

A Bilateral, Non-syndromic, Type III Second Branchial Arch Sinus in a Neonate: a Case Report

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ABSTRACT

The incidence of a second branchial arch sinus accounts for 26–60% of all existing congenital malformations deriving from the branchial apparatus. They are most usually detected between 14 months and 7 years of age, while their incidence during neonatal period and infancy accounts for 0.06% of all cases.

The aim of this case study is to emphasize three rare characteristic features: the manifestation during neonatal period, the bilateral localization and the ultrasonographic diagnostic documentation.

A 25 days old girl was admitted by her parents due to the presence of mucoid excretion from two small openings found on the neck. These openings were found bilaterally, between the mid and lower third of the anterior border of sternocleidomastoid muscle. Diagnosis was confirmed via ultrasonography. The patient underwent elective surgery during early infancy and both branchial fistulas were excised. Patient's postoperative course was uneventful.

In conclusion:

- in cases of a bilateral second branchial arch sinus, the branchio-oto-renal (BOR) or branchio-otic (BO) syndromes must be excluded;
- ultrasound scan can be used for the thorough evaluation of the sinus anatomic course and the relationship with the adjacent anatomic structures;
- prompt diagnosis and early therapeutic intervention, even during neonatal period, ensures an uneventful post-operation course.

KEYWORDS

bilateral branchial sinus; second branchial arch; ultrasonography; neonate

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INTRODUCTION

“Branchia” is the Greek word for gill, and the same word describes the corresponding anatomic structures, due to their resemblance to fish gills. Branchial apparatus plays a vital role in the development of head and neck structures. Six paired branchial arches, develop from the 4th to the 6th fetal week. Each branchial arch consists of a mesenchymatous core covered externally by ectoderm and internally by endoderm (1).

The incidence of second branchial arch deformities accounts for 26–60% of all existing congenital malformations deriving from the branchial apparatus. They are most frequently detected between the 14th month and the 7th year of life, while their incidence during neonatal period and infancy accounts for 0.06% of all cases (2) Second branchial sinuses are the most common branchial anomalies (up to 97% of all second branchial apparatus anomalies) and they are usually found unilaterally (2, 3).

The aim of this case study is to emphasize four uncommon features: the manifestation during neonatal period, the bilateral localization, the ultrasonographic diagnostic documentation and the absence of branchio-oto-renal (BOR) syndrome features.

CASE REPORT

A 25 days old girl was admitted to our Department by her parents due to the presence of mucoïd excretion from two small openings found on the neck. These openings were found bilaterally, between the mid and lower third of the anterior border of sternocleidomastoid muscle (Figures 1, 2).

Ultrasonography revealed the presence and anatomic course of two branchial sinuses, 1.2–1.3 mm in diameter. They were located subcutaneously, penetrating the platysma, passing between the carotid bifurcations and leading to the peritonsillar fossa (Figure 3).

Preoperative evaluation did not reveal any abnormality, while familial predisposition was not documented. More specifically, in the context of thorough physical examination there were no distinctive features indicative of cranio-facial deformities or ocular abnormalities. ENT examination was also normal, without indications of hearing loss. Ultrasonographic evaluation of the urinary tract did not reveal any pathology.

The patient then underwent elective surgery under general anaesthesia. The two branchial sinus tracts were initially catheterized with the use of a 4 Fr catheter. These were then used to lead in, while the sinus tract was dis-



Fig. 1, 2 External opening of the right (figure 1 – arrow) and left sinus (figure 2 – arrow), respectively.



Fig. 3 Ultrasonographic image of the right branchial sinus.

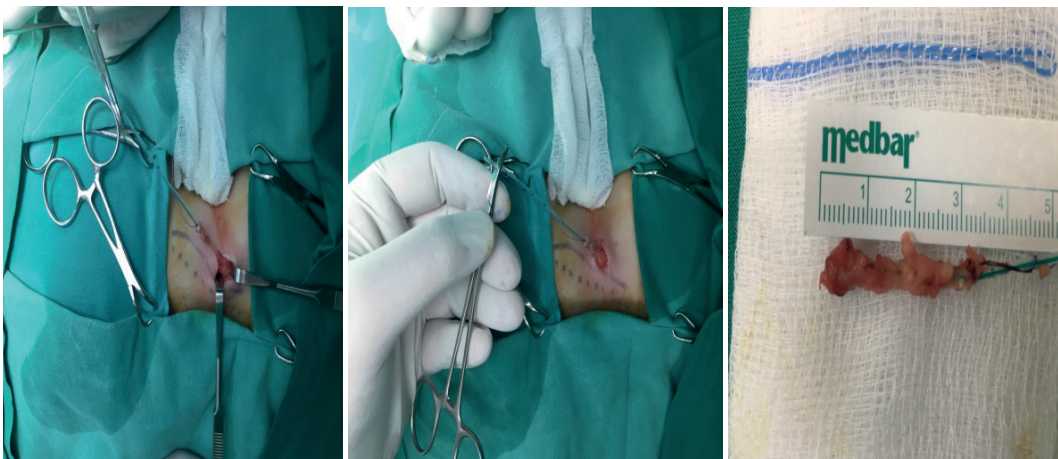


Fig. 4–6 Notice the step by step meticulous dissection of the left branchial sinus.

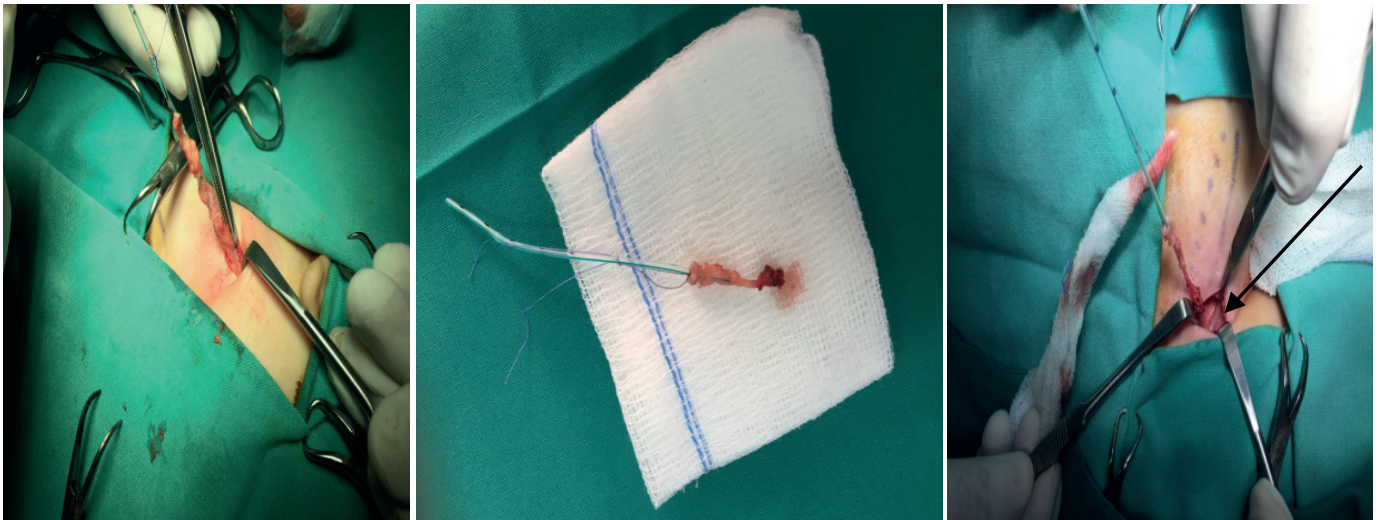


Fig. 7-9 Notice the step by step meticulous dissection of the right branchial sinus, passing between the carotid bifurcation. Figure 8 depicts the internal jugular vein and under this, the internal carotid artery (arrow)

sected through a step-ladder incision until the complete excision of each tract at the level of tonsillar fossa. A second incision was not required. Traction of the leading catheter assisted to the better dissection of the branchial sinus (Figures 4-9).

Patient's postoperative course was uneventful and she was discharged home the next day in good general condition. The excised specimens were subjected to histopathological examination, which showed that they were lined by squamous epithelium, while cartilage remnants and lymphoid tissue were found in the subepithelium.

One year later, patient remains asymptomatic without indications of recurrence.

DISCUSSION

Anomalies of the second branchial arch can be found in the anterior-lateral surface of the neck along with the anterior border of the sternocleidomastoid muscle. Classification of second branchial apparatus sinuses is based upon their anatomic course and the relationship with the adjacent anatomic structures (Table 1). In this case report, we present a type III second branchial sinus (3).

After meticulous research of the relevant literature, there are less than 7 reports of second branchial sinuses found bilaterally, while 6 of them were associated with familial predisposition (4-8). Bilateral manifestation along with absence of familial predisposition were the two key points in our case.

Clinical manifestation of a second branchial fistula during neonatal period is very rare, even when it manifests unilaterally. After meticulous bibliographic research, we did not find another case of second branchial fistula diagnosed during neonatal period (9-13). Prasad and colleagues reported another case of remnant of the second branchial apparatus, which was diagnosed and surgically treated at the age of 5 months, without determination of the specific type of this anomaly (3).

Tab. 1 Classification of second branchial apparatus sinuses.

Type	Characteristics
I	Branchial sinus penetrates the platysma and ends to the deep cervical fascia
II	Branchial sinus extends medially between the external and internal carotid artery
III	Branchial fistula passes between carotid bifurcation, then penetrates the stylohyoid muscle and the posterior surface of the digastric muscle towards ipsilateral pharynx to end up to the tonsillar fossa. Rarely, there are two complete openings. The sinus passes next to the glossopharyngeal and hypoglossal nerves, behind the stylohyoid ligament.
IV	Branchial sinus passes between the carotid artery bifurcation and ends blindly.

In the largest relevant retrospective study, Yang and colleagues report 28 cases of branchial apparatus anomalies that they encountered, while, only 5 of them (16.7%) derived from the second branchial apparatus. The youngest patient of their study group was 1 year old (14). Smith and Kielnovitch reported a case of a newborn with branchial cyst, remnant of second branchial apparatus (15).

In a retrospective study conducted by Maddalozzo and colleagues, including 28 cases in total, three patients (11%) had bilateral second branchial arch sinus. In two out of those three cases (66%) there were features indicative of BOR syndrome. Mutation of EYA1 gene is implicated as the commonest cause of BOR syndrome. In cases of this syndrome, hearing loss's incidence reaches up to 90%, with the mixed type being the most prevalent. Hearing loss may progress during the course of life in 12.8% of all cases (12). There are also cases, in which no renal involvement is documented, thus it is called branchial-otic (BO) syndrome (16).

In terms of exclusion of BOR syndrome in our case, we used the criteria proposed by Chang and colleagues. Based upon those criteria, in order to establish the diagnosis, the patient must fulfill three major criteria or two major and

two minor criteria, when there is no familial predisposition. Our patient fulfilled only one major criterion, the presence of bilateral second branchial arch anomalies, with no minor criteria. Thus, the clinical diagnosis of BOR syndrome was excluded. After that, we did not consider as necessary the performance of genetic testing in our patient (17).

Determination of the anatomic course of a second branchial sinus constitutes the basis for safe and successful treatment. Most frequently used methods for this aim are: a) conduction of fistulogram preoperatively and b) intraoperative injection of blue de methylene via the external opening of the sinus (1-3).

However, Prasad et al., in their series consisting of 17 cases of second branchial sinus, documented the anatomic course via ultrasonography in 9 patients (3). In our case, ultrasonography contributed crucially to the preoperative determination of the anatomic course of the sinuses.

There is indication of surgical excision of second branchial sinus early after diagnostic documentation. Major risks are: suppuration development of solid adhesions that lead to laborious and unsafe surgical excision, due to the adjacency to major vessels and hypoglossal nerve. Another main risk that must be avoided in clinical practice is the possible development of branchial carcinoma in the future (3). Alternative treatment methods such as endoscopic electro-coagulation and chemical cauterization are considered as unsafe in pediatric population (18).

In conclusion:

- in cases of a bilateral second branchial arch sinus, the branchio-oto-renal (BOR) or branchio-otic (BO) syndromes must be excluded;
- ultrasound scan can be used for the thorough evaluation of the sinus anatomic course and the relationship with the adjacent anatomic structures;
- prompt diagnosis and early therapeutic intervention, even during neonatal period, ensures an uneventful post-operation course.

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