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Congenital neck masses in children and their embryologic and clinical features

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Abstract. *Congenital neck masses in children and their embryologic and clinical features.* Neck masses of congenital origin can be diagnostic and therapeutic challenges for internists, paediatricians and surgeons. Treatment modalities of congenital neck masses are different depending on their nature, symptoms and location. Differential diagnosis includes a variety of diseases that can cause cervical masses such as infectious and neoplastic neck tumours. Our objective is to review the embryologic and clinical features of some of the most common congenital neck masses such as the haemangioma, branchial cleft anomalies, thyroglossal duct cyst, ectopic thyroid, congenital midline cervical cleft, congenital cervical teratoma, lymphangioma, cervical thymic cyst, dermoid cyst and congenital muscular torticollis.

Introduction

Neck masses are common clinical findings in children. The differential diagnosis includes congenital, inflammatory and neoplastic lesions, which can present as palpable cystic or solid masses, infected masses, draining sinuses or fistulae. Congenital neck lesions most commonly found in the paediatric population include the thyroglossal duct cyst and the branchial cleft and arch anomalies. These can be present at birth or may manifest later in life. They can be diagnostic and therapeutic challenges for internists, pediatricians and surgeons. Differential diagnosis includes a variety of diseases that can cause cervical masses such as infectious and neoplastic tumours. Treatment modalities of congenital neck masses depend on their nature, symptoms and location and they generally require early excision to avoid complications of infection-inflammation, airway obstruction, nutritional compromise or the risk of malignant transformation. Our objective is to review the embryologic and clinical features of some of the most common congenital neck masses.

Haemangioma

Haemangiomas are the most common tumours of childhood, affecting as many as 10% of infants and of these, 60% occur in the head and neck. They manifest mostly within the first few weeks to months of life and may undergo a proliferative phase for 6 to 18 months followed by a slow period of involution over years. Haemangiomas can be infantile and congenital. The infantile haemangiomas are more common and appear as a stain at birth, which grow rapidly for several months, and may begin to involute with time. Congenital haemangiomas are fully formed at birth and can be either rapidly involuting or non-involuting.[1]

Haemangiomas are histologically characterized by endothelial cell proliferation. They can be localized, segmental and multifocal. Localized haemangiomas are papules or nodules that appear spatially contained, as if arising from a single focus point. Multiple haemangiomas are defined as six or more lesions. A segmental haemangioma consists of a plaque-like lesion that demonstrates a linear or geographic cutaneous patterning. Segmental lesions can be associated with complications such as ulceration.[1]

Haemangiomas will usually involute spontaneously by the age of 3 to 7 years.[2,3] While about 50% of haemangiomas regress spontaneously without sequelae, which justifies not treating them, the evolution in the other 50% of cases can be less favourable. Symptomatic problems such as ulceration, infection, bleeding, obstruction of orifices or psychosocial factors may be indications for treatment.

Compression treatment for facial or cervical haemangiomas is difficult and not used.[4] Medical management is preferred for lesions causing cosmetic or

functional deformity or in surgically challenging lesions. The corticosteroids control growth and regression of haemangiomas and are also used for palliation.[5-7] Corticosteroids, given orally or as local or systemic injections, are widely used for inducing early involution in large and rapidly growing haemangiomas.[8] They are used even in the 1st months of life if there is rapid proliferation or risk for the functional development of an organ. Intralesional corticosteroid injections are recommended for the management of infantile haemangiomas.[9] However, a corticosteroid injection may cause ulceration of the lesion.

Vincristine and interferon can be organ-sparing and are used when corticosteroid treatment fails. Interferon alfa-2a is recommended for the treatment of life-threatening or function-threatening corticosteroid-resistant haemangiomas.[10] An immunomodulator, imiquimod may be used as a topical cream.[11,12] Photodynamic therapy and lasers are used to treat superficial haemangiomas and ulcerations.[13] Becaplermin, a recombinant platelet-derived growth factor, 0.01% gel is a potential therapeutic option for ulcerated haemangiomas, including the refractory cases.[14] Various types of laser therapy are effectively used to treat both superficial and deep vascular tumours.[15] Good results may be obtained with surgical treatment when facial haemangiomas cannot be controlled by conservative therapy, interfere with an orifice or have a social or psychological impact on infants.[16] Stage, size and location of the lesion, presence of functional impairment and the amount of disfigurement are important in deciding on surgical treatment. In the proliferation stage, surgery is reserved for lesions that cause a functional problem. It may also be important to remove ulcerated areas that are not responding to medical or laser treatment. Once haemangiomas have stopped proliferating and begun with involution, surgery is important to help correct any disfigurement. When haemangiomas are protrusive, they slowly turn into fibrofatty tissue. Once this has occurred, liposuction of the fatty tissue will minimize scars.[17] A pyogenic granuloma (lobular capillary haemangioma) may begin proliferating as early as 2 weeks of age but the mean age of onset is 6.7 years. These lesions are small (6.5 mm). A tufted angioma, haemangiopericytoma or fibrosarcoma may be confused with haemangioma. Although the natural history of an haemangioma often differentiates it from other tumours and malformations, radiologic imaging is indicated if in doubt. Biopsy is indicated if malignancy is suspected. Immunostaining against erythrocyte-type glucose transporter (GLUT 1) can differentiate an haemangioma from other vascular malformations and tumours. An haemangioma appears sonographically as a soft tissue mass with decreased arterial resistance and increased venous flow. Unlike venous and lymphatic malformations, an haemangioma will have increased blood flow. During the proliferating phase, an haemangioma shows dilated feeding and draining vessels, appears solid and intermediately intense on T1-spin echo images and hyperintense on T2-weighted images on MRI. Fast flow and arteriovenous shunting are illustrated by flow voids. An involuted haemagioma appears as an avascular fatty mass on MRI.[6]

Branchial Cleft Anomalies

Incomplete obliteration of the branchial cleft during embryological development results in cysts, sinuses or fistulas of the neck.[18] The second branchial anomalies account for about 90% of all branchial anomalies. The other branchial anomalies are considerably rare.[19]

First branchial cleft anomalies

These account for 5 to 12% of all branchial cleft anomalies. Type I lesions are ectodermal in origin and lined by a squamous epithelium with or without accessory skin structures. They are a duplication anomaly of the membranous external auditory canal, located antero-inferior to the ear lobe in association with the parotid gland and superficial to the facial nerve. Type II anomalies are of ectodermal and mesodermal origin, and are duplications of the membranous and cartilaginous external auditory canal. They may present as a cyst, sinus or fistula and extend from the submandibular region, over the ramus of the mandible through the parotid gland with variable relation to the facial nerve, to end below the skin of the ear canal where there may be a sinus.[20] A branchial cyst has no internal or external openings, a branchial sinus has an opening on the surface ectoderm with or without cyst formation, and a branchial fistula has a communicating tract from the surface epithelium to the skin of the external auditory canal.[21]

The age of initial presentation ranges from birth to the second decade, with a mean age of

2.5 years. The common clinical presentation is swelling in the peri-auricular (24%), parotid (35%) or cervical regions (41%).[22] There may be external drainage from infection or desquamating epithelial debris or a skin pit in the case of a sinus or fistula.[23] There may be a history of recurrent otorrhoea, otitis externa or hearing loss.[24] Almost 35% to 48% undergo recurrent surgical intervention before the correct diagnosis is made.[25]

Differential diagnosis includes any other cystic mass of the parotid gland like obstructive or traumatic retention cysts and benign lympho-epithelial parotid cysts, benign parotid tumours like cystic Warthin's tumour, malignant parotid lesions with central liquefactive necrosis and lymphangiomas.[23] CT is useful in the differential diagnosis, confirmation of the diagnosis and definition of both location and extent of the lesion.[26] Pre-auricular pits anterior to the tragus of the ear are usually not related to a branchial cleft but represent aberrant development of the auditary tubercles. These pits are lined with squamous epithelium, may contain hair and other skin appendages and may form a cyst. They extend from the skin surface down through the subcutaneous tissue. The tract is deep and tortuous, leading to the cartilage of the external auditory canal. They are frequently familial, bilateral and asymptomatic and excision is not routinely needed, but when drainage or infection occurs, they often persist or recur and require excision. Draining or infected sinuses may consist of a chain of several small cysts linked together and to the skin by the sinus tract. These squamous cysts contain keratinaceous material. An uninfected sinus is excised electively and the tract is readily identified and traced down to the cartilage. There is often considerable adherence to the cartilage and a small portion of the cartilage must be removed in order to evade a recurrence.[24]

The risk of secondary infection of branchial lesions is about 25% and definitive excision should be performed before the occurrence of multiple episodes of infection, which cause scarring and difficulties in dissection.[27] Infection may produce severe adhesions between the lesion and facial nerve, resulting in permanent weakness of the facial nerve postoperatively. The position of the facial nerve in relation to the sinus or fistulous tract is variable.[22] The only effective treatment is complete excision of the lesion in the absence of infection or inflammation.[23] The overall recurrence rate after operation is about 3% to 5% with a slight increase for sinuses and fistulas compared to cysts.[22]

Second branchial cleft anomalies

Six paired branchial arches, mesodermal in origin, appear between the fourth and sixth week of intrauterine development. The external and internal invaginations through the arches give rise to the development of internal endodermal pouches and external ectodermal clefts.[18,19] Second branchial fistulas occur when caudal growth of the second pharyngeal arch over the third and fourth arches is interrupted, leaving the remnants of the second, third and fourth clefts in contact with the surface by a narrow canal.[28,29]

Second branchial anomalies are the most common of all branchial anomalies and are usually located on the lateral side of the neck directly anterior to the superior one third of the sternocleidomastoid muscle (Figure 1).[28,30,31] Two to 13% of the second branchial anomalies are bilateral.[29,32,33] The course of the second branchial fistula begins at the external opening of the lesion, coursing cephalad along the lateral carotid sheath, passes between the internal and external carotid arteries and travels lateral to the glossopharyngeal, hypoglossal and superior laryngeal nerves. After moving deep to the stylohyoid muscle, it pierces the upper border of the middle pharyngeal constrictor muscle to enter the tonsillar fossa.[28,30,34] Sinuses and fistulas occur at younger ages while cysts in adults. The majority of second branchial anomalies are detected by the age of 5 years. Most of them manifest as an intermittent mucous or purulent discharge during the newborn period.[28] Almost 80% of the patients have cysts and the rest has fistulas.[33] Sometimes a skin tag or a cartilaginous remnant is present around the external orifice.

Although some claim that intervention is not needed in asymptomatic patients, most of these cases need surgery for cosmetic reasons or recurrent infections.[29,30] Early diagnosis and treatment decreases recurrence by preventing infection, which causes adhesions in the tissue planes. Operative complications include glossopharyngeal, hypoglossal or facial nerve injury (3.5%), fistula formation, internal jugular vein trauma, infection, seroma, haematoma and scar formation (3.3%).[35] The overall recurrence rate after surgery is 3 to 4.9% but increases to 25% if surgery is performed after the first infection.[28,35,36]

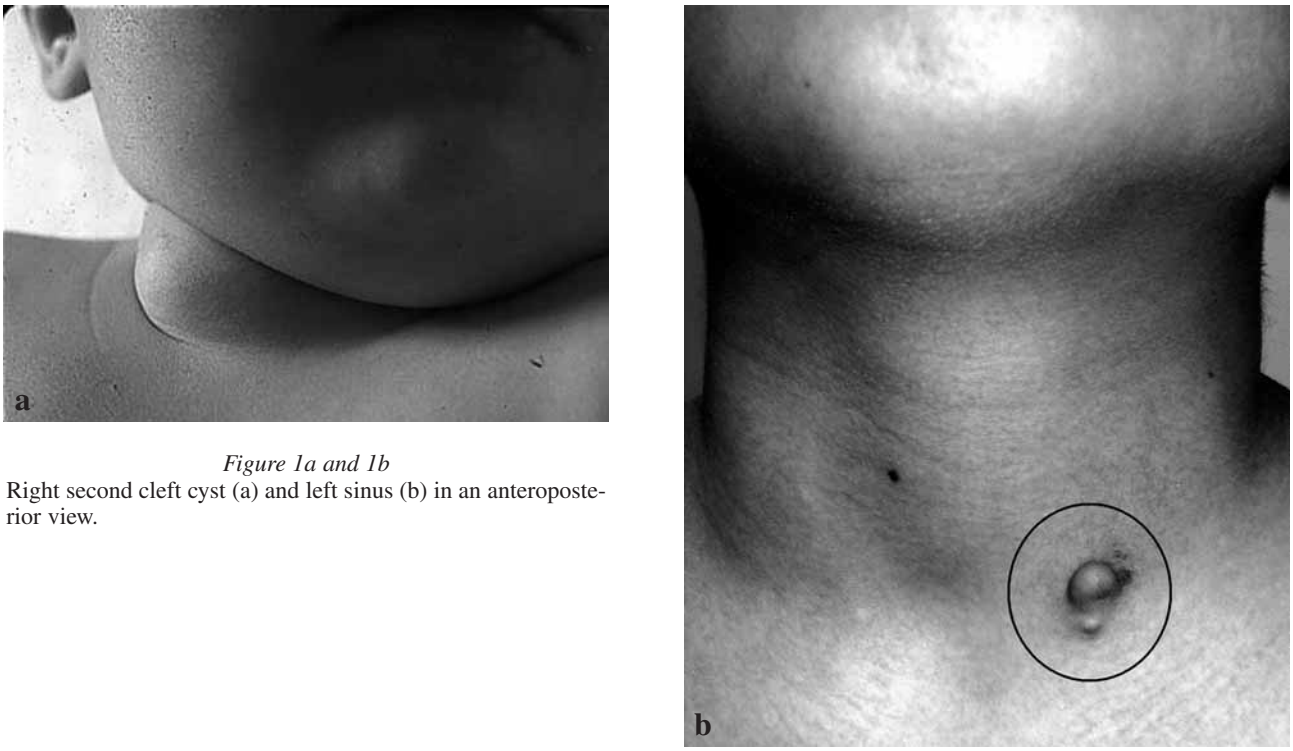

Figure 1a and 1b
Right second cleft cyst (a) and left sinus (b) in an anteroposterior view.

Third and fourth branchial cleft anomalies

Failure of the third and fourth branchial pouches to obliterate *in utero* results in cysts or sinus tracts that lie in close proximity to or inside the thyroid gland. When present, the sinus tract originates in the pyriform fossa, also known as the pyriform sinus.[32,33,36] These remnants occur more frequently on the left.[37] Both the third and fourth pouches connect to the pharynx by the pharyngobranchial duct, which involutes during the seventh week of development.

Third pouch remnants cross the superior laryngeal nerve posterior to the common carotid artery. Fourth pouch remnants emerge caudal to the thyroid cartilage and cricothyroid muscle and pass between the superior and recurrent laryngeal nerves.[38] The sinus tract is lined by stratified squamous epithelium, which may be replaced in areas with respiratory epithelium.[39] The third and fourth branchial remnants present at any age. In neonates, these anomalies can be dangerous because rapid enlargement may cause tracheal compression and respiratory distress as the infant swallows fluids. Non-communicating or non-infected communicating cysts may present as cold thyroid nodules, be inside the thyroid gland, and confused with thyroglossal cysts.[32,39,40] The lesions may manifest with recurrent painful thyroid or neck masses, thyroiditis and cellulitis.[41] When the child presents with an acute infection, aggressive antibiotic therapy is followed by elective resection.

Thyroglossal duct cyst

Thyroglossal duct cysts (TGC's) were considered the most common congenital midline neck swelling in children, but the incidence of thyroglossal and branchial anomalies is approximately equal in recent large pediatric series.[42]

The thyroid gland begins its embryological development at the level of the foramen caecum of the tongue. Subsequent descent of the thyroglossal diverticulum will allow the thyroid gland to reach its normal position in the inferior neck area. The thyroglossal duct, connecting the thyroid gland with the foramen caecum, normally involutes beginning inferiorly. If this involution is incomplete, a remnant of the duct may persist in the upper neck area, and contribute to formation of the TGC. While TGC's are usually recognized by age 5 years, at least 50% of the lesions are diagnosed in the second decade of life.[42,43] The majority of TGC's lie close to the hyoid bone although they may be at any site along the path of descent of the thyroid gland.[44]

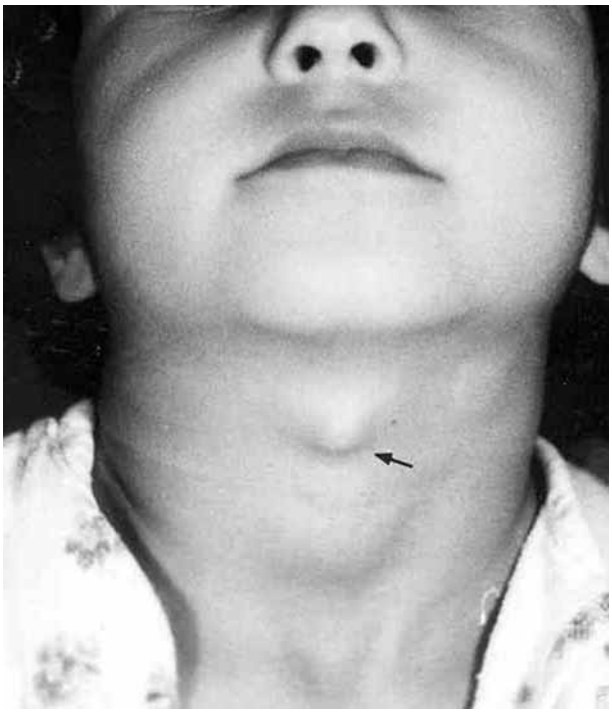

Figure 2
Thyroglossal duct cyst in a child

TGC's present most often with a painless swelling, a draining sinus or a tender mass in the midline of the neck (Figure 2).[42,45] Unusual locations of TGC's have been reported.[40,46,47] Movement of the cyst with swallowing is a reliable diagnostic sign whilst movement of the mass with protrusion of the tongue is unreliable, especially in children.[42] The TGC may be confused with a dermoid cyst, lymphadenopathy or cystic hygroma when they are located near the hyoid bone.[44,45,48-51] Co-existence of a dermoid cyst and a TGC is reported.[40,44] Ultrasonography (USG) may be valuable and sufficient in the assessment of the thyroid gland. USG will reveal the normal thyroid localization and pattern.[52]

Excision of the hyoid body and cyst was first proposed by Schlange in 1893 (recurrence rate 20%) and Sistrunk popularized the operation in 1920, advising removal of a tissue core from the hyoid bone, thus reducing the recurrence rate to 3%.[44,45] The recurrence rate increases with dermal involvement, young patient age, rupture of the cyst during the operation, lobulation of the cyst, inflammation and/or infection, incomplete excision of the suprahyoidal tract, and presence of a fistula.[52,53]

Ectopic thyroid

Ectopic thyroid (ET) is the presence of functioning thyroid tissue in a site other than its normal pretracheal position. ET tissue can exist anywhere in the midline between the foramen caecum and anterior mediastinum and is localized to the floor of the mouth in 90% of cases.[54-56] It is rare in the sublingual, pre-laryngeal, pretracheal, intratracheal and mediastinal regions.[5] ET may cause respiratory distress, dysphagia and haemorrhage.[54,55] The patients are usually euthyroid and excessive swelling of the ET may cause hyperthyroidism.[54,55]

Treatment is mainly medical with thyroid hormone replacement in cases of hypothyroidism. Close follow-up is advised in euthyroid and asymptomatic patients. Surgical excision is necessary when the ET leads to a neck mass, dysphagia, haemorrhage or airway obstruction.[54,55]

Congenital midline cervical cleft

The congenital midline cervical cleft (CMCC) is a rare anomaly also described as a medial cleft, median fissure of the neck, congenital midline cervical cord, midline cervical webbing and pterygium colli medianum. There are less than 100 cases reported to date.

Midline clefts may be the consequence of an anterior fusion defect of the two first pairs of branchial arches during the third and fourth weeks of intrauterine life;[57-60] They may result from pressure of the primitive heart on the branchial arches, which in turn could generate adhesions leading to a defect of fusion.[57,58,61]

CMCC is clinically typical and diagnosed at birth. The length and width can differ from one child to another, but clinically the cleft presents as linear vertical areas of thin and erythematous skin. Nipple-like projections are usually noted over the clefts, as well as sinus or fistula openings at their lower ends. Fibrous bands are often found and can restrict cervical extension (Figure 3). Tractions

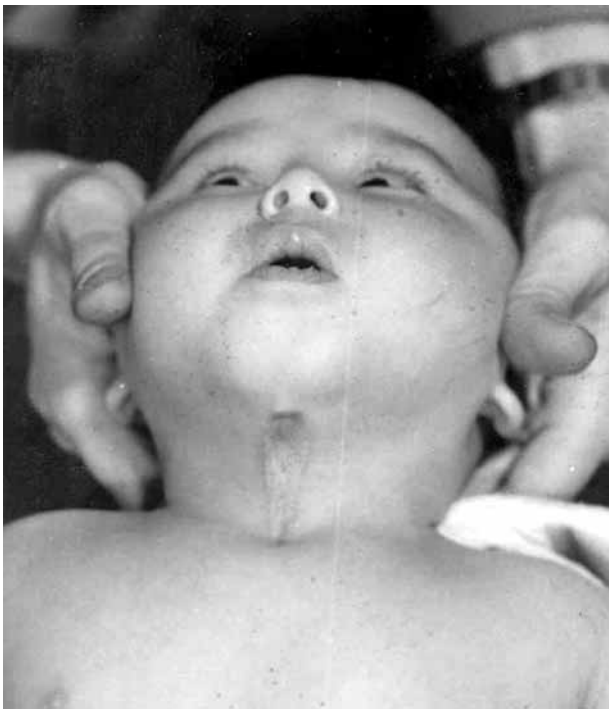

Figure 3
Midline cervical cleft in an infant

performed by these fibrous bands on the lower part of the mandibular symphysis can generate a bony spur seen on plain roentgenograms.[57,58,62,63] There may be associated pathologies like a cleft of the lower lip, tongue, mandible and sternum or hypoplasia of the hyoid bone. Complete aplasia of the hyoid bone and thyroid cartilage is possible.[64]

The lesion is covered by epithelium without pilosebaceous apparatus.[58] Secretions produced by accessory salivary glands may drain from a fistulous tract.[58,65] Treatment of CMCC is surgical, and early intervention is recommended for aesthetic reasons, and in order to avoid subsequent limitation of extension of the neck and impairment of mandibular growth. In the surgery, all pathologic tissue, including the fibrous cord must be removed to prevent formation of subcutaneous inclusion cysts and recurrence. Skin closure by Z-plasty prevents a retractile straight-line scar.[57,58]

Congenital cervical teratomas

Teratomas are a heterogeneous group of rare neoplasms originating from pluripotent cells and foreign to the anatomic site in which they arise. Only 3 to 5% of teratomas occur in the neck and frequently present as large and deforming neoplasms. Of these, over 90% are found in neonates.[66-68] Less than 300 cases of cervical teratomas have been reported.[69]

Prenatal sonography identifies the presence of the tumour.[66-68] The most common finding is a pharyngeal mass, consisting of semi-solid and semi-cystic elements with well-defined borders and calcifications in about 50% of cases.[67] A plain radiograph of the neck after birth demonstrates a soft tissue mass with calcifications. They usually present as a large solitary mass but may also be multifocal (Figure 4).[67] Malignancy occurs in 5 to 10% of teratomas and causes an elevation in serum alpha-fetoprotein. Large lesions may cause hyperextension of the neck of the fetus, oesophageal obstruction, swallowing disturbance and polyhydramnios. Polyhydramnios is seen in 18 to 40% of teratomas.[67,68] Cervical teratomas are usually large and may give rise to difficult labour and problems with the airway after birth. Perinatal morbidity is high and usually related to the size of the tumour. Untreated cervical teratomas are associated with an 80 to 100% mortality[70] and is usually related to airway problems.

The treatment is complete excision. A multidisciplinary team must be present in the delivery room for optimal treatment. When endotracheal intubation is impossible, a tracheostomy is performed. An *ex utero* intrapartum treatment (EXIT) procedure provides time until an adequate airway can be established whilst the baby is still attached to the umbilical cord, thus maintaining uteroplacental gas exchange.[59,60,61] If the airway is secured and the tumour removed in the immediate postnatal period, the survival rate is as high as 85%.[71] In some of the patients, there may be pulmonary hypoplasia, a hypoplastic larynx and tracheomalacia due to tumour compression.[66]

Lymphangioma

The lymphangioma (LA) is a congenital lymphatic malformation, accounting for 6% of all benign

Figure 4
Giant cervical teratoma in a child

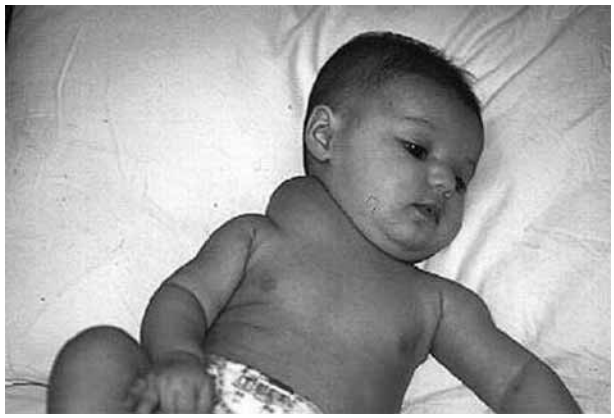

Figure 5
Lymphangioma in the right cervical region

lesions of infancy and childhood.[72,73] Both sexes are equally affected, 60% are present at birth and 90% are detected before the end of the second year.[73] The LA results from the failure of embryonic lymph channels to establish a communication with the venous system.[74] The likely aetiologies include failure of the lymphatic system to connect with or separate from the venous system, abnormal budding of lymphatic structures from the cardinal vein, abnormal sequestration of lymphatic tissue in early embryogenesis and acquired processes including trauma, infection, chronic inflammation or lymphatic obstruction.[75] LA's are divided into three categories based on histologic appearance; capillary lymphangiomas, cavernous lymphangiomas and cystic hygromas.

LA's are soft, cystic and usually translucent. Almost 50% of all LA's occur in the head and neck,[73] are mostly in the posterior triangle of the neck and may suddenly increase in size with infection or haemorrhage (Figure 5).[76] They do not become malignant nor do they have a familial tendency, but often infiltrate adjacent tissue planes.[77] LM's may cause polyhydramnios or may be associated with chromosomal abnormalities like Turner syndrome (45, X; 31%), trisomy 21 (15%), trisomy 18 (7%), trisomy 13 (2.5%), and other chromosomal abnormalities (3.5%).[78]

LA's mostly present with a mass or diffuse swelling, which initially grows slowly along with the child, and after some time, may slowly regress.[79] Respiratory tract infection and trauma may cause enlargement of the mass.[73,79] The history and physical examination suggest the diagnosis of a LA. The children are normal except for the presence of the mass.[66] Macroglossia due to a LA of the tongue can cause dyspnoea and swallowing difficulty.[73,79]

Careful monitoring for at least 2 to 6 months is recommended as the mass may cause dyspnoea and dysphagia.[73,80] Asymptomatic cases are followed up in the hope of spontaneous regression, which is rare. Complete resection is achieved in 77%, and recurrence can occur in 20 to 50% of cases.[73] Injection with agents such as bleomycin, tetracyline, steroids, and 50% dextrose, results in a 70% recurrence and even worsening of the symptoms in some cases. These agents may be used in recurrent cases.[73,74] Treatment of LA's with OK-432 seems promising. In 96% of patients with macrocystic LA, complete regression is achieved with OK-432, whereas patients with microcystic disease generally do not respond and should therefore not be injected with this agent.[81]

Surgical intervention is indicated for life-threatening symptoms. In an emergency situation, needle drainage may assist in establishing an airway. A mass without other symptoms is observed for 18 to 24 months to allow for spontaneous resolution. If the lesion is decreasing in size, additional time is allowed before a surgical intervention. Macrocystic lesions in all locations may respond to sclerosing agents such as the currentlyfavoured OK-432. However, complete surgical excision of macrocystic lesions is equally effective.[82]

Cervical thymic cyst

The thymus gland is the central organ of the lymphoid system

during infancy. It develops from the third and fourth pharyngeal pouches.[83,84] The dorsal wing of the 3rd pouch elongates into a tubular structure known as the thymopharyngeal duct.[84,85] In the 7th to 10th week of intra-uterine life, primordial thymus tissue in the neck descends to the mediastinum whilst its duct becomes atrophic and disappears. During this process, some pieces of thymic tissue may remain in the neck, which may lead to a cervical thymoma and thymic tissue cysts.[82,83]

A cervical thymic cyst is a rare congenital anomaly presenting as a neck mass in children and there are less than 100 cases reported to date.[86] However, cervical thymic masses are relatively more common and mostly asymptomatic. Cervical thymic tissue remnants are seen in almost 30% of the autopsies of asymptomatic children. The majority of thymic masses occur in infancy and childhood as the thymus is at its largest.[82,87]

The thymus weighs 30-40 grams in puberty and 15 grams in adulthood, when it becomes fibrofatty tissue. Almost 75% of the patients are less than 20 years of age (mean age 12 years) and 10% of the masses occur in infancy.[82,88] Typically, thymic cysts are located in the anterior cervical triangle,[82,88] are soft, uni- or multilocular masses, and contain a straw-coloured to dark brown fluid and sometimes semisolid or necrotic debris and cholesterol crystals. The cyst wall is lined with squamous, cuboidal or columnar epithelium. Thymic elements are found in a close contact with the cyst wall. Hassall's corpuscles are present.[83] Embryonic remnants of branchial clefts, thymopharyngeal ducts, sequestration of the thymus, degenerating Hassall's corpuscles, mesenchymal elements arrested in development and neoplastic changes in the solid cervical thymic tissue have been postulated as the origin of thymic cysts.[89-91]

Congenital thymic cysts are usually asymptomatic. In less than 10% of the patients, they may cause dysphagia, dyspnoea, odynophagia, pain or hoarseness and rarely respiratory distress in neonates. Infections and haemorrhages may cause sudden enlargement of the cysts.[82,83]

Complete surgical excision of the cysts does not cause a problem in adults. However, the thymus is essential in childhood for its immune function. Therefore, a conservative approach is recommended.[82,88] Recurrence or malignant transformation is not reported in children after surgical excision of the cyst. However, these are possible in less than 2% of adults.[82,85]

Dermoid cyst

The dermoid cyst and its epidermoid variant that arise in the head and neck area are believed to result from congenitally included ectoderm trapped during bilateral embryologic fusion of the 1st and 2nd branchial arches.[92-94] The congenital dermoid cyst is classified into three categories. First, the epidermoid cyst is an epithelium-lined cavity with a capsule that contains no skin appendages like hair, sweat and hair follicles and sebaceous glands. In the second group, the cyst differs only in that it contains skin appendages. In the third group, the cyst contains skin appendages in addition to bone and muscle tissue.

The most common localizations are the orbits and the floor of the mouth, and they rarely occur in the submandibular region.[91] Dermoid cysts are thought to originate in the midline in the oral cavity, above the mylohyoid muscle, and then shift position laterally during expansion and extension into the neck. They usually manifest as neck masses in the 2nd and 3rd decades of life. The well-circumscribed, painless and uni-locular cyst grows slowly, varying in size from several millimetres to as much as 12 cm.[91] Dermoid cysts in the lateral side of the neck are larger than the midline masses. The mass occasionally causes dysphagia and airway compression. Rarely, infections may cause fistula formation between a laterally located cyst and the oral cavity, and these may be confused with a submandibular gland disorder. Surgical excision is both diagnostic and therapeutic.[95,96]

Congenital muscular torticollis

Torticollis in Latin means twisted neck and was first defined by Tubby in 1912 as "a congenital or acquired deformity, characterized by lateral inclination of the head to the shoulder with torsion of the neck and deviation of the face". Congenital muscular torticollis (CMT) is the third most common congenital problem of the musculoskeletal system in neonates and infants with a reported incidence of 0.3 to 1.9%.[97,98] The typical lesion is a hard mass within the substance of a tight sternocleidomastoid muscle. It is often recognized when a child is one to four weeks old. The size of the lesion ranges from 1 to 3 cm in its largest transverse diameter. It is firm and

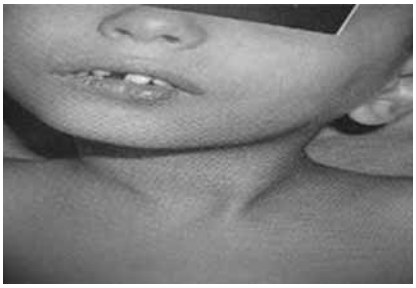

Figure 6
Torticollis in a 9-year-old boy

smooth, mobile beneath the skin and tender on palpation.[97,98] Cervical hemivertebrae, cervical adenitis, acute fasciitis and strabismus lead to torticollis in childhood. However, neonatal torticollis is the most common type of torticollis and results from fibrosis of the sternocleidomastoid muscle, which produces a fibrous mass and shortening of the muscle.[99]

CMT may result from fetal malposition, birth trauma (breech delivery), hereditary problems (10% have a family history of torticollis), infections, congenital and acquired lesions of the central nervous system (cervical spine instability, neural compression, neurologic injury) and cervical hemivertebrae.[97,100-102] Bilateral CMT is rare.[90]

The diagnosis of CMT is made clinically by pulling the ear toward the clavicle on the ipsilateral side whilst the face looks upwards towards the contralateral side (Figure 6). In torticollis that occurs early in childhood, the deformity may be preceded in the early weeks of life by the formation of a mass in the mid-portion of the sternocleidomastoid muscle. This occurs in 20 to 43% of the patients. Differential diagnosis includes inflammation, trauma, psychiatric problems, neurological torticollis, ocular problems, Grisel's syndrome (non-traumatic atlanto-axial rotatory subluxation) and Sandifer syndrome (abnormal body movements and contortions of the neck, associated with gastro-oesophageal reflux). CMT coexists with other congenital musculoskeletal anomalies in 6 to 20% of the patients. Congenital hip dysplasia is the most common associated anomaly.[96,102] There is facial asymmetry in 14 to 70% of cases.[96,103] CMT and plagiocephaly (flattening or obliquity of one or more sides the skull) may occur together in 1 per 300 live births.[104]

Although 8% of all CMT resolves spontaneously, 60 to 70% of the persistent cases have cranial or facial asymmetry. The initial therapy consists of physical therapy. If contraction of the sternocleidomastoid muscle persists beyond 1 year of age or a craniofacial anomaly develops, surgical treatment is required.[103,105] Early diagnosis of CMT is important because almost 95% of children treated by physical therapy in infancy recover. Otherwise, surgery is needed with a resulting increased recurrence rate. Additionally, surgery is also indicated in the presence of head tilt after 6 months of physical therapy, deficit in passive rotation and lateral bending of the neck >15º, and in cases of a mass.[97] The most common surgical treatment for CMT is muscle release or sternocleidomastoid muscle tenotomy. Postoperatively, a neck collar is applied and exercise prescribed for 4 to 8 weeks.[96-98,101]

Conclusion

The clinical examination of congenital cervical lesions allows diagnosis in most cases without requiring further studies. In suspected or undetermined cases, USG, CT or MRI help achieve a differential diagnosis. During acute inflammation (infection and abscess), surgery is more difficult and increases the recurrence and nerve injury rate.

Figures 1-5 are with the courtesy of ACB.

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