymphoma with one of the fastest growth rates amongst all malignancies, doubling in size in 12–24 h. Endemic BL is found in young African children. There is an association with Epstein–Barr virus (EBV) infection although the exact relationship remains unclear. Endemic BL commonly presents as a large jaw mass involving and eroding the mandible, maxilla and developing dentition. Coexistent abdominal involvement and peripheral lymphadenopathy are common. Sporadic BL is found worldwide; like endemic BL there is an association with EBV infection. It is an extranodal disease and although head and neck involvement is rare, pharyngeal and tonsillar tumours can occur. Immunodeficiency-associated BL is seen in patients with HIV, congenital immunodeficiencies and transplant recipients.

Involvement of the head and neck region in children with DLBCL and anaplastic large cell lymphoma usually occurs in the context of diffuse disease. Bulky cervical and supravacular lymphadenopathy is characteristic of lymphoblastic lymphoma, usually with concurrent anterior mediastinal disease.

**Rhabdomyosarcoma**

Rhabdomyosarcoma (RMS) is the commonest soft tissue sarcoma in children below the age of 15 years and accounts for 8% of all childhood malignancies. Most cases occur in the first decade of life and there is a slight male predominance. RMS arises from mesenchymal precursor cells and three histological types are recognized: embryonal, alveolar and pleomorphic. Staging of disease is complicated, with some historical variation in systems used by North American and European cancer groups but these have unified in recent years. In essence the disease is staged both pre-operatively and post-operatively. Incorporated into the staging systems are disease site and extent of residual disease following primary surgery, and a traditional TNM system. The Intergroup Rhabdomyosarcoma Study (IRS) trials have established the prognostic value of the rhabdomyosarcoma staging system with three prognostic groups (low, intermediate and high risk), defined according to staging and histological subtype. Favourable disease sites include the orbit or non-parameningeal head and neck sites. Tumours of the embryonal subtype have a better prognosis. The prognostic group is used to guide multimodality treatment with surgery, radiotherapy and chemotherapy. Refinements in risk stratification and treatment have led to dramatic improvements in prognosis, with survival rates increasing from approximately 25% in the 1970s to approximately 70% in IRS IV. Radiology has a key role in the initial staging of children with RMS and in their long-term follow-up and assessment of treatment-related complications.

The head and neck is the primary site of disease in 40% of children with RMS. Location of disease within the head and neck is categorized into orbital, parameningeal and non-parameningeal sites. Specific sites of parameningeal disease are the nasal cavity, paranasal sinuses, pterygoid fossa, nasopharynx and middle ear. Children may present with a mass lesion, pain, proptosis, deafness, recurrent sinusitis or cranial nerve palsies.

MRI is the technique of choice in the assessment of head and neck RMS, providing excellent depiction of the mass and its anatomical relations, in particular the assessment of intracranial extension. The mass typically demonstrates high signal intensity on T2-weighted sequences and is isointense or minimally hyperintense to skeletal muscle on T1-weighted sequences. A fat-suppressed sequence is particularly helpful in evaluating orbital disease. Following intravenous contrast administration there may be moderate-to-intense enhancement (Fig. 3), occasionally similar to that seen in juvenile angiofibroma. Accurate assessment of disease extent and volume is crucial in pre-operative planning and in defining the field for radiotherapy. CT, although less effective in determining the extent of the mass, is important in demonstrating bony involvement and many tumours at the skull base merit both CT and MRI for full evaluation at diagnosis. Up to 14% of all RMS patients have metastatic disease (stage IV) at presentation. Staging is completed with chest CT, abdominal ultrasound and bone scintigraphy. MRI may be more sensitive than bone scan in the demonstration of bony metastases but this is uncertain at present. Rhabdomyosarcomas show variable degrees of FDG uptake, with the head and neck being a high-risk site.

![Figure 3](image-url)  
**Figure 3** Parameningeal RMS in a 6-year-old girl. T1-weighted post-gadolinium axial MRI shows an avidly enhancing mass in the left sphenoid sinus extending into the posterior nasal space and pterygopalatine fossa (arrow). Histologically this mass was rhabdomyosarcoma but similar MRI appearances may be due to angiofibroma.
uptake\textsuperscript{[5]}. The exact role of FDG-PET in RMS has yet to be defined but in a recent prospective trial PET was superior to conventional imaging in detecting lymph node involvement and bone manifestations although there was no difference between modalities in depicting the primary tumour\textsuperscript{[25]}.

Orbital tumours are usually non-invasive and confined to the orbit (Figs. 4, 5). The mass may be intracranial or extracranial with the superonasal part of the orbit being the most common site of disease. The embryonal form is the most common histological subtype in orbital RMS. Lymphatic spread and lymph node metastases are very rare. Treatment usually involves a combination of chemotherapy and radiotherapy. Although surgical debulking may be indicated, improvements in chemotherapy and radiotherapy have rendered traditional surgical exenteration obsolete. Prognosis is excellent with a 3-year survival rate greater than 90\%\textsuperscript{[20]}.

Parameningeal tumours tend to be large and invasive, with intracranial extension commonly seen (Fig. 6). Cervical lymph node metastases may be seen, occurring in 21\% of patients in one series\textsuperscript{[26]}. Improvements in surgical techniques have allowed more parameningeal tumours to be resected with clear margins but adjuvant chemotherapy and radiotherapy are used to achieve local control and improve survival. Prognosis of parameningeal disease is worse than that of orbital and non-parameningeal disease. In the fourth IRS trial, overall 5-year survival for patients with parameningeal disease treated with radiotherapy was 73\%\textsuperscript{[27]}.

Follow-up imaging should be performed no sooner than 6 weeks post-treatment to avoid confusion between post-treatment change and residual disease. The same imaging modality should ideally be used for pre-treatment and post-treatment assessment of disease. Enhancement in the tumour bed more than 6 weeks after treatment is suspicious for residual tumour but further follow-up imaging rather than early biopsy is often more appropriate in this circumstance\textsuperscript{[28]}.

**Nasopharyngeal carcinoma**

Nasopharyngeal carcinoma (NPC) in children is rare with only 5–12\% of cases of NPC occurring in patients aged 30 years or younger. Non-epithelial tumours

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**Figure 4** Orbital RMS. T2-weighted axial MRI shows a predominantly intracranial right-sided orbital mass of intermediate signal intensity causing proptosis of the right globe.

**Figure 5** Orbital RMS. T1-weighted post-gadolinium axial MRI in a different patient showing avid enhancement of a left orbital mass proven to be RMS.

**Figure 6** Parameningeal RMS. Coronal T1-weighted post-gadolinium image shows a large avidly enhancing mass involving the skull base.
(e.g. lymphoma, RMS, angiofibroma) occur more commonly in the nasopharynx than NPC in this age group. The median age at diagnosis for childhood NPC is 13 years and there is a slight male predominance. Wide geographic and ethnic variation is seen in the distribution of NPC, in adults and children, implicating genetic and environmental factors, including a recognized association with EBV infection[29].

There are several features of childhood NPC that distinguish it from NPC in adulthood. Advanced locoregional disease at presentation is common in children with over 80% presenting with bulky disease, often with skull base erosion, cranial nerve involvement and regional lymph node involvement. Presentation is most commonly with an upper cervical mass but pain, conductive deafness, sinus problems or cranial nerve palsies can also bring the child to medical attention. Histologically, most NPC in children is undifferentiated carcinoma (WHO subtype III) characterized by undifferentiated round cells with prominent nucleoli. This contrasts with adults in whom keratinizing squamous cell carcinoma is the most common subtype, followed by non-keratinizing epidermoid carcinoma. There is an association between EBV and NPC with the association being stronger in children than in adults and in the undifferentiated form of disease.

CT and MRI are used to assess the extent of disease. MRI is better at demonstrating intracranial extension through the skull base foramina and involvement of the cranial nerves, whereas CT is superior in demonstrating bony erosion. Cervical lymph node involvement may be detected with both techniques. At contrast-enhanced CT, NPC in children is demonstrated as a homogeneously enhancing soft tissue mass with infiltration of adjacent soft tissues and skull base erosion seen in the majority[30]. Necrosis in involved lymph nodes is less common than in adult NPC, possibly a result of the different histological subtypes observed in adults and children. At MRI, the mass is slightly hyperintense to skeletal muscle on T1-weighted and T2-weighted sequences and demonstrates relatively homogeneous enhancement following intravenous gadolinium administration (Fig. 7). Staging CT of the thorax is indicated in childhood NPC.

Treatment of childhood NPC is with high-dose radiotherapy to the head and neck region with chemotherapy using a cisplatin-based regime. Whilst the aim of treatment is cure and prognosis of NPC in children is better than in adults, a proportion of cases relapse. Presentation with metastatic disease is rare but the outlook in these cases is poor.

Other malignant tumours of the paediatric head and neck

Neuroblastoma is the third most common childhood malignancy but only 5% of primary lesions arise in the head and neck. Metastatic involvement of the head and neck region is more common[7]. Presentation may be with a mass, commonly in the lateral neck or retropharyngeal space, cranial nerve involvement or Horner syndrome. Imaging evaluation includes US, CT or MRI (Fig. 8). Metaiodobenzylguanidine (mIBG) scintigraphy is valuable in assessing bone and bone marrow involvement and in following response to therapy. Primary cervical neuroblastoma has a favourable outcome. Localized
Lesions are frequently amenable to complete surgical excision. Adjuvant radiotherapy and/or chemotherapy maybe required for more advanced local disease where complete resection may not be possible.

Retinoblastoma is the most common intraocular neoplasm of childhood. Although an aggressive tumour, early detection and appropriate treatment have resulted in cure rates of over 90%\[31\]. About one-third of tumours are bilateral with multifocal involvement of the eye in one-third of cases\[32\]. Approximately 5–10% of cases are familial. Clinical examination and ultrasound of the orbit are used to establish the diagnosis of retinoblastoma and are invaluable in assessing intraocular disease status. MRI can also depict intraocular tumour spread and is the best modality in delineating extraocular (including intracranial) tumour spread, with important treatment and prognostic implications\[31\]. CT will depict the typically calcified intraocular mass but is not routinely used in disease assessment.

Thyroid cancer in children is rare, accounting for 5–10% of all thyroid cancers. It is most common in female adolescents and presentation is usually with a solitary nodule. Risk factors for developing thyroid cancer include previous neck irradiation and genetic disorders, e.g. familial adenomatous polyposis and Cowden disease. Thyroid malignancy in childhood tends to be well differentiated with papillary thyroid carcinoma predominating. Differentiated thyroid cancers are often more advanced at presentation in children than in adults with cervical lymph node metastases present in more than 50% and pulmonary metastases in 20–30%\[33\]. Despite this, prognosis is excellent with 100% survival for all registered UK cases. Ultrasound-guided FNA of thyroid nodules may be helpful in older adolescents but in younger children the yield is lower and the presence of a thyroid nodule should be considered an indication for diagnostic surgery. The current recommendation of the British Society of Paediatric Endocrinology and Diabetes and the Children’s Cancer and Leukaemia Group (previously known as the United Kingdom Children’s Cancer Study Group) is that confirmed cases of thyroid cancer should be discussed at the adult cancer centre thyroid multidisciplinary team\[34\].

Primary tumours of the salivary glands are uncommon in children although the proportion of malignant lesions compared with benign disease is slightly higher in children than in adults. Most tumours involve the parotid gland. The commonest primary malignancy is mucoepidermoid carcinoma\[35\]. Adenocarcinoma, acinic cell carcinoma and adenoid cystic carcinoma are less common. The salivary glands may also be involved as an extranodal site in NHL. US and MRI are used in the imaging evaluation of salivary tumours.

**Benign tumours**

*Juvenile angiofibroma*

Juvenile angiofibroma (JA) is a highly vascular tumour involving the posterior nasal cavity and nasopharynx. It occurs almost exclusively in adolescent and young adult males. Although JA is a benign tumour, it displays locally aggressive behaviour, eroding the skull base and infiltrating the CNS, and has a tendency to recur following...
The diagnosis is suspected in an adolescent male presenting with nasal obstruction or recurrent epistaxis. A nasopharyngeal mass may be clinically detected. In most cases, the characteristic imaging appearances confirm the diagnosis without the need for biopsy, a potentially dangerous procedure given the highly vascular nature of the tumour. Definitive surgical resection, often using an endoscopic approach, is the gold standard management. Radiotherapy and pre-operative embolization may be used in combination with surgery to treat large tumours, particularly those with intracranial extension.

CT and MRI are used to confirm diagnosis and define tumour extent. MRI better defines soft tissue extent but CT, using a bone algorithm with coronal reconstructions, is particularly valuable in depicting the inevitable bone erosion at the skull base and extension into the sphenoid. This information is crucial to the surgeon in planning their approach. The site of origin of the tumour is key to making the diagnosis. JA classically arises in the pterygopalatine fossa in the recess behind the sphenopalatine ganglion. Growth and expansion into the nasal cavity medially occurs through the sphenopalatine foramen and by erosion of the palatine bone. This characteristic combination of a mass in the nasal cavity and pterygopalatine fossa and erosion of bone behind the sphenopalatine foramen is diagnostic for JA. The mass typically also causes anterior bowing of the posterior wall of the maxillary antrum (Fig. 9). Invasion of the sphenoid occurs as the tumour grows posteriorly along the pterygoid canal to invade and expand the body and greater wing of sphenoid. Extension into the orbit and infratemporal fossa, via the inferior orbital fissure and pterygomaxillary fissure, respectively, may be seen. Intracranial extension is uncommon but clinically significant and should be sought on MRI, usually occurring into the middle cranial fossa via the foramen rotundum or vidian canal.

On MRI, characteristic flow voids are seen within the soft tissue mass and there is marked enhancement following intravenous gadolinium administration (Fig. 10). Both features reflect the highly vascular nature of JA. MRI may better illustrate the extent of soft tissue abnormality than CT and is particularly helpful in detecting recurrence following surgery. Previous series have reported recurrent tumour growth in more than one-third of patients but a more recent series reported recurrence in just 15% of patients, probably reflecting improvements in pre-operative imaging, embolization and surgical techniques. Recurrence is thought to occur due to incomplete resection of tumours with high growth rates. Sphenoid invasion detected on pre-operative imaging is a predictor of recurrence. As recurrence may occur in the first few months following resection, early follow-up with contrast-enhanced MRI is recommended with serial examinations indicated in those patients with residual radiologic abnormality but absence of symptoms or a clinically apparent mass.

**Vascular abnormalities**

Vascular abnormalities are classified into vascular tumours arising as a result of endothelial hyperplasia (haemangioma) and vascular malformations, congenital lesions with normal endothelial turnover. Vascular malformations are further classified according to whether they are high-flow or low-flow lesions and the type of vessel (artery, vein, capillary or lymphatic) that they involve.

![Figure 9](image1.png) **Figure 9** JA. Axial CT (bone algorithm) shows abnormal soft tissue expansion of the left nasal cavity and pterygopalatine fossa with anterior bowing of the posterior left maxillary wall (open arrow) and involvement of the left medial and lateral pterygoid plates (solid arrows).

![Figure 10](image2.png) **Figure 10** JA. Axial T1-weighted post-gadolinium MRI shows an enhancing soft tissue mass in the sphenoid and ethmoid sinuses with extension into the middle cranial fossa.
arise from. Complex vascular malformations containing more than one vessel type may occur. Complex vascular malformations containing more than one vessel type may occur.

Haemangiomas are benign endothelial tumours that undergo cellular proliferation and enlargement in the first year of life followed by gradual involution during early childhood. They are the commonest tumours of infancy and more than one-half occur in the head and neck. Thirty to forty percent of haemangiomas are present at birth; the remainder manifest in the first few months of life. Cervicofacial haemangiomas involving the chin, jaw line and pre-auricular areas (beard distribution) are associated with glottic and subglottic haemangiomas. These may impinge directly on the airway, prompting the need for surgery. The diagnosis of haemangioma is usually based on the clinical appearance: superficial lesions tend to be soft, red and raised, whereas deep lesions may be bluish in colour. Imaging may be used to confirm diagnosis and define extent, particularly if there is clinical concern regarding involvement of the glottis and subglottic region. Ultrasound of superficial lesions shows a mass lesion, which is highly vascular on colour and power Doppler imaging, notably during the proliferative phase in infancy, when these growing mass lesions cause clinical concern. On MRI, proliferating haemangiomas are discrete mass lesions, isointense to muscle on T1-weighted sequences and homogeneously hyperintense on T2-weighted images (Fig. 11). Internal flow voids may be present and homogeneous enhancement follows contrast injection. Given their natural history of involution, most haemangiomas do not require treatment. Lesions that are large or impinge on vital structures may be treated by systemic steroids, surgery or rarely with chemotherapy.

Lymphatic malformation is not a tumour but a benign vascular lesion that arises from embryological disturbance in lymphatic development. It is included here as it may present with a large cervical mass lesion, mimicking a tumour, and also to underline the distinction between this lesion, a type of vascular malformation, and haemangioma, a vascular tumour. The neck is the commonest location for lymphatic malformations with most lesions sited in the posterior cervical triangle. Diagnosis may be made on antenatal ultrasound or a mass lesion may be present at birth. If the lesion is large it may cause airway compromise. Most lymphatic malformations present before 2 years of age. Pathologically, lymphatic malformation is composed of multiple cystic spaces lined by endothelial cells with minimal intervening stroma. Lesions grow by insinuating around adjacent structures and may extend into the floor of the mouth, axilla or thorax. On US, a multilocular, predominantly cystic mass with internal septa is seen. Fluid-fluid levels with layered echogenic debris suggest internal haemorrhage. MRI is the best modality to demonstrate the extent of the lesion. Most lymphatic malformations are hyperintense on T2-weighted images and low-to-intermediate signal intensity on T1-weighted images. Unlike haemangioma, lymphatic malformations do not spontaneously regress and treatment is by surgical excision or sclerotherapy.

Teratomas

Teratomas are congenital neoplasms containing tissues derived from all three germ-cell layers. The sacrococcygeal region is the most common site for teratomas with less than 10% of teratomas in the newborn occurring in the cervical region. Cervical teratoma is often detected on antenatal ultrasound. Otherwise, it is clinically apparent at birth as a large cervical mass, which if of sufficient size, may cause airway compression. Malignant transformation in cervical teratoma is rare, and the major threat to life is airway compression. Ultrasound shows a heterogeneous mass with solid and cystic components. Calcification may be present. CT and MR appearances vary depending on the internal composition of the lesion. At CT, a unilocular hypoattenuating mass to a

Figure 11 Infantile haemangioma. (A) Axial T2-weighted MRI shows bilateral superficial, plaque-like facial haemangiomas (arrows) in a 10 month-old girl. (B) The same lesion shows uniform enhancement on an axial T1-weighted sequence following gadolinium administration.
heterogeneous mass containing areas of fat attenuation and calcification may be seen. On MRI the cystic components of a teratoma are often hypo- to isointense on T1-weighted images and hyperintense on T2-weighted images (Fig. 12). These lesions are often large and infiltrate widely.

Dermoid cysts also form part of the teratoma spectrum. These lesions are derived from two germ-cell layers; they are lined by squamous epithelium and contain variable skin appendages (sebaceous glands, hair follicles and sweat glands). Within the head and neck, the upper orbital margin is the commonest location of dermoid cysts, followed by the floor of the mouth. Lesions in the floor of the mouth are midline and most commonly occur above mylohyoid. Unlike thyroglossal ducts, there is no intimate relation with the hyoid bone and therefore dermoid cysts do not move with tongue protrusion. Initial assessment of any superficial structure should be with ultrasound but MRI is the best imaging modality to delineate the relationship of a cyst to the floor of mouth musculature. Dermoid cysts show variable signal intensity on T1-weighted sequences and are hyper-intense on T2-weighted sequences. On CT, a thin-walled, unilocular mass is seen in the submandibular or sublingual space. Coalescence of fat into small nodules within the fluid matrix gives rise to the pathognomonic ‘sack of marbles’ appearance.

Other benign tumours of the head and neck

A number of other non-malignant conditions may present with a mass lesion in the head or neck and thus must be differentiated from the malignant tumours already described. In addition, they may cause morbidity through local mass effect and potentially aggressive local growth. Langerhans cell histiocytosis (LCH) refers to a group of disorders characterized by the proliferation of histiocytes of the Langerhans type. Localized and disseminated forms of the disease occur with isolated bony involvement being the commonest form of localized disease in children. The cranial vault is the commonest location of osseous LCH but the orbit, maxilla, mandible and temporal bone are also frequently involved. Lytic bone lesions are depicted on radiographs but a combination of CT and MRI is often invaluable in determining the extent of bony destruction and associated soft tissue masses involving the head and neck.

Plexiform neurofibromas are benign peripheral nerve sheath tumours, virtually pathognomonic of neurofibromatosis type I. The commonest site in the extracranial head and neck is the first division of the trigeminal nerve in the region of the orbital apex. The infiltrative growth pattern of this tumour results in a diffuse mass of thickened nerves that may be superficial or deep in location, or a combination of both. MRI successfully depicts the extent of lesions, which are characteristically high signal intensity on T2-weighted imaging and low signal intensity on T1-weighted imaging. The deep lesions have a nodular appearance, whereas the superficial lesions have more diffuse, infiltrating morphology involving the subcutaneous tissues and the skin.

Fibrous dysplasia and juvenile ossifying fibromas are variant forms of benign fibro-osseous lesions characterized by replacement of bone with fibrous tissues and varying amounts of mineralization. Both conditions may arise in the craniofacial bones. Craniofacial involvement occurs in up to 25% of patients with mono-ostotic fibrous dysplasia and 50% of cases of polyostotic disease. Radiographs typically show expansile medullary lesions with ground-glass appearance and areas of sclerosis. CT may better delineate anatomical involvement. Juvenile ossifying fibromas are benign tumours commonly involving the paranasal sinuses and less frequently the orbit. Radiologically these are well-demarcated, expansile lesions of mixed lytic and sclerotic density.
Conclusion

A number of neoplastic lesions may occur in the extra-cranial head and neck in children. Imaging often has a key role in establishing diagnosis and in guiding management. We have described the more frequently occurring benign and malignant paediatric head and neck tumours and presented their imaging findings. Technical developments in US, CT, MR and PET will continue to expand the role of radiology in this field.

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