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Branchial anomalies in children: A report of 105 surgical cases

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ABSTRACT

Background: Branchial anomalies (BAs) account for 20% of all congenital masses in children. We sought to review the incidence of involvement of individual anomalies, diagnostic methods, surgical treatment, and complications of BAs in children. In addition, we also classified our study and analyzed a congenital lower neck cutaneous fistula near the sternoclavicular joint that was thought to be the skin-side remnant of the fourth BAs.
Methods: We conducted a retrospective analysis of 105 children who were referred to our hospital from June 2009 to December 2016 for the treatment of BAs.
Results: In this series, there were 51 males and 54 females. The age at the time of operation varied from 19 days to 13 years, and the mean age was 4.5 years. A total of 33 (31.4%) cases presented with first BAs, 13 (12.4%) presented with second BAs, and 59 (56.2%) presented with third and fourth BAs, including 6 cases of congenital lower neck cutaneous fistula. Fistulectomy under general anesthesia was performed on all of them. For post-operative complications, 2 cases had temporary facial paralysis, 1 case had permanent facial paralysis, 4 cases had temporary recurrent laryngeal nerve injury. Recurrence occurred in 2 patients with first BAs after medium follow-up time of 3.6 years (6 months–8 years).
Conclusions: BAs are common congenital head and neck lesions in children, and there are four distinct types (first, second, third and fourth anomalies). The incidence of third and fourth BAs in Asia maybe higher when compared with literature reports, second BAs seem rare in this population, but more research is needed to confirm this perspective. Diagnosis is not difficult with a proper knowledge of the anatomy of the BAs. The surgical procedures should be tailored depending on the various types, and complete excision of the fistula is the key to prevent recurrence.

1. Introduction

Branchial anomalies (BAs) account for 20% of all pediatric congenital masses and are the second-most common mass after thyroglossal duct cysts and sinuses [1]. The incidence rates in males and females are the same. BAs encompasses first, second, third and fourth branchial cleft cysts, fistulas, and sinuses. Although some theories about the origin of BAs have been reported (thymopharyngeal duct theory, parotid gland inclusion theory, degenerative cystic changes of cervical lymph nodes theory) [2]. The most widely accepted theory is the incomplete involution of the branchial apparatus during embryogenesis.

Branchial structures begin to develop early in the fourth week of the embryonic stage. By the end of the fourth week, there are four unequivocal pairs of branchial arches and two extra rudimentary arches that are invisible on the surface of the embryo, and the mesoderm of the arches is separated by endoderm-lined pouches internally and ectoderm-lined clefts externally. By the seventh week of embryonic life, the pouches and clefts are gradually obliterated by invasion of the surrounding mesenchyme. BAs are residuals of the four primary pairs of branchial pouches and branchial clefts that fail to regress or develop normally [3]. When a cleft or pouch fails to obliterate, it may communicate with either the mucosa of the upper airway or skin, forming a sinus. When both a cleft and pouch fail to obliterate, it may form a communication between the skin and mucosa, forming a fistula. When a cleft remnant forms an epithelial-lined space without communication to mucosa or the skin, a cyst is formed [1].

This article retrospectively analyzed 105 cases of BAs operated on at Shanghai Children's Hospital over the past 8 years. In addition, we also classified our study and analyzed a congenital lower neck cutaneous fistula near the sternoclavicular joint that was thought to be the skin-side remnant of the fourth branchial anomalies [4].

2. Patients and methods

This study was conducted at the pediatric ENT department at Shanghai Children's Hospital, China. All cases who underwent surgery

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Table 1
Features of 105 cases with BAs.

Characteristic	First BAs	Second BAs	Third and fourth BAs	Lower neck cutaneous fistula
No. of patients	33	13	59	6
Sex				
Male	13	8	30	2
Female	20	5	29	4
Age surgery	4.6	4.2	4.6	3.2
Side				
Left	23	4	55	6
Right	10	5	4	0
bilateral	0	4	0	0
Previous treatment				
Incision and drainage	23	2	34	4
Surgical resection	4	0	11	1
Type of anomaly				
Cyst	3	2	0	0
Sinus	16	13	24	6
Fistual	14	2	35	0
Imaging examination				
Enhanced CT	11/33	2/13	17/59	1/6
Ultrasound	1/11	0/2	5/38	0/2
Magnetic resonance	0	0	2/2	0
Complications	2 cases had temporary facial paralysis, 1 case had permanent facial paralysis	0	4 cases had temporary recurrent laryngeal nerve injury	0
Recurrence	2	0	0	0

BAs: Branchial anomalies.

for BAs from June 2009 to December 2016 were included in this study. Eligible patients included those under 18 years of age. The intraoperative course of the fistula tract and pathology results confirmed the diagnosis of BAs. Congenital lower neck cutaneous fistulas were also included. The cases of BAs who just received abscess incision and drainage and who did not undergo surgical therapy during hospitalization have been excluded. The patients with preoperative diagnosis of BAs and postoperative pathological diagnosis of thyroglossal duct cyst, lymphangioma, teratoma and others were excluded. All the surgeries for BAs in our study were accomplished by a senior physician. The patient's medical records were analyzed for demographic data, clinical presentations, diagnosis method, surgical managements of the types of BAs and complications. In addition, the incidence of involvement of individual anomalies was also analyzed.

This study was approved by the local institutional review board, and an informed consent form was signed by the parents of the children.

3. Results

The summary of patients in this series was depicted in Table 1. A total of 105 patients were surgically treated for BAs, and there were 51 males and 54 females. The age at the time of operation varied from 19 days to 13 years, and the mean age was 4.5 years. According to the classification of BAs, 33 (31.4%) presented with first BAs, 13 (12.4%) presented with second BAs, 59 (56.2%) presented with third and fourth BAs, and we included 6 cases of congenital lower neck cutaneous fistula. The BAs occurred on the left side in 82 (78.1%) cases, the right side in 19 (18.1%) cases and was bilateral in 4 (3.8%) cases. A total of 23 were on the left side and 10 were right in the first BAs. 9 were unilateral (4 left and 5 right) and 4 were bilateral in the second BAs. In the third and fourth BAs, 55 were left and 4 were right.

Of the BAs, 59 (56.2%) had a history of abscess incision and drainage, 23 had more than one procedure, and the mean number of times was 1.7 (1–6 times). 15 (14.3%) had a history of surgical resection in other institutions, and the mean number of times was 1.4 (1–3 times). A total of 21 cases were misdiagnosed as other diseases, such as dermoid cyst, lymphangioma, suppurative thyroiditis, mumps, thyroglossal duct cyst, etc. There were 5 (5/109, 4.6%) cysts, 51 (51/109, 46.8%) fistulas, and 53 (53/109, 48.6%) sinuses, of which 4 cases of bilateral lesions of the second BAs were sinuses.

Preoperative fiberoptic laryngoscopy was performed to check the presence of the internal opening of the pyriform sinus for the 53 cases of third and fourth BAs, from which lower neck cutaneous fistula was excluded, but the diagnostic rate was only 36.5%. Computed tomography (CT) scanning with contrast was performed on all cases. CT imaging provides accurate anatomical details of the location, extent, and the importance of the relationship between organizational structures, and there were 30 (28.6%) cases diagnosed as BAs. A total of 51 had an ultrasound, which can determine the nature and size of the lesion, and 6 (11.8%) were diagnosed as BAs. Magnetic resonance imaging was performed in 2 cases, all diagnosed as BAs.

Fistulectomy under general anesthesia were performed on all the patients as a conclusive treatment. The mean hospital stay was 3 days (1–7 days) postoperatively. The nasal feeding tube was placed on day 3 after the operation in the third and fourth BAs, which excluded lower neck cutaneous fistulas. For postoperative complications, in the first BAs, 2 cases had temporary facial paralysis that disappeared after one month, 1 case had permanent facial paralysis, and recurrence was reported in two cases. In the second BAs, the postoperative period was uneventful, and the patients had no relapse. In the third and fourth BAs, 4 cases had temporary recurrent laryngeal nerve injury; satisfactorily, no recurrence occurred postoperatively. All patients were followed up for 6 months to 8 years, and the median time was 3.6 years. The follow-up time of some patients was relatively short, Therefore, the curative effect and the recurrence should be further tracked.

4. Discussion

Second BAs, which account for 95%, are the most common; first BAs account for 1–4%, and third and fourth branchial anomalies are rare [5]. Y. Bajaj reported 80 cases of BAs in the United Kingdom. Second clefts account for 77.5%, first clefts account for 18.8%, and fourth clefts account for 3.7% [6]. In our study, 31.4% presented with first BAs, 12.4% presented with second BAs, and 56.2% presented with third and fourth BAs; the incidence of the third and fourth BAs was the highest, while that of the second BAs was the lowest. We found that the incidence of various types of BAs in China was different from the most of the published literature, and there was only one literature report that suggested the incidence of third and fourth BAs appeared to be higher in local Asian populations, while that of second BAs was lower [7]. Therefore, we also support that the incidence of third and fourth BAs in Asia may be higher, and attention should be paid to reduce the misdiagnosis rate of the third and fourth BAs in the clinical diagnosis and treatment process. However, our study population, including the data of China, does not represent all Asian countries and races, so more research is needed to confirm this perspective.

The third and fourth BAs have variations in the anatomy. The third BAs are often proposed to originate from the base of the pyriform fossa (cranial end), which passes through the thyrohyoid membrane and is located above the superior laryngeal nerve, but the fourth BAs originate from the apex of the pyriform fossa (caudal end), which passes through the cricothyroid membrane and is located beneath the superior laryngeal nerve [8]. Repeated infections of third and fourth BAs are prone to occur in the initial part of the lateral pharyngeal wall, which makes it difficult to clearly identify the course of the fistula tract. Therefore, it is not easy to distinguish the third and fourth BAs in the surgical operation based on the relationship between the fistula tract and superior laryngeal nerve [9,10]. We consider that the third and fourth BAs are the same on the clinical manifestations and surgical methods, the