Second Branchial Anomalies: A Study of 94 Cases

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Abstract Ninety-four patients with second branchial anomalies were retrospectively analysed at a tertiary care centre from January 2006 to September 2016 to determine the demographical data and management. Branchial sinus and fistula presented earlier as compared to branchial cyst. The mean age at presentation in case of branchial sinuses, fistulae and cysts was 5.07, 5.79 and 7.31 years respectively. There was preponderance in males as compared to females, more so in bilateral cases. Male to female sex ratio was 2.91:1. The branchial fistulae were the most common type of lesions, followed by the branchial sinuses. The branchial anomalies were more on the right side (65.96%) probably due to right handedness of the population. Only eight patients (8.51%) had bilateral anomalies. Four patients had familial association, it was seen in bilateral cases and they presented earlier than unilateral cases. Early and complete surgical excision is the treatment of choice. Preoperative sinogram/fistulogram and intraoperative methylene blue dye injection is not mandatory for excision of a branchial sinus/fistula. Post-operative wound infection was the most common complication (4.25%).

Keywords Branchial anomalies · Branchial sinus · Branchial fistula · Branchial cyst · Children

Introduction

Branchial anomalies are common congenital lesions of the neck in the pediatric age group causing approximately 20% of the cervical lesions in children [1]. Second branchial complex anomalies are most common, accounting for up to 95% of these lesions and can present as a cyst, sinus or fistula in the neck [2]. Despite the prevalence of these anomalies, diagnosis is not always straightforward, as clinicians may not consider them in the differential diagnosis. Pathogenesis of branchial anomalies is not entirely certain. Several theories proposed to explain their development include the branchial apparatus theory, cervical sinus theory, thymopharyngeal duct theory and inclusion theory [3]. Out of these, the most widely accepted theory is that branchial anomalies result from incomplete involution of the branchial apparatus [4]. Surgical excision is the definitive mode of treatment for the second branchial anomalies as these lesions do not regress spontaneously and are associated with a high rate of recurrent infection.

Patients and methods

The present study included 94 patients of second branchial anomalies, which presented at a tertiary care centre of North-India, from January 2006 to September 2016. The available records of the patients were retrieved from the department. The patient's age, sex, presenting symptoms, family history, previous history of infection and surgery were noted. Physical examination included the site and side of the cervical lesion and associated anomalies in other parts of the body. Radiological investigations, medical and surgical treatment including surgical findings, post

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operative morbidity and outcome were reviewed. The histopathological reports were also retrieved.

Results

A total of 94 patients of second branchial complex anomalies were studied retrospectively. The youngest patient in our study was 3 months old, while the oldest patient was 14 years old. The mean age was 5.56 years. The mean age at presentation in case of branchial sinuses, fistulae and cysts was 5.07, 5.79 and 7.31 years respectively (Fig. 1). Out of 94 cases, 70 patients (74.47%) were males, while 24 (25.53%) were females (Fig. 2). Branchial cysts presented as a lateral neck mass. Branchial fistula and sinus presented with a skin opening located close to the anterior border of the sternocleidomastoid muscle with or without discharge from the cervical opening. In one of our case, milk was dribbling from the external opening of the fistula. Out of the 86 unilateral cases, branchial fistulae were observed in 44 (51.16%), branchial sinus in 34 (39.53%) and branchial cysts in 8 patients (9.3%) (Fig. 3). Branchial anomalies were more on the right side (62 cases i.e. 65.96%), while 24 (25.53%) occurred on the left side. Bilateral anomalies were observed in 8 patients (8.51%). Among eight bilateral cases, four patients had fistulae on both the sides while other four had fistula on one side and sinus on the opposite side. Out of the eight bilateral cases, four patients (50%) had familial association, six patients (75%) were male. The mean age of presentation in the bilateral cases was early (4.73 years) as compared to the unilateral cases. One patient had anomaly of the external ear, while one patient was diagnosed with pelvi-ureteric junction obstruction. Surgical excision in all of the patients

Fig. 1 Bar diagram showing mean age at presentation (in years)

Fig. 2 Pie chart showing gender distribution of branchial anomalies

was carried out under general anaesthesia. Wound infection occurred in four patients (4.25%). The recurrence was observed in two patients (2.12%) who had multiple pre-operative infections.

Discussion

The branchial apparatus appears in the fourth week of embryonic life, has a vital role to play in the development of head and neck structures. The branchial apparatus consists of grooves (ectoderm), arches (mesoderm) and pouches (endoderm). The branchial complex anomalies result from the developmental abnormalities of the branchial apparatus during the embryonic period and so it is a misnomer to use only the word “cleft” when describing these congenital malformations, as this implies that there is no contribution from the branchial arch or pouch. The branchial cysts are considered to be entrapped remnants of branchial clefts or sinuses; sinuses are remnants of clefts or pouches; and fistulae result from persistence of both pouch and cleft [4]. During embryonic development, the second arch grows caudally, enveloping the third, fourth and sixth

Fig. 3 Distribution of unilateral branchial anomalies

arches and fusing with skin caudal to these arches, forming a deep groove (cervical sinus). It is generally accepted that most branchial anomalies are derived from remnants of this sinus [5].

The branchial anomalies are usually diagnosed in infancy and sometimes in childhood. However some lesions may not be recognised until adulthood. The mean age at presentation in our study was 5.56 years. Karabulut et al. [6] reported that the mean age at presentation was 5.30 years. A sinus or fistula tends to be diagnosed earlier than a cyst, as cysts may take a longer time to enlarge and present clinically [4]. The mean age at presentation in our study was 5.07, 5.79 and 7.31 years for branchial sinuses, fistulae and cysts respectively. Schroeder also reported that the mean age of presentation of cysts was late compared to that of fistulae and sinuses [7]. There were 70 males and 24 females with a male:female ratio of 2.91:1. Simpson reported a 3:1 male:female ratio, which is almost similar to the ratio of 2.91 in the present series [8]. Careful history and physical examination are the most important elements in the diagnosis of a branchial anomaly. Equally important is high index of suspicion and clinical awareness. Choi and Zalzal [4] reported that the most common presenting features were discharge from the cervical openings, cervical mass, and repeated infections. In our study the most common presenting features were drainage from a cervical opening and a cervical mass. Choi and Zalzal [4] reported a maximum incidence of sinuses, followed by the fistulae. However we observed that branchial fistulae were the most common lesions followed by branchial sinuses. Ford et al. reported that branchial anomalies occur more on the right side of neck. They had 60% incidence on the right side and 40% on the left side [9]. In the present study, we observed almost a similar picture with right side preponderance of

65.95% as compared to 34.04% on left side in unilateral cases. The right side preponderance is described in the literature, however, there is no explanation for this. We propose that right sided preponderance may be related to that fact that majority of population is right handed in Northern India. The incidence of bilateral branchial anomalies is reported in the range of 1–30% [10], we observed an incidence of 8.51%. The branchial anomalies are very infrequently associated with familial tendency [8]. The familial association was observed in two pairs of siblings in the present study, all of these patients had a bilateral involvement.

Pre operative sinogram/fistulogram is helpful in defining the anatomy of a sinus or fistula [11]. But these were not used in our study as children are often too uncooperative to cannulate, and require sedation or anaesthesia for cannulation. Further, inflammation, scarring and recent infections can lead to false-negative results. Potentially harmful radiation exposure to the children can be avoided if fistulogram is not carried out. In our study ultrasound was used in all of the patients with branchial anomalies. The second branchial cysts may very rarely be associated with the mediastinal extension and can lead to diagnostic challenge, so MRI scan neck/thorax should preferably be done if there is doubt of mediastinal extension on USG neck.

The definitive treatment for the branchial anomalies is early and complete surgical excision. The lesions that remain unresected have a high risk of infection, and the incomplete resection results in high recurrence rate. An elliptical cervical incision was used for excision of the branchial fistulae and sinuses. The complete excision of a branchial fistula requires gentle traction on the cord like tract to avoid breaking it and it can be patiently dissected up to the tonsillar fossa (Fig. 4). The termination of tract in

Fig. 4 Operative photograph showing excision of branchial fistula

patients of branchial sinus was observed as loss of firmness and fanning of soft tissues from the medial end of the tract. The sinus or fistula tract may be injected with methylene blue, it helps in delineating the course of tract. But methylene blue can stain surrounding tissues and make the rest of the dissection more challenging. Hence methylene blue was not used in any case. We used a transverse cervical incision within the natural skin fold of the neck for branchial cyst. A branchial cyst may sometimes be associated with a fistulous tract, which needs special attention for complete excision to avoid recurrence.

In our series, the most common postoperative complication observed was wound infection in 4.25% patients. Schroeder et al. [7] also reported that wound infection is the most common minor complication, it developed in 6.75% of the cases. Recurrence rate after excision vary from 3% in the fresh cases to 20% in second surgical attempts [3]. Recurrence rate of 2.12% was observed in our series. The low incidence of complications in our series was probably due to tertiary nature of our institute and extensive experience of the senior surgeon.

Conclusion

In conclusion branchial sinus and fistula presented earlier as compared to branchial cyst. There was familial association in bilateral cases. Early and complete surgical

excision is the treatment of choice to prevent repeated infections, scarring and multiple surgical procedures. Pre-operative sinogram/fistulogram and intraoperative methylene blue dye injection is not mandatory for excision of a branchial sinus/fistula.

Compliance with Ethical Standards

Conflict of interest The authors declare no conflict of interests.

Informed consent Informed consent was taken at the time of surgery. This study is a retrospective analysis of operated patients. This surgery is routinely done in our hospital and worldwide and so no ethical aspect is involved in this study.

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