

## Bilateral branchial cleft anomaly type two and type three seen together

### *Tip iki ve tip üçün birlikte görüldüğü bilateral brankial yarık anomalisi*

Cüneyt Kucur<sup>1</sup>, Vefa Kınış<sup>2</sup>, Yusuf Eren<sup>3</sup>, Ali Okan Gürsel<sup>3</sup>

<sup>1</sup>Zeynep Kamil Women and Children Diseases Hospital, Dept. Otorhinolaryngology, İstanbul, Turkey

<sup>2</sup>Dicle University, School of Medicine, Dept. Otorhinolaryngology, Diyarbakır, Turkey

<sup>3</sup>Fatih Sultan Mehmet Education and Research Hospital, Dept. Otorhinolaryngology, İstanbul, Turkey

#### ABSTRACT

Branchial apparatus begins to develop at about second week of gestation and each complex will transform into different structures in the head and neck. Branchial cleft anomalies develop due to defect in the closure of these structures by time. Branchial cleft anomalies may be diagnosed at any age but most of them are seen in pediatric population. Although, branchial cleft anomalies are frequently seen, bilateral cases, which have been reported are very rare. We present a 14 years old boy who was diagnosed and operated due to bilateral branchial cleft anomaly. *J Clin Exp Invest* 2012; 3(1): 99-101

**Key words:** Branchial cleft anomaly, bilateral, type 2, type 3

#### ÖZET

Yaşamın ikinci haftasından itibaren oluşmaya başlayan her bir brankiyal yapı, baş ve boyun bölgesinde farklı yapıları oluşturmaktadır. Brankiyal yarık anomalileri zamanla kapanması gereken bu yapıların kapanmaması sonucu gelişmektedir. Her yaş grubunda görülebilmekle beraber sıklıkla çocukluk döneminde görülmektedir. Brankiyal yarık anomalileri sık görülmekle birlikte bilateral olarak bildirilen vakalar oldukça nadirdir. Biz 14 yaşında bilateral brankiyal yarık anomalisi teşhisi konulan ve bu nedenle opere edilen erkek hastayı sunduk.

**Anahtar kelimeler:** Brankiyal yarık anomalisi, bilateral, tip 2, tip 3

#### INTRODUCTION

The branchial apparatus made up of pouches (endoderm), arches (mesoderm) and grooves (ectoderm).<sup>1</sup> The development of branchial apparatus begins at about 2nd week of gestation and it is completed by the 6-7 th weeks. Each branchial arch, pouch and groove complex will develop into specific structures in the head and neck region. Branchial cleft anomalies may present as a cyst, sinus or fistula. A branchial cyst is lined by epithelium and has no external or visceral opening and thus retains secretions. Sinus is a blind pocket, communicate either with skin or pharyngeal lumen. Fistula is a tract that has both internal and external openings.

#### CASE

A 14 years old boy is brought to our outpatient clinic with swelling and purulent discharge on both sides of his neck (Figure 1). The family informed us that especially after upper respiratory tract infections, a serous discharge comes from the fistula openings.

The complaints of the patient had started in his 5 years of age and become more frequent and annoying in the last 2 years. On physical examination his vital signs were normal. On head and neck examination there was nontender, soft and mobile mass on both sides of the neck. The mass size was 2x3 cm in the right and 1x1 cm in the other side of the neck. There were openings of fistulous tract located at the anterior-inferior 1/3 part of the sternocleidomastoid muscle (SCM) in the right side and at the anterior middle 1/3 part of the SCM in the left side. On laboratory, WBC (white blood cell count) was 13.000 and there were no other abnormalities. First of all, 1 gr amoxicilline-clavulonic acid two times per day was given for 2 weeks. Then an USG and CT with contrast of the neck were performed. The masses were cystic in character but there was no noticeable tract on radiologic examination. According to the patients history, our physical examination and the radiological findings branchial cleft anomaly (BCA) was thought in the early diagnosis. The family was informed about the diagnosis and surgery was recommended. An elliptical skin incision was

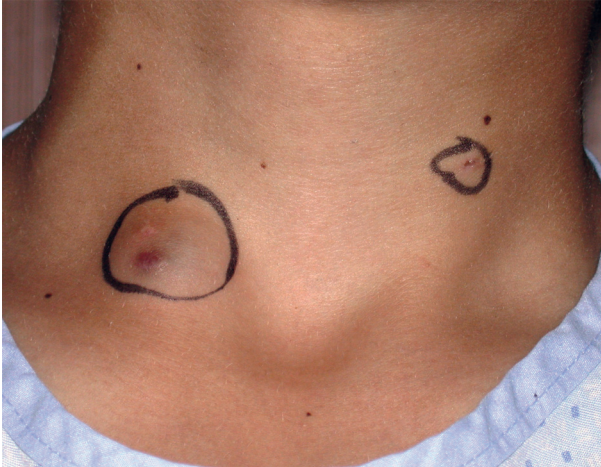
**Correspondence:** Dr. Vefa Kınış

Dicle Üniversitesi Tıp Fakültesi, KBB Kliniği, Diyarbakır, Türkiye Email: vefakinis@hotmail.com

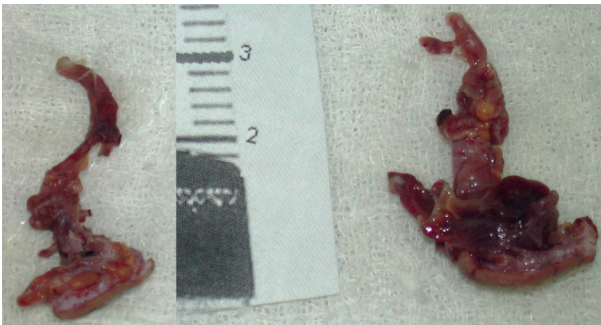
Received: 01.05.2011, Accepted: 05.08.2011

Copyright © JCEI / Journal of Clinical and Experimental Investigations 2012, All rights reserved

done around the fistula opening on both sides. Than tracts were followed until the tonsillar fossa at left side and until the pyriform sinus at right side. Tracts were ligated and excised at both sides (Figure 2). No complications were seen postoperatively.



**Figure 1.** Fistulous openings are seen on both sides of neck.



**Figure 2.** Cysts and their tracts were excised on both sides.

## DISCUSSION

Four type of BCA have been described. Work<sup>2</sup> described 2 types of first branchial anomaly; which are designated type I and type II. Type I anomalies are characterized by duplication of external auditory canal. They occur in the preauricular area and they line in a plane parallel to external auditory canal and lateral to facial nerve. Type II anomalies appear as an opening posterior or inferior to the angle of mandible. The tract is intimately associated with parotid gland and facial nerve.

The second branchial cleft anomaly typically seen at a side along the anterior border of SCM.

The tract crosses superiorly- lateral to the common carotid artery (CCA), the glossopharyngeal nerve and the hypoglossal nerve and it lies between the internal and external carotid arteries. The sinus often ends close to the middle constrictor muscle; in other cases, the sinus opens into the region of tonsillar fossa.

The third branchial anomaly opens into the pharynx in the region of the pyriform sinus. Fistulas of the third branchial cleft, which are rare, also appear at the anterior border of SCM. They ascend lateral to the CCA and they pass posterior to the internal carotid artery, superior to the hypoglossal nerve and inferior to the glossopharyngeal nerve. The tract terminates by piercing the lateral thyroid membrane of the pyriform sinus.<sup>3</sup>

Presumed anomalies of the fourth branchial cleft have been described as traveling below the arteries of the fourth arch (aortic arch, subclavian artery) in the mediastinum along the ascending recurrent laryngeal nerve to the upper esophagus.<sup>4</sup>

Branchial remnants are derived from the first arch in %1-18 of cases. In %54-95 of cases the anomaly is derived from second arch. Abnormalities derived from the third and the fourth arches are quite uncommon- less than %8.<sup>3,5</sup>

Developmental anomalies of the branchial apparatus are not uncommon. In fact, they account %17 of all pediatric cervical masses. They are the most common type of congenital cervical mass.<sup>5</sup> Branchial cleft anomalies are often confused with other causes of masses and infections in the neck such as cystic hygroma, teratoma and lymphoma. There are two clinical presentations; the first and main is a fistulous tract which ends in the skin of the anterior part of SCM, typically at the middle third. Second clinical presentation is an isolated cyst. The symptoms associated with these tracts include permissive or intermittent mucoid drainages, recurrent infection attacks, cellulitis, abscess formation.

In diagnosis CT, USG, and fine needle aspiration can be useful to distinguish cysts from carotid body tumors, cystic metastases of squamous cell carcinoma, adenopathy, cystic hygroma, lymphoma and neurofibromas.<sup>6</sup> But preoperative diagnosis primarily based on physical examination and history.

Almost half of the early diagnosis changes after operation. Definitive diagnosis is made by perioperative findings and with histopathological results.

Widely used and accepted treatment is complete tract excision and high suture ligation of tracts at its end points in the neck.

Although branchial cleft anomalies are common, bilateral cases are very rare. Huang et al. presented 3 bilateral cases in their series of 37 patients.<sup>7</sup> In different studies ratio of bilateralism is %2-3. Most of them are seen in familial cases.<sup>8</sup> In literature there are a few number of bilateral cases which have been reported.<sup>6,9,10</sup> In only 2 of them<sup>9,10</sup> the anomalies seen in right and left neck were different types which were first and second. Our patient had not a family history or any other disease. In addition, our patient is an extremely rare case among bilateral ones, because of different types of BCA were present on left and right sides. We used complete tract excision and high suture ligation of tracts. By this method, recurrence rate is very low and patient had not any problems during follow up.

## REFERENCES

1. Hyndman OR and Light G. The branchial apparatus. *Arch Surg* 1929;19(3):410-52.
2. Work WP. Newer concepts of first branchial cleft defects. *Laryngoscope* 1972;82(9):1581-93.
3. Ford GR, Balakrishnan A, Evans JN, Bailey CM. Branchial cleft and pouch abnormalities. *J Laryngol Otol* 1992;106(2):137-43.
4. Proctor B, Proctor C. Congenital lesions of the head and neck. *Otolaryngol Clin North Am* 1970;3(2):221-48.
5. Kenealy JF, Torsiglieri AJ Jr, Tom LW. Branchial cleft anomalies; a five-year retrospective review. *Trans Pa Acad Ophthalmol Otolaryngol* 1990;42(3):1022-5.
6. L.Shankor, R.Josephson and M.Hawke. The branchial cleft cyst. *J Otolaryngol* 1991;20(1):62-4.
7. Huang RY, Damrose EJ, Alavi S, Maceri DR, Shapiro NL. Third branchial cleft anomaly presentin as a retropharyngeal abscess. *Int J Ped Otorhino* 2000;54(2-3):167-72.
8. Vent J, Grier CG, Leopold DA, Heywood BB. Congenital familial bilateral branchial tracts: a rare case. *Ear Nose Throat J* 2008;87(1):48-50.
9. Randall P, Royster HP. First branchial deft anomalies. A not-so-rare and a potentially dangerous condition. *Plast Reconstr Surg* 1963;31(2):497-506.
10. Gupta AK, Kumar S, Jain A. Bilateral first and second branchial cleft fistulas: a case report. *Ear Nose Throat J* 2008;87(5):291-3.