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Fourth branchial pouch or cleft anomalies

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Fourth branchial cleft and pouch anomalies are rare but clinically important entities that require a high index of suspicion. A missed diagnosis can prolong patient morbidity resulting in incorrect treatment and probable recurrence. These anomalies are secondary to errors in embryogenesis from incomplete involution of the fourth branchial cleft or pouch, resulting in the formation of a cyst, sinus, or fistula. They commonly present as a recurrent lateral neck swelling in the pediatric population. Surgery is the mainstay of treatment.

Introduction

Branchial anomalies are common causes of pediatric neck masses, accounting for approximately 17% of all pediatric cervical masses.[1] Fourth branchial anomalies are extremely rare; however, accounting for only 1%-2% of branchial malformations.[2-4]

Despite being first described more than 40 years ago,[5] there have only been just more than 500 cases of fourth branchial anomalies reported in the literature.[6] They commonly present as an infected neck swelling in childhood that can easily be mistaken for suppurative lymphadenitis. Otolaryngologists must be vigilant, as errors in diagnosis can lead to increased patient morbidity and unproductive interventions.

As is the case with all congenital anomalies, knowledge of embryology is key to understanding the pathogenesis, presentation, and treatment of the resultant condition. By the fourth week of gestation the 6 arches, clefts, and pouches making up the branchial apparatus have formed. The fourth branchial arch is responsible for the formation of many important cervical structures, including the superior laryngeal nerve and the pharyngeal constrictors.[2,7] The fourth branchial pouch gives rise to the superior parathyroid glands bilaterally as well as the ultimobranchial body, which itself is in part responsible for the development of parafollicular C-cells in the thyroid gland.[2] Bearing this in mind, fourth branchial anomalies characteristically present in close association with the thyroid gland, with a tract ascending to the pyriform sinus. Consequently, consideration must be taken to address the full extent of the tract, often including the thyroid gland.

Anatomy

Fourth branchial anomalies are diagnosed by their anatomical course. They are classically described to begin in the apex of the pyriform sinus, pierce the larynx near the cricothyroid ligament, and then pass between the superior and recurrent laryngeal nerves.[8] Right and left anomalies differ in their course beyond this point. A left sided anomaly descends in the tracheoesophageal groove, loops around the aorta in a posterior to anterior direction, ascends posterior to the common carotid before passing over the hypoglossal nerve (cranial nerve [CN] XII), and exiting the neck anterior to the sternocleidomastoid muscle (SCM). A right-sided anomaly also descends in the tracheoesophageal groove to the level of the subclavian artery where it loops around in a

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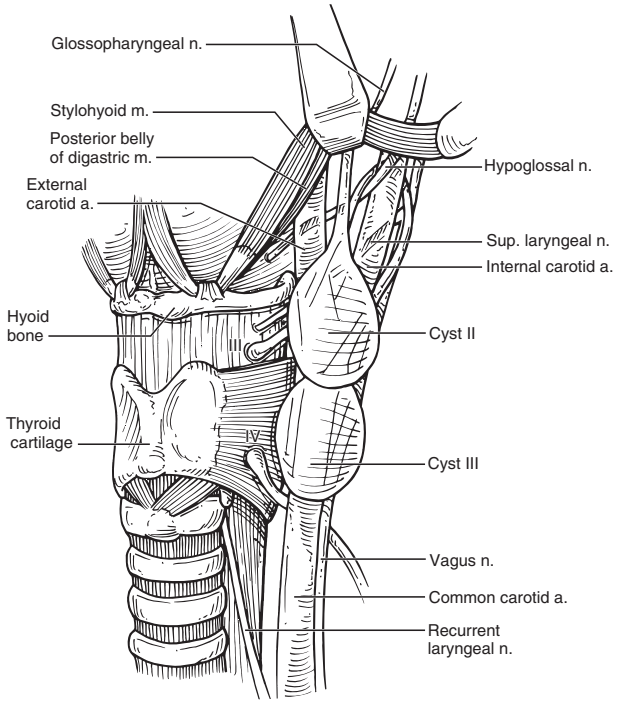

Figure 1 Courses of brancial anomalies.

posterior to anterior direction before ascending with the common carotid artery and following the same course as on the left side.[2,9] The relationship of the tract to the piriform apex, cricothyroid joint, and superior laryngeal nerve are the main points of differentiation between a third and fourth branchial anomaly; a third branchial anomaly passes cephalad to the superior laryngeal nerve whereas a fourth passes caudally (Figure 1).[9-11]

Histologically, ectopic parathyroid, thyroid, or thymic tissue, or all of these can be found in a fourth branchial anomaly.[2] Although they mainly contain stratified squamous epithelium or respiratory ciliated epithelium, the presence of ectopic tissue(s) can aid in the diagnosis of a branchial anomaly, but cannot help distinguish a fourth from a third anomaly.[6]

Presentation and evaluation

Fourth branchial anomalies present most commonly as a neck mass in childhood, with more than 90% of cases occurring on the left.[2,9,12,13] The mean age of symptom onset is 9 years old and the mean age of diagnosis is 14 years old.[6] Presentation varies based on the age of the patient. Neonates with fourth branchial anomalies are likely to experience respiratory distress, whereas older children tend to present with either a lateral neck abscess or suppurative thyroiditis.[6] A range of symptoms has been described, including upper respiratory tract infection, cervical pain, thyroid tenderness, cellulitis, hoarseness, and odynophagia.[9,12,14] Mediastinal abscess has also been reported in advanced cases.[15,16]

Physical examination must include a thorough head and neck examination, including flexible laryngoscopy. Typical neck findings including lateral neck swelling, tenderness, overlying erythema, warmth, induration, and possible fluctuance. Respiratory symptoms are possible and flexible laryngoscopy may reveal unilateral supraglottic swelling, erythema, or purulent debris, or all of these if there is a connection to the piriform sinus. The combination of unilateral neck swelling and supraglottic findings should raise a high index of suspicion for a third or fourth branchial anomaly. Laboratory tests are of limited use in this setting. Radiological investigations of use include ultrasound, contrast swallow studies, magnetic resonance imaging, and computed tomography. Timing of investigative studies is also important. Magnetic resonance imaging and computed tomography are most useful during acute infection. Although the actual sinus or fistula tract typically cannot be easily seen on imaging, the combination of an infected neck cyst with adjacent thyroid inflammation and inflammation of the tissues along the expected course of the tract is pathognomonic. Direct laryngoscopy and contrast swallow studies have the best positive predictive values of all investigative tests, with values of 90% and 88%, respectively.[6] Contrast swallow studies can be helpful in illustrating fourth branchial anomalies after the acute infection has subsided, as contrast can pool in the pyriform fossa sinus and proximal tract. Direct laryngoscopy can be performed during an acute infection, and if a sinus tract is identified it confirms the diagnosis.[17]

Treatment options

Definitive treatment for fourth branchial anomalies is surgical. Open and endoscopic approaches have both been described. Endoscopic intervention is used either as an initial intervention or as an adjunct to open excision.[18,19] Endoscopic interventions as an initial approach are advocated as there is significantly less associated morbidity.[19]

Resection should be avoided, if possible, in an acutely infected anomaly.[2,9] In the setting of infection, aggressive treatment with antibiotics should be the initial management strategy with a planned definitive resection performed several weeks later.[20] Antibiotic choice should be dictated by culture and sensitivity testing, if available. Typical organisms found are oral flora susceptible to penicillin or related β-lactamase–resistant antibiotics.[6] If sepsis, respiratory compromise, or progression of the infection is present, needle aspiration or incision and drainage can be considered, but it may complicate future definitive resection.[2,6,10]

In an open transcervical surgical resection, both proximal and distal ends of the lesion are addressed in an effort to avoid recurrence. This is technically challenging, as the anatomical course of fourth branchial anomalies can be quite circuitous. Distal lesion excision usually involves removal of the cystic mass as well as the ipsilateral thyroid gland, as the fistula tends to be either immediately medial, lateral, or through the gland itself.[21,22] Recurrence is greatly increased when thyroid tissue is not removed.[9,23,24] Exposing the inferior horn of the thyroid cartilage can aid