Review Article

Congenital Craniofacial and Cervical Cysts, Sinuses and Fistulas- A Review Article

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Abstract

Congenital cysts, sinuses and fistulas of the head and neck are anomalies that may be encountered in pediatric life. They represent abnormal development of the brachial apparatus or inclusion of the ectoderm at any site of the lines of fusion of the face. In the head, preauricular, salivary and median nasal fistulas as well as dermoid cysts and Van der Woude syndrome are examples. In the neck, median thyroglossal cyst and fistula, and lateral branchial cyst and fistula are well-known congenital anomalies. Proper diagnosis of these malformations is essential to decide the line of treatment. Standard and modified surgical procedures are adopted for excision and repair of these anomalies with the possibility of incidence of complications, recurrence and malignancy.

Keywords: Congenital; Cysts; Sinuses; Fistulas; Craniofacial; Cervical; Thyroglossal; Branchial

Abbreviations: CT: Computed Tomography; MRI: Magnetic Resonance Imaging; MULFs: Median Upper Lip Fistulas; TGDCs: Thyroglossal Duct Cysts; VWS: Vander Woude Syndrome

Introduction

A fistula is an abnormal tract communicating the cavities of two internal organs or the cavity of an organ with the body surface whereas a sinus is one-end blind tract that opens on the surface. Congenital cysts, sinuses and fistulas of the head and neck are uncommonly detected in children. They include congenital craniofacial and cervical anomalies; the latter may be medio-cervical or latero-cervical in position. Surgical excision of these lesions is the only option of treatment.

Congenital Craniofacial Cysts and Fistulas

Congenital periauricular cysts, sinuses and fistulas occur commonly in children are developmental anomalies of the first branchial cleft and first pharyngeal arch [1,2]. Children with recurrent postauricular abscesses show associated congenital anomalies like postauricular sinuses, post auricular dermoid cysts, first branchial cysts, aural fistulas and aural atresia [3]. Congenital salivary fistulas of the main part of the parotid gland are rarely reported; surgical excision of the fistula and parotid gland is an effective line of treatment for this anomaly [4]. Odontogenic cutaneous fistula results from abnormal tooth canalization caused by chronic periapical infection; it may be wrongly diagnosed as a congenital fistula [5]. Abnormal protrusion of a part of the brain and its meninges through a defect in the skull bones is termed meningo-encephalocele. A case of an anterior meningo-encephalocele presenting as a small swelling in the right lower eyelid was reported; if it was mistreated as a lachrymal swelling it might result in meningitis and cerebrospinal fluid leakage [6]. The nasal dermoid cyst and median nasal sinus are rare congenital anomalies that are attributed to early embryonic inclusion of the ectodermal germ layer during closure of the anterior neuropore of the neural tube [7]. Congenital nasofrontal fistulas (also termed nasofrontal dermal sinuses) are extremely rare malformations that may be complicated with serious meningitis [8]. Patients with congenital nasal fistulas and dermoid cysts are operated upon through vertical midline or horizontal glabellar; and nasofrontal osteotomy [9]. A craniofacial dermoid cyst at the lateral wall of the orbit was reported to be associated with a cutaneous frontotemporal fistula that extended to the dura of the temporal lobe of the brain through the temporal and sphenoidal bones; lateral orbitotomy was surgically performed in this case [10].
Congenital sinuses and fistulas of the lip are uncommon and median upper lip fistulas (MULFs) are extremely rare. A case of congenital MULF, at the philtrum of a girl infant, was surgically removed. The fistulous tract was histologically lined with squamous epithelium that contained sebaceous and mucous glands as well as hair follicles. [11,12] Van der Woude syndrome (VWS) is a single gene autosomal dominant congenital craniofacial abnormality that is featured by labial cysts, accessory salivary glands, and lower lip pits, fistulas and paramedian sinuses; it is commonly associated with cleft lip or palate or both [13,14]. Excision of lower lip sinuses and accessory glands, reconstruction of the lip, nose, and associated cleft lip and palate are the components of VWS surgery [15]. Surgical excision of congenital lower lip sinuses has an increased rate of mucocoeles formation and recurrence [16].

**Congenital Cervical Cysts and Fistulas**

Neck masses are frequently met with in children and they may be congenital or acquired in origin. Congenital neck swellings include branchial cysts, thyroglossal cysts, hemangiomas and cystic hygromas. They may be complicated into sinuses and fistulas [17,18]. Congenital cervical cysts, sinuses and fistulas are not common. Thyroglossal duct cysts (TGDCs) are the commonest malformations followed, in frequency, by branchial cleft anomalies and dermoid cysts [19]. Congenital cysts and fistulas of the neck are divided, according to their locations, into midline or medio-cervical and latero-cervical anomalies [20,21]. In man during the 3rd to the 4th week of development, a pharyngeal (branchial) apparatus is formed in the wall of the pharyngeal gut. It consists of six mesodermal arches, four ectodermal clefts and five endodermal pouches. The derivatives of these branchial elements include essential structures in the head and neck while their remnants may develop into congenital neck anomalies like branchial cyst (and fistula) and thyroglossal cyst (and fistula) [22]. Latero-cervical congenital cysts and fistulas are mentioned to be due to abnormal development of branchial clefts particularly the 2nd one; they are usually found at the anterior border of the lower third of the sternodeidomastoid muscle. The TGDCs (and fistulas) are the most common medio-cervical congenital anomalies; they are usually found in the median thyrohyoid region. Surgical resection of the cysts and fistulas should be complete to avoid recurrence and risk of malignancy [23]. Congenital cervical salivary fistulas result from ectopic non-branchial salivary tissue that has a sinus draining serous or mucous secretion to the anterior part of the base of the neck [24]. In congenital dermoid fistula of the anterior chest region, a skin orifice could be seen at the anterior border of the sternocleidomastoid muscle with subcutaneous extension of the fistulous tract till the sternoclavicular joint [25].

**Median Neck Cysts and Fistulas**

TGDCs are the commonest congenital swellings of the neck [26]. They represent 2% of cervical masses and 70% of congenital neck swellings [27,28]. TGDCs result from patency of a part of the embryonic thyroglossal duct that extends from the foramen cecum (on the dorsum of the tongue) till the thyroid primordium (pouches). They are painless and commonly diagnosed in children below the age of 5 years. A TGDC may ulcerate spontaneously leading to the formation of a thyroglossal fistula that rarely communicates with foramen cecum [28]. Thyroglossal fistulas may be suprahypoid, hyoid or infrahyoid in position [29]. It is agreed that congenital mento-ternal fistulas are embryologically and pathologically different from thyroglossal fistulas. Mento-ternal fistulas are most probably caused by midline cervical inclusion of ectodermal or endodermal tissue during fusion of the branchial elements [30]. Excision of the TGDC and fistula is performed through Sistrunk procedure which includes removal of the body of the hyoid bone. The lining epithelium of the excised cysts varies according to the sites of the cysts; it is cuboidal, columnar, pseudo stratified or stratified squamous in type [31]. In an adult female patient, during a Sistrunk procedure for a TGDC with recurrent purulent discharge, two fistulous tracts terminating at the hyoid bone were discovered; it was mentioned that similar cases were missing in the literature [32]. Moreover, a branched thyroglossal duct terminating in two separate cysts with a chronic fistula was diagnosed in a young woman with a history of an incomplete surgical excision of a TGDC [33].

Clinically, most of the thyroglossal fistulas are considered as the sequel of interventional procedures: incisions, punctures, enucleations or radiations of TGDCs. Radical Sistrunk procedure is an effective option for removal of a TGDC [34-37]. Standard or modified Sistrunk operation includes removal of the TGDC and/or fistula together with the middle part of the hyoid bone; the duct is microscopically or macroscopically dissected till the foramen cecum to avoid recurrence [38-43]. The recurrence rate of TGDCs removed by Sistrunk procedure is reported to be 4% [44]. In a group of patients with TGDCs and fistulas, the size of the cyst varied from 1 to 4 cm; Sistrunk procedure had the advantages of low rates of complications (9.08%) and recurrence (1.82%), avoidance of antibiotics usage and short hospital stay [45]. Preoperative neck ultrasonography is recommended as a diagnostic tool of a TGDC whereas postoperative histopathology of the excised cyst and fistula is mandatory to exclude rare occurrence of malignancy [46,47]. An unusual case of a TGDC at the base of the tongue and fistulizing to the anterior part of the tongue was described and successfully excised via a combined trans-hyoid and intra-oral surgical approach [48].

Piriform sinus fistulas are congenital anomalies of the third and fourth branchial arches. They are usually unilateral and associated with thyroiditis and recurrent abscesses in the lower part of the front of the neck. A case of rare bilateral congenital piriform sinus fistulas is mentioned in the literature [49].

**Lateral Neck Cysts and Fistulas**

Branchial anomalies of the 1st to 4th branchial clefts and arches represent 20% of all congenital head and neck swellings in children [50-52]. Branchial cleft anomalies can present as a neck swelling, [319x463]Clinically, most of the thyroglossal fistulas are considered as the sequel of interventional procedures: incisions, punctures, enucleations or radiations of TGDCs. Radical Sistrunk procedure is an effective option for removal of a TGDC [34-37]. Standard or modified Sistrunk operation includes removal of the TGDC and/or fistula together with the middle part of the hyoid bone; the duct is microscopically or macroscopically dissected till the foramen cecum to avoid recurrence [38-43]. The recurrence rate of TGDCs removed by Sistrunk procedure is reported to be 4% [44]. In a group of patients with TGDCs and fistulas, the size of the cyst varied from 1 to 4 cm; Sistrunk procedure had the advantages of low rates of complications (9.08%) and recurrence (1.82%), avoidance of antibiotics usage and short hospital stay [45]. Preoperative neck ultrasonography is recommended as a diagnostic tool of a TGDC whereas postoperative histopathology of the excised cyst and fistula is mandatory to exclude rare occurrence of malignancy [46,47]. An unusual case of a TGDC at the base of the tongue and fistulizing to the anterior part of the tongue was described and successfully excised via a combined trans-hyoid and intra-oral surgical approach [48].

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Branchial anomalies of the 1st to 4th branchial clefts and arches represent 20% of all congenital head and neck swellings in children [50-52]. Branchial cleft anomalies can present as a neck swelling,
sinus, or fistula. Second branchial cleft anomalies account for about 95% of all anomalies of the branchial apparatus. Repeated preoperative infections of these anomalies lead to a higher rate of postoperative recurrence [53]. Branchial cysts are supposed to result from incomplete developmental obliteration of the branchial cleft or trapping of cell nests in the branchial apparatus that can later form branchial cysts [54]. Second branchial cleft cysts and sinuses are the most common type of branchial cleft anomalies. Failure of obliteration of the cervical sinus of His leads to the formation of a branchial cleft and failure of fusion of the 2nd branchial arch with the 5th arch results in a branchial sinus (fistula) [55]. A rare coexistence of TGDC and branchial cleft with fistula in the same child is reported in the literature [56]. Unilateral first and bilateral second branchial cleft fistulas were concomitantly observed together with a preauricular sinus and moderate bilateral hearing loss in branchio-otic and branchio-oto-renal syndromes [57]. A case of an ectopic tooth in a branchial cleft anomaly was described in a young girl who had Townes-Brocks syndrome [58]. Another case of second branchial cleft sinus remained asymptomatic till the sixth decade of life when it was surgically excised together with ipsilateral carotid endarterectomy [59]. Furthermore, a congenital branchial fistula may be complete with an internal orifice at the tonsillar fossa; complete surgical excision of the fistulous tract could be performed in one setting [60].

**Conclusion**

Congenital cysts, sinuses and fistulas of the head and neck must be properly diagnosed and managed to diminish the rate of complications and recurrence, and to avoid the risk of malignancy.

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