KEY WORDS

Midline neck lumps, Thyroglossal cyst, Sistrunk procedure, dermoid cyst, ectopic thyroid,

Introduction:

Cervical swellings are a common source of referral to paediatric surgical clinics. The majority will be caused by lymphadenopathy in the jugular nodes located laterally in the neck, but there are several conditions that can cause a swelling in the midline. Careful clinical assessment of a midline cervical swellings often enables an accurate diagnosis to be made, and informs subsequent management.

Causes of Midline Swellings

Congenital abnormalities are the cause of most midline masses, with inflammatory lesions being less common. Malignancy and other more serious causes are rare in this anatomical location in children.

Table 1

Thyroglossal Duct Cyst

Most midline cervical swellings in children are caused by thyroglossal duct cysts, accounting for about 70-75% in most reported series.^{1 2}

The cyst normally presents as an incidentally-noticed asymptomatic soft mass. A small number will present as an abscess or intermittently draining sinus because of spontaneous rupture due to infection.³

Embryology

Knowledge of the embryology of the thyroid gland helps explain the pathogenesis of these cysts. The thyroid gland is formed in the fourth week of gestation by the median thyroid anlage at the foramen caecum and the lateral thyroid anlage from the fourth and fifth branchial pouch complex.² Between the fourth and seventh weeks of gestation the thyroid anlage descends from the foramen caecum at the junction between the anterior and posterior parts of the

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tongue. It descends along the midline, to its final position low in the anterior neck as the embryo elongates. During this time, the hyoid bone develops from the second branchial arch and the thyroglossal tract becomes intimately related to it. The path of descent of the thyroid anlage is anterior to the developing hyoid but it loops inferiorly and posteriorly around the bone before continuing its descent anterior to the thyrohyoid membrane.⁴ The thyroid subsequently develops with lateral lobes connected by an isthmus and the tract disappears by the end of the 10th week of gestation.⁵ A remnant of the tract may persist as a pyramidal lobe in 50% of people.⁶ Post mortem studies have suggested that the thyroglossal tract fails to completely involute in 7% of the adult population.⁷ A cyst may form at any point along this residual tract.

Histopathology

Thyroglossal duct cysts can be lined by a variety of different epithelial linings. At the superior end of the tract cysts tend to be lined with pseudostratified ciliated epithelium while thyroidal acinar type epithelium can occur in cysts near the thyroid gland.⁶ Small foci of thyroid tissue can be found in the tract in 1-2% of cases.⁸ Secretions accumulate to form a cyst which is characteristically full of distinctive viscous material.

Figure 1

Anatomical location

The majority of cysts will present in the anterior midline near the hyoid bone.⁹ Most more or less overlie the hyoid. A small number will appear in the suprasternal area, with lingual and intrathyroid thyroglossal cysts also described, but are extremely rare.¹⁰

Dermoid Cyst

Dermoid cysts account for most of the remaining midline cervical swellings. They are slow growing inclusion cysts of epithelial elements in the line of fusion of the embryo and are lined with epithelium containing appendages such as hair follicles and sebaceous glands. The cysts are mainly filled with keratin and sebaceous material forming a characteristic cheesy liquid content. At surgery, the different content may help differentiate them from a thyroglossal cyst. Clinically, there may be a yellow hue seen through the skin. They are non-tender and virtually never get infected. They may be difficult to differentiate from thyroglossal cysts and mistakenly be thought to have an attachment to the hyoid bone..

Figure 2

Lymphadenopathy

Cervical lymphadenopathy is common in childhood, particularly in the lateral neck, but less so near the midline. The submental lymph nodes drain the lower lip, floor of mouth and apex of the tongue and can cause midline swellings just behind the mandible anteriorly. Thus, a tender swelling in the submental region demands careful inspection of the lip, floor of mouth, gums and tongue for evidence of infection.¹¹

Ectopic Thyroid

Abnormal thyroid descent can result in thyroid tissue located in an ectopic position. Ectopic thyroid is more common in females of Asian descent.¹² Patients with ectopic thyroid are commonly hypothyroid and an elevated TSH may stimulate enlargement of the tissue during childhood bringing it to clinical attention. By far the most common abnormality is lingual thyroid, which most series report as accounting for 90% of cases, but tissue may present in the cervical midline and other locations.^{13,14} In the majority of patients with lingual thyroid it represents the only functioning thyroid tissue and a normal thyroid gland is not palpable in the neck.¹⁵ The incidence of a midline neck swelling being the only thyroid functioning tissue has been suggested to be much lower at 50% of patients with ectopic thyroid and 0.17% of all those with midline cervical swellings.¹⁴ Whilst rare the unrecognised removal of the sole functioning thyroid tissue in a child can have devastating consequences.¹⁶

Lymphatic Malformations

Lymphatic malformations are low flow congenital cystic abnormalities of the lymphatic system. They commonly present as a ballotable subcutaneous mass. Rapid enlargement can be caused by haemorrhage or by infection. Most lymphatic malformations appear before the age of 2 and they tend to be more diffuse masses infiltrating tissue rather than being contained within a capsule.

Plunging Ranula

A ranula is the result of mucus being extravasated from a sublingual gland and forming a pseudocyst. The sublingual gland is the only salivary gland that secretes continuously and the high protein content aids cyst formation. The aetiology is usually obscure and they are

comparatively rare in childhood. A mucus filled cyst that affects the submandibular space is known as a plunging ranula. This can extend to the submental space and present as a midline swelling.¹⁷

Clinical Examination

Location

Thyroglossal duct cysts are attached to the hyoid bone, with the majority being immediately adjacent to it. Some thyroglossal cysts will sit slightly to one side of their midline attachment. Submental nodes are present in the submandibular area only. Dermoid cysts are commonly located in the submental and suprasternal areas but can occur at any level in the midline of the neck.

Inspection

Dermoid cysts frequently have a yellow hue that is more apparent in larger superficial lesions. Inflammation around a dermoid cysts is rare but can occur in the event of rupture (which is rare in this location in contrast to external angular dermoid cysts). An infected thyroglossal duct cyst can be readily identified by inflammation and skin erythema that spreads laterally. This is pathognomonic and allows these lesions to be distinguished from other lesions such as an infected node where inflammation tends to spread in a more circular fashion.¹¹ Progression of infection may result in an abscess which may spontaneously erupt to produce an intermittent discharge and subsequent sinus formation.

Movement

Movement of the tongue will cause minor movement of all midline neck lesions. The relationship of the thyroglossal cyst to the hyoid bone means its movement in swallowing is pronounced both with tongue protrusion and swallowing. Movement of the cyst is better palpated rather than observed: if the cyst is held between the fingers during swallowing the strong tug on it from movement of the hyoid bone can be felt. Unlike thyroglossal cysts, dermoid cysts do not move significantly with tongue protrusion as they do not have attachment to the deeper structures: this feature is useful in distinguishing the two conditions.

Transillumination

Transillumination can help distinguish a large uninfected thyroglossal cysts from other lesions. The viscous content of a thyroglossal cyst will usually glow brilliantly while the sebaceous material in a dermoid cysts and the solid nature of other lesions prevent transmission of light.

Figure 3

Palpation

Cysts should be palpated to gain more information about their nature, in particular their degree of tenderness, mobility, relationship to anatomical structures and the presence of fluctuance. Fluctuance is rare in the absence of infection but can signify a lymphangioma or plunging ranula.

Palpation of the thyroid is an essential part of examination. Palpation is best performed from behind, though some children prefer an anterior approach. The isthmus is the most easily palpated part of the thyroid in children in what can sometimes be a difficult examination to perform.

Examination of the Oropharynx

Examination of the oropharynx should not be forgotten during clinical examination. Isolated submental lymphadenopathy is commonly associated with an intraoral infection such as dental caries for which reason careful examination of the mouth is important. It is also rare for a plunging ranula to appear without an oral component which can be readily seen in the floor of the mouth.

Investigation

Imaging for thyroglossal cysts is controversial. Some authors maintain it is not required because the diagnosis can nearly always be made with certainty on clinical examination. Nevertheless, a survey of United Kingdom ENT surgeons found virtually every surgeon ordered an initial ultrasound study¹⁴ despite there being no firm evidence to justify this as a routine investigation. It may be considered where there is a delay or difficulty in obtaining a surgical opinion.

Thyroglossal cysts are highly variable in their sonographic appearance. They are variable in position, be unilocular or multilocular and have variable echogenicity.⁶ The majority of

thyroglossal cysts are unilocular and demonstrate posterior enhancement.¹⁸ This variability can be challenge to diagnostic accuracy and is one reason for questioning the utility of scans.

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Arguably, its main value may be to confirm the presence of a normal thyroid gland where there has been difficulty palpating it during clinical examination. An absence of normal thyroid gland with the presence of a solid component or calcification within the cyst raises the possibility of an ectopic thyroid gland or a carcinoma. In these extremely rare instances, further investigation with cross sectional imaging such as CT or MRI and functional studies such as radioisotope scanning should be performed, depending on the presumed pathology. The presence of a normal thyroid gland is sufficient to exclude a diagnosis of ectopic thyroid tissue being the sole functioning tissue.¹⁸

Some surgeons perform routine thyroid function tests during their assessment. Evidence would suggest this is not necessary except in those patients in whom an ectopic thyroid is suspected.

Management

A thyroglossal cyst should be excised to prevent infection and because of the small risk of malignancy. Cosmetic considerations are secondary.

Thyroglossal cysts can become infected by oropharyngeal organisms, in which situation they must be managed with antibiotics prior to surgery. The most common organisms include Haemophilus influenza, Staphylococcus aureus and Staphylococcus epidermidis.² If they form an abscess, aspiration or formal surgical drainage is needed as an initial procedure, and once the inflammation has resolved, definitive surgical excision is performed which includes excising the drainage tract. Inflammation at the time of surgery is a significant risk factor for recurrence and is best avoided.^{20, 21}

In adult populations, the incidence of malignancy in thyroglossal duct cysts is approximately 1%. There have been multiple case reports of children and adolescents with a thyroglossal cyst carcinoma with the majority being papillary carcinoma.^{5, 2, 22} A Sistrunk procedure may be adequate therapy though some authors propose completion thyroidectomy. The results of treatment results²³. As thyroglossal cysts are routinely excised in childhood, and usually shortly after diagnosis, the true incidence of thyroid tract carcinoma will always be unknown.

Complete surgical excision the cyst and tract is the treatment of choice. Walter Sistrunk in 1920 described the procedure which now carries his name: it involves resection of the cyst, the middle portion of the hyoid bone and the tract running towards the foramen caecum.²⁴ Although the technique has been refined over the years the fundamental principles remain sound and have been proven to reduce the risk of recurrence ten-fold.²⁵ Therapies such as sclerotherapy remain as yet unproven, and innovative approaches using cosmetic incisions or robotic surgery are only recently being published, with little data available on their value.²⁶

Management of ectopic thyroid diagnosed during a Sistrunk procedure is more controversial. If suspected, it should be biopsied for frozen section. The diagnosis can then be confirmed on histology. If the patient is known to have normal thyroid tissue then the tissue should be excised as part of the Sistrunk procedure. If there is concern that this might be the sole functioning thyroid tissue it is best left *in situ*, or divided and rotated on its vascular supply to lie behind the strap muscles. Some surgeons have advocated autotransplantation such as to the rectus abdominis or quadriceps muscle.¹³ Given that most require long-term thyroid hormone supplementation anyway, some surgeons recommend excision of the remnant to prevent malignant degeneration within the tissue.²

Dermoid cysts are excised to improve cosmesis and remove the risk of rupture and subsequent inflammation. Dermoid cysts excised from the hyoid area may be opened or sent for frozen section during surgery to confirm whether a Sistrunk procedure is required, but if a cyst appears to be attached to the hyoid most surgeons would proceed to a Sistrunk procedure anyway.

Surgical management of other lesions can also be required. Plunging ranula are treated by excision of the mass and its underlying sublingual gland. Lymphangiomas are usually managed with sclerotherapy with surgical excision reserved for selected cases. Submental lymphadenopathy is treated with the same principles as elsewhere in the body.

Outcomes

The general outcome of midline neck swellings with appropriate surgical management remain excellent.

The most troublesome complication of thyroglossal cysts treated with a Sistrunk procedure is recurrence which occurs in most series with a frequency of approximately 10%.^{27 28} Incomplete

excision, intraoperative rupture, surgical proficiency and experience, misdiagnosis, the presence of infection and variation in duct path have all been suggested as contributing factors.

⁶ Iuzzi:2013iv} ³ ²⁹ Unfortunately, recurrence can occur after an apparently technically proficient procedure. In general, repeat Sistrunk procedures involve wider resection of tissue and more aggressive resection of the hyoid bone.³⁰ ^{29,31} It has an increased risk of failure than a primary operation. Other complications can include wound infection and seroma formation.

^{3,8} Larygotracheal injury is much less common, but potentially devastating as it is usually caused by the erroneous resection of thyroid cartilage instead of hyoid bone. Early surgical exposure of the thyrohyoid membrane reduces the risk of this complication. Hypoglossal nerve injury has also been reported.¹³

Recurrence is rarer in dermoid cysts. Intraoperative rupture or incomplete excision of all epithelial elements is a risk factor for further recurrence of these lesions.

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