International Surgery Journal
Shenoy NS et al. Int Surg J. 2017 Oct;4(10):3234-3237
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: http://dx.doi.org/10.18203/2349-2902.isj20174108

Original Research Article

Anomalies of branchial cleft: our experience and review of literature

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Received: 06 July 2017
Revised: 13 August 2017
Accepted: 23 August 2017

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ABSTRACT

Background: Anomalies of branchial arches are uncommon anomalies of embryonic development and may present as cysts, sinus tracts, fistulae or cartilaginous remnants. We describe our experience with 30 children with branchial cleft anomalies.
Methods: Case records of all patients were retrospectively reviewed and analysed with respect to age, sex, clinical presentation, duration of symptoms, investigations, management and follow up. All patients underwent complete excision of the tract/cyst.
Results: There were 15 males and 15 females (ratio of 1:1). The average age of presentation was 5 years. Majority of the patients presented with discharging sinuses (n=26). Twenty-six patients had branchial sinuses, three patients had branchial cysts and one had pyriform fistula. The anomalies were lateralized to left side in 17 patients (56.66%), right side in 11 patients (36.66%) and bilateral in 2 patients (0.066%). Complete excision was done in all patients. Four patients presented with abscesses and required incision and drainage. Definitive surgery was done after 6 weeks. There were no complications.
Conclusions: Abnormal development of branchial apparatus lead to formation of different anomalies which remain asymptomatic and present later in life as cysts, sinuses or fistulae in neck. Diagnosis is mostly clinical and complete excision provides definitive cure.

Keywords: Branchial apparatus, Cyst, Fistula, Sinus

INTRODUCTION

Anomalies of branchial arches are uncommon anomalies of embryonic development.[1] They represent the embryological precursors of face, neck and pharynx and are the second most common congenital lesions of head and neck in children.[2,3] They account for approximately 17% of all paediatric cervical masses.[1,2]

The branchial arches develop in the fourth week of gestation and are the embryological precursors of ear and muscles, blood vessels, bones, cartilage and mucosal lining of the face, neck and pharynx.[2] In total, six pairs of branchial arches are formed on either side of the pharyngeal foregut in cranio-caudal succession.[2] The fifth pharyngeal arch is usually rudimentary, or disappears and the sixth arch is often represented as part of the fourth arch due to its small size.[2-4]

Thus, depending on the anatomic location, branchial anomalies have been classified into first, second, third and fourth arch anomalies.[1] Second branchial arch anomalies are the most common accounting for approximately 95% of cases. First branchial arch anomalies account for 1-4% cases; third and fourth branchial arch anomalies are extremely rare.[2-5] They may

present as cysts, sinus tracts, fistulae or cartilaginous remnants.[2] Cysts are the entrapped remnants of branchial clefts; sinuses are remnants of clefts or pouches; and fistulae can result from persistence of both pouch and cleft.[1] The typical clinical and radiological presentation depends on the arch which is involved.[2]

We describe our experience with 30 children with branchial cleft anomalies treated at a tertiary institution.

METHODS

This is a retrospective observational study of 30 patients with branchial cleft anomalies who were managed at a tertiary centre from January 2012 to June 2017. Case records of all patients were reviewed for age, sex, clinical presentation, duration of symptoms, investigations, management and follow up. Family history and previous history of infection and/or surgery were noted. Patients with sinus and fistulas underwent sino- / fistulogram, if required. Cystic lesions were investigated with ultrasound (USG) and CT scan. All patients underwent complete excision of the tract/cyst.

Patients with acute infection were started on antibiotics; incision and drainage was done in patients with abscesses. Surgical excision of the tract was done after six-week interval in these patients.

RESULTS

Case records of total 30 patients with branchial cleft anomalies were retrospectively reviewed and analysed. There were 15 males and 15 females (ratio of 1:1). The average age of presentation was 5 years with range being 6 months to 11 years. However, the history dated back to infancy in most of the patients.

Majority of the patients presented with discharging sinuses (n=26); 13 patients had an associated swelling. Solitary cysts were present in three patients only. One patient had pyriform sinus fistula.

Four patients presented with abscesses and required incision and drainage. All four of these patients were infants which implies that the narrow sinus opening may have contributed to their blockage leading to infection and early presentation.

Twenty-six patients (86.66%) had branchial sinuses. Four sinuses were seen at preauricular region and three at clavicular region (Figure 1); rest 19 sinuses were at anterior border of lower two-thirds of sternocleidomastoid muscle. Three patients had branchial cysts (0.1%). One patient had pyriform sinus fistula. There were no patients with branchial fistula. The anomalies were lateralized to left side in 17 patients (56.66%), right side in 11 patients (36.66%) and bilateral in 2 patients (0.066%).

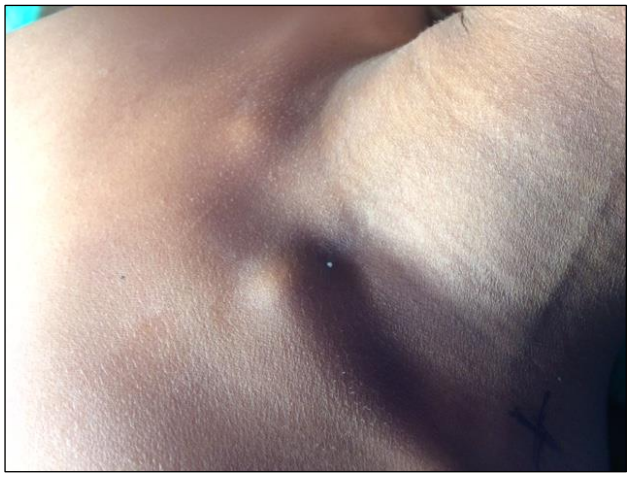

Figure 1: Clinical image of a patient with left clavicular sinus.

Ultrasound (USG) was done for patients with swelling. Two patients were referred with a sino-/fistulogram and CT/MRI. One had a recurrent branchial cyst and the other had a pyriform fistula.

The patient with pyriform fistula presented with incision and drainage done elsewhere for neck swelling followed by intermittent discharge of orally ingested fluids. MRI was suggestive of pyriform fistula. The tract was successfully excised.

Surgical excision of the tract was done in all patients. Histopathology confirmed branchial pathology. There were no complications or recurrences.

DISCUSSION

Branchial anomalies were first described in the early nineteenth century; however, their origin and classification are a subject of controversy even today. Several theories have been proposed for the development of branchial anomalies.[1] These include branchial apparatus theory, cervical sinus theory, thymopharyngeal theory and inclusion theory. Amongst these, the widely accepted theory is that branchial anomalies develop as a result of incomplete involution of the branchial apparatus.[1,6,7]

Most branchial arch anomalies arise from the second branchial cleft (92.45%).[1,8] Anomalies of first arch remnants (4.72%), third (1.87%) and fourth arch anomalies (0.94%) are quite rare [d]. Bajaj et al, have reported a higher incidence of second branchial anomalies (78%) in their series of 80 patients.[1,9] Although Choi and Zalzal reported a higher incidence of first branchial arch anomalies (25%) in their series; they still had the maximum incidence of second branchial arch anomalies (40%).[1,6]

Although congenital, branchial anomalies usually present later in life.[1] The age of onset of symptoms depend on the type of lesion.[1] Branchial cysts usually present in older children/young adults, whereas fistulas typically present in infants/young children. Most of these anomalies are confined to the left side; the same was seen in our study.[1] These are seen more common in females; however, in this study, we had equal number of girls and boys.[1]

Branchial anomalies can present as cysts, fistulas, sinuses or cartilaginous remnants.[2] A sinus is a blind ending tract which may connect with either the skin (branchial cleft sinus) or with the pharynx (branchial pouch sinus).[2] A fistula is a communication between two epithelialized surfaces, i.e., between a persistent pouch and cleft.[2] If there is no communication with the inner mucosa or outer skin, then the trapped branchial arch remnant will form a cyst.[2,7] Choi and Zalzal have reported a maximum incidence of sinuses, followed by fistula.[1,6] In this study, majority of patients presented as discharging sinuses (26/30).

Second branchial cleft anomalies are the most common and usually present as sinuses.[2,11] Most of these present within the submandibular space but can occur anywhere along the course of the second branchial arch tract, i.e., from the skin overlying the supraclavicular fossa, between the internal and external carotid arteries, to pharynx at the level of the tonsillar fossa.[2,4] Thus a sinus or fistula in the lower anterior or lateral region of the neck is most likely to represent a second branchial cleft anomaly.[2] Majority of patients in this study had second branchial cleft anomaly.

Sinuses or fistulae usually present in infancy/ childhood with drainage of secretions or purulent material from an opening at the anterior border of the sternocleidomastoid within the lower third of the neck.[2] Cysts are most often diagnosed as a painless, compressible lateral neck mass in a child/young adult that may become tender and increase in size if they get infected.[2,11] Histologically, they contain a turbid yellow fluid containing cholesterol crystals and are lined by stratified squamous epithelium.[2]

First branchial cleft anomalies can occur anywhere along the course of the first branchial arch tract. This extends from a cutaneous opening in the submandibular triangle, superolateral to the hyoid bone, ascending to the region of the parotid salivary gland to terminate at the cartilaginous/bony junction of the external auditory canal. The tract may pass above or below the facial nerve.[2,12]

Third and fourth branchial cleft anomalies appear similar to second branchial cleft anomalies externally with a cutaneous opening in the supraclavicular area; however, internally, they enter the pharynx through the pyriform sinus below the hyoid bone.[2] There were three patients with clavicular sinuses in this study.

Third and fourth branchial cleft anomalies are distinguished anatomically by their relationship to the superior laryngeal nerve with third pharyngeal cleft anomalies above and fourth pharyngeal cleft anomalies below.[2] The internal opening helps to determine their origin and a third branchial cleft sinus arises from the rostral end of the pyriform fossa.[2,4] Third branchial cleft fistulae/sinus tracts typically present earlier than third branchial cleft cysts. Most third branchial cleft cysts present in the posterior cervical space, posterior to the sternocleidomastoid muscle as a painless, fluctuant mass that may enlarge and become tender if infected. An infected third branchial cleft cyst should be considered if a patient presents with an abscess in the posterior triangle of the neck.[2]

A fourth branchial cleft fistula/sinus tract arises from the pyriform sinus apex and descends inferiorly to the mediastinum in the path of the tracheo-oesophageal groove.[2,13,14] They are commonly left-sided.[1,2] Only one patient had left pyriform sinus fistula in this study.

The diagnosis of branchial anomalies is usually clinical.[1] No radiological investigations are usually required. However atypical lesions can be misdiagnosed.[1] An initial accurate diagnosis is crucial because recurrence rates after surgical excision of branchial anomalies are 14% and 22% with previous infection and surgery respectively, whereas recurrence rate for primary lesion is 3%.[1,15] Physical examination and proper history remain the most important elements in a correct diagnosis. The radiological investigations help to add valuable information in the evaluation of a neck mass.[1]

On ultrasound (USG), second branchial cleft cysts are typically well-circumscribed, thin-walled and anechoic with evidence of compressibility and posterior acoustic enhancement.[2,16] They may contain internal echoes compatible with internal debris. On CT imaging, they are well- circumscribed, low-density cystic masses with a thin wall.[2,7] On MRI, T1- weighted imaging may show low to high signal depending on the proteinaceous content of the cyst, but are typically hyperintense on T2-weighted imaging.[2,13] In both CT and MRI imaging, mural thickening and enhancement varies with inflammatory change and typically occurs in the setting of infection.[2] A tissue 'beak' between the internal and external carotid arteries is pathognomonic of Bailey type III cysts.[2]

Surgery is the definitive mode of management.[1] The natural history of these lesions reveal that there is lack of spontaneous regression, a high rate of recurrent infection, possibility of other diagnoses and rare malignant degeneration.[1] Acute inflammation should be treated medically unless incision and drainage or aspiration of an abscess is required.[1] Three to four weeks should pass after an acute infection before a definitive surgical exploration is undertaken.[1] In this study, four patients

presented with abscesses requiring incision and drainage followed by definitive surgery after 6 weeks.

Surgery involves complete surgical excision of the cyst and sinus/fistula encompassing the external opening with dissection of the tract.[2,17] Complications following surgery include recurrence, wound infection and facial nerve paralysis (specifically in patients requiring superficial parotidectomy for first arch anomalies).[1] There were no complications in this study.

CONCLUSION

Branchial apparatus plays an important role in the development of head and neck structures. Abnormal development of these structures lead to formation of different anomalies which remain asymptomatic and present later in life as cysts, sinuses or fistulae in the neck. Surgery is the treatment of choice for these lesions due to the fact that these lesions do not regress spontaneously and they have a high incidence of recurrent infection.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the institutional ethics committee

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Cite this article as: Shenoy NS, Tiwari C, Gandhi S, Dwivedi P, Shah H. Anomalies of branchial cleft: our experience and review of literature. Int Surg J 2017;4:3234-7.