

Bilateral first and second arch anomalies: a rare presentation

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ABSTRACT Branchial sinuses are one of the most common congenital anomalies present. They are usually unilateral; bilateral cases are present but are rare. The presentation of bilateral branchial sinus anomalies along with bilateral first arch anomalies is very rare. Here, we present a case of bilateral first arch anomalies co-existing with bilateral second arch anomalies in a patient with no related family history and no associated syndrome.

Keywords: bilateral, branchial, first arch, second arch
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INTRODUCTION

A branchial anomaly is a congenital developmental defect that arises from the primitive branchial apparatus.⁽¹⁾ Fistulas of the second branchial cleft are the most common branchial anomalies, accounting for as many as 90% of all branchial cleft fistulas.^(2,3) However, bilateral presentation of branchial fistulas is very uncommon, with an incidence of 2%–3% and a familial component in most cases.⁽⁴⁾ Pre-auricle sinus, a common first arch anomaly, occurs bilaterally in 25%–50% of the presentations. Not only is the incidence of bilateral branchial sinus presentation rare, but the simultaneous presence of both first and second arch anomalies, both bilaterally, is even rarer. A review of the literature yielded only two such cases.^(5,6)

CASE REPORT

A 15-year-old Caucasian boy presented to our department with bilateral pinpoint openings in the lower one-third of the neck. The patient reported having these small openings since birth, with occasional fluid discharge. The amount of discharge would increase during episodes of upper respiratory tract infections and subside with antibiotic treatment. The patient also had bilateral pre-auricular sinuses with no discharge (Fig. 1). The pinna was normal in shape, size and position, and the tympanic membrane was normal. Pure-tone audiometry showed that the patient had normal hearing. Bilateral fistulography indicated the presence of complete branchial tracts with spillage of contrast material in each tonsillar fossa. Ultrasonography of the abdomen and renal examination revealed normal kidney function, thus ruling out the possibility of branchio-oto-renal syndrome. Both the branchial fistulas were excised via stepladder incisions on both sides of the neck (Fig. 2). The patient's postoperative course was uneventful, and he remained symptom-free at the two-year follow-up.

DISCUSSION

The embryonic development and differentiation of the branchial

apparatus occurs between the third and seventh week in the human embryo. The first developing feature is the appearance of five ridges on the ventrolateral surface of the embryonic head. These ridges represent branchial arches; each consists of a core of mesenchyme, covered externally with ectoderm and internally with endoderm. Thus, each adjacent arch is separated externally by ectodermic clefts and internally by endodermic pouches. The clefts and pouches approach each other to form a closing membrane. Invasion of the surrounding mesenchyme gradually obliterates the cleft and pouch in humans.

The second arch first increases in thickness and proliferates caudally to meet the enlarging epiperocardial ridge of the fifth arch with further embryonic development. As it proliferates, the second arch extends over the third and fourth branchial clefts, resulting in the formation of an enclosed ectodermally lined cavity, the cervical sinus of His. With further development, the sinus obliterates by the opposition and fusion of its walls, yielding a smooth uniform contour to the external surface of the neck. Failure of the tract to obliterate would result in the formation of a sinus, termed as the branchial sinus. These sinuses are typically seen at a site along the anterior border of the sternocleidomastoid muscle. The tract crosses superiorly lateral to the common carotid artery, glossopharyngeal nerve and hypoglossal nerve, and lies between the internal and external carotid arteries. The sinus often ends close to the middle constrictor muscle, but in some cases, the sinus may open into the region of the tonsillar fossa.

The first branchial anomalies were classified into two groups by Work in 1972.⁽⁷⁾ Type I is considered a duplication of the membranous external auditory canal. A cystic mass in the postauricular area extends medially and anteriorly along the external auditory canal. It usually passes lateral to the facial nerve and ends at the bony meatus. The cyst is lined with stratified squamous epithelium, indicating ectodermic origin. Type II anomalies are considered duplication of the membranous external auditory canal and pinna.⁽⁷⁾ A sinus passes from an

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Fig. 1 Pre-operative photograph shows the clinical setup of the patient, with the bilateral pre-auricular and bilateral sinuses being cannulated for depiction of sinus tracts.

external opening high in the neck along the anterior border of the sternocleidomastoid muscle, superficial or deep to the facial nerve and closely related to the parotid gland. It usually ends blindly near the floor of the membranous external auditory canal, or opens into the canal in the same area as a complete fistula. It is lined with skin and cartilage, indicating ectodermal and mesodermal origin.

It is sometimes difficult to decide whether these pre-auricular sinuses and cysts are inclusion dermoids resulting from epithelium trapped between the developing auricle tubercles or remnants of the first branchial groove epithelium that have failed to re-absorb. Attempts have been made by Work⁽⁷⁾ in 1972 and Aronsohn et al⁽⁸⁾ in 1982 to support one or the other pathogenesis. However, if the pre-auricular sinuses do arise from a lack of fusion of the six hillocks of His, then such sinuses are usually very small and present as pits or blind superficial sinus. However, in our patient, the sinus tract reached the bony meatus. The length of the cannula inserted in the pre-auricular region (Fig. 1) attests to the fact that the sinus tract ran deeper. Work, in his classification of Type 1 branchial cleft sinus, stated that these sinuses were a result of duplication of the membranous external auditory meatus and that the tract reached the bony meatus in these cases.⁽⁷⁾ This fact has been dually supported by Eills⁽⁹⁾ and Gupta et al⁽⁶⁾ in their studies.

The inheritance pattern of pre-auricular sinuses have shown incomplete autosomal dominant pattern, with reduced penetration and variable power of expression. They can arise spontaneously. Bilateral sinuses occur in 25%–50% of cases and



Fig. 2 Peri-operative photograph shows dissection of the branchial sinus tract in the neck via stepladder cervical incision.

are more likely to be hereditary.⁽¹⁰⁾ This is unlike our case where there was no family history. A complete fistula of the third arch and pouch has yet to be reported; however, if it were to occur, the possible route would be caudal to the glossopharyngeal nerve, over the superior laryngeal nerve, posteromedial to the internal carotid artery and piercing the thyrohyoid membrane to enter the pharynx just cephalic to the superior laryngeal nerve.⁽¹¹⁾ Complete fourth arch fistula has also yet to be described in the literature, but there is an established clinical pattern for internal sinuses, which are almost always left-sided and present as suppurative thyroiditis.⁽¹¹⁾

Branchial cleft anomalies such as branchial cysts, sinuses and fistulas are typically seen in the paediatric population.^(10,11) About 96%–97% of these anomalies are unilateral and only 2%–3% have a bilateral presentation; the rate of bilateralism is higher in familial cases.⁽⁴⁾ A review of the English literature found only six cases of bilateral branchial cleft fistulas.^(4,6,12–15) Similar to the cases reported in these studies, our patient also presented with bilateral branchial sinuses. In all the previously reported cases of bilateral cleft anomalies, the patients had a family history of such anomalies, unlike our patient, who had no such history. Similar findings have also been observed by Gupta et al in their case report.⁽⁶⁾

The treatment protocol for such lesions is surgical excision. Antibiotics are only used to treat infections in the tract. Surgical excision is a definitive treatment, which is best delayed until the patient is at least three months old. Of the five reported cases of bilateral branchial sinus, Gatti and Zimm reported two cases,⁽¹³⁾ both of which exhibited the classic constellation of symptoms and were successfully treated with complete surgical excision. A series of horizontal incisions, known as a stair-step or stepladder incision, was made to fully dissect the occasionally tortuous path of the tract. The tract was pulled sequentially from this stepladder incision and followed up till the tonsillar fossa, as shown in Fig. 2. However, definitive surgery should not be attempted during an episode of acute infection or if an abscess is present. Surgical incision and drainage of the abscess is indicated, if present, usually along with concurrent antimicrobial therapy.

In conclusion, simultaneous presentation of first and second arch anomalies is very rare. To the best of our knowledge, only three cases with similar findings have been reported. Such cases should therefore be documented and a thorough investigation done to rule out any other congenital anomalies.

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