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Second branchial cleft anomalies

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Second branchial anomalies represent the most common type of branchial anomalies. Their anatomical course is well understood based on knowledge of the embryologic branchial apparatus. These anomalies tend to present in children, though slow-growing cysts may first be noted well into adulthood. The most common presenting sign is a slowly growing lateral neck mass. Surgical excision remains the mainstay of therapy.

Introduction

Second branchial anomalies include cysts, sinuses, and fistulae. They represent the most common branchial anomalies. Understanding the embryology and resulting anatomy of these anomalous structures can allow one to predict their course and implement safe, effective surgical interventions with low recurrence rates.

Second branchial cleft anomalies commonly occur along the anterior border of the sternocleidomastoid muscle. Sinus and fistulous tracts of second, third, and fourth branchial anomalies all exit near one another, owing to their common embryologic origin in the cervical sinus of His. Second branchial anomaly tracts course between the external and internal (derived from the third arch) carotid arteries, superior to the glossopharyngeal and hypoglossal nerves, and finally end in the tonsillar fossa, usually along the posterior pillar. Some authors have further subclassified second branchial anomalies based on minor anatomical variations of the tract,[1] but these classifications have not been broadly applied.

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Histology

Squamous epithelium, pseudostratified columnar (respiratory) epithelium, and combinations thereof have been reported to line second branchial lesions. Lymphoid, sebaceous, and salivary tissues have also been associated with second branchial anomalies. Cysts are more commonly lined by squamous epithelium, whereas sinuses and fistulae tend to be lined by ciliated, columnar epithelium.[1] Cholesterol crystals may be seen in cystic fluid.

Presentation

Generally, second branchial cleft cysts present as nontender, stable-to-slowly-enlarging lateral neck masses that may acutely increase in size during an upper respiratory tract infection, leading occasionally to respiratory compromise, torticollis, and dysphagia. Fistulae and sinuses are usually diagnosed earlier than cysts, owing to the more obvious presence of a skin opening with chronic drainage along the anterior border of the sternocleidomastoid. In a review of 52 patients, the 3 most common presenting symptoms of branchial anomalies were drainage from a cervical sinus opening, neck mass, and repeated infection.[2]

Second branchial anomalies are the most common and have been reported to account for 95% of all branchial anomalies.[1] However, a more recent review of 74 branchial

anomalies found that second branchial anomalies accounted for 69% of cases.[3] Reviews of the proportion of cysts, sinuses, and fistulas have also varied widely. One review of mostly adults noted cysts accounting for approximately 80% of anomalies,[4] whereas another review of children found sinuses, fistulae, and cysts accounting for 45%, 27%, and 27%, respectively.[3] Another review of pediatric patients also found sinuses were more common.[2] Perhaps this discrepancy relates to the fact that cysts often are found much later in life than sinuses and fistulae, as it can take many years for cysts to enlarge and become noticeable.

In a retrospective review of 74 branchial anomalies, the average age of presentation for was 4.1 years for a cyst, 3.6 years for a sinus, and 2.6 years for a fistula.[3] Approximately one-third of second branchial anomalies had been previously infected before diagnosis. Of that third, 71% of patients had more than 1 infection. Of the anomalies with a skin opening, 85% presented with drainage and 35% presented with a history of infection. For those with both a lateral neck skin opening and drainage, 59% were ultimately diagnosed as a sinus and 37% as a fistula. Sinuses and fistulas were more likely to present with infection (67%) compared with cysts (21%).

Previous reviews, including a relatively large Spanish series, have noted that second branchial anomalies are seen more commonly in women, with a female to male ratio of 2:1 or even higher.[4] However, this female predominance has not been duplicated in later studies.[3,5] There also appears to be no predilection for right or left laterality.

Evaluation

The diagnosis of a second branchial anomaly can be straightforward. A careful history and physical examination, as well as a high index of suspicion and clinical awareness, are all important factors in establishing a diagnosis. Any unexplained neck mass, recurrent neck infection, or abscess should be suggestive of a possible branchial anomaly.

Although not exhaustive, the differential diagnosis of second branchial anomalies includes reactive lymphadenopathy, lymphadenitis, abscess, lipoma, lymphatic or vascular malformation, lymphoma, ectopic thyroid and thyroglossal duct cysts, tuberculosis adenitis, and cervical thymic cysts.[3] In adults, it is always important to exclude a cystic cervical lymph node metastasis by fine needle aspiration to avoid surgical misadventure.

Clinical examination alone has been reported as 50%-60% accurate when used to diagnose a second branchial anomaly.[6,7] Consequently, several authors have examined the role of preoperative imaging in establishing an accurate diagnosis. Computed tomography scans with and without fistulography, magnetic resonance imaging, and ultrasound have all been employed in the workup of second branchial anomalies. Computed tomography is the best-studied imaging modality, and preoperative diagnosis of branchial anomalies has a reported accuracy rate as high as 93%.[3] Ultrasound can be a useful tool in preoperative evaluation due to its low cost, absence of ionizing radiation, and ability to be performed in an office setting. A large review from China found that the diagnostic accuracy of ultrasound was 66.2%.[8] Use of magnetic resonance imaging in the evaluation of second branchial anomalies has been described in case reports as accurate, though not necessarily practical due to its expense and possible need for sedation in young children.[9]

Routine laboratory testing is not required in the workup of most patients with second branchial anomalies. Most often, second branchial cleft anomalies are not associated with additional anomalies. However, one syndrome is worth noting. Branchiootorenal syndrome, an autosomal dominant disorder with variable expressivity, is characterized by hearing loss (conductive, sensorineural, or mixed), ear defects (outer, middle, and inner have all been described), branchial anomalies, and renal abnormalities.

Treatment options

Surgical excision has been shown to be safe and effective for addressing second branchial anomalies and remains the mainstay of treatment. However, several studies have trialed sclerotherapy as an alternative to surgery in branchial cleft cysts. Kim et al[10] injected OK-432, a lyophilized mixture of low-virulence Group A *Streptococcus pyogenes* and penicillin G potassium, directly into branchial cleft cysts in a prospective study of 23 patients. Fourteen of 18 patients with unilocular cysts experienced complete regression. Of the 5 patients with multilocular cysts, 2 showed a partial response and 3 showed no response to OK-432 sclerotherapy. Side effects of OK-432 therapy are noted to be minor and include mild fever, bloody drainage, and temporary injection site pain. Based on the best available evidence, sclerotherapy is not the standard of care and should only be offered to patients with unilocular branchial cleft cysts.

Indications for surgery

Once discovered, surgical excision of second branchial anomalies is almost uniformly recommended given the propensity of recurrent infection and cosmetic deformity associated with these lesions. Generally, surgical excision is delayed until a child is at least 3 months old. Surgical excision should not be performed during an acute infection. Rather, one should allow adequate time for the infection and associated inflammation to subside with appropriate antimicrobial therapy before proceeding with surgical excision, typically 2-4 weeks after resolution of the acute infection.

Details of surgical technique

Before incision, the patient should be placed supine on the operating table with the neck extended by placement of a small shoulder roll. If it is known that a fistulous tract is