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CASE REPORT

Bilateral second branchial cleft cysts without any associated congenital abnormality: a case report

Eşlik eden başka bir doğuştan anormalliği olmayan bir hastada iki taraflı ikinci brankiyal yarık kisti

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A thirty-year-old male patient with bilateral second branchial cleft cysts is presented, in whom no association was found with any other congenital abnormality, in particular branchio-oto-renal syndrome. He had a complaint of painless swelling on both sides of the neck. Physical and radiological examinations showed bilateral branchial cleft cysts. A coexisting congenital syndrome could not be documented despite investigations including physical examination, blood biochemistry and complete blood count, audiologic tests, temporal bone computed tomography, and abdominal and renal ultrasonography. Surgical excision was performed on both sides with a three-week interval. Histopathologic examination confirmed the diagnosis. The postoperative period was uneventful, with no recurrence during a follow-up period of 12 months.

Key Words: Branchial region/abnormalities; branchioma/surgery; cysts; head and neck neoplasms/diagnosis/surgery.

İki taraflı ikinci brankiyal yarık kisti saptanan 30 yaşındaki erkek hasta sunuldu; olgunun ilginç yanı, kistlere başka bir doğuştan anormalliğin, özellikle de brankiyo-oto-renal sendromun eşlik etmemesiydi. Hasta boynun iki tarafında ağrısız şişlik şikayetiyle başvurdu. Fizik muayene ve radyolojik incelemeler sonucunda iki taraflı brankiyal yarık kisti tanısı kondu. Genel fizik muayene, rutin kan testleri, odyolojik değerlendirme, temporal kemik bilgisayarlı tomografisi, abdominal ve renal ultrasonografi ile araştırılmasına rağmen hastada başka bir doğuştan anormalliğe rastlanmadı. Boyundaki kistlere üç hafta arayla cerrahi uygulandı. Patolojik değerlendirme sonucu brankiyal yarık kistiyle uyumlu idi. Ameliyat sonrasında herhangi bir sorunu olmayan hastanın 12 aylık izleminde nüks görülmedi.

Anahtar Sözcükler: Brankiyal bölge/anormallik; brankiyom/cerrahi; kist; baş-boyun neoplazileri/tanı/cerrahi.

The branchial system consists of five pairs of mesodermic archs separated by invagination of the ectoderm and endoderm. This system comprises the lower part of the face and muscular and visceral parts of the neck. For this reason, any anomaly in the development of the branchial apparatus may result in the formation of a sinus, fistula, or a cyst.[1]

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Second branchial cleft anomalies are considered the most common anomalies of the branchial system accounting for 95%.[2] They may develop bilaterally, the incidence of bilaterality being different for cleft cysts (2% to 3%) and fistulas (10% to 15%).[3,4]

Bilateral branchial anomalies are generally associated with branchio-oto-renal (BOR) syndrome, which has an autosomal pattern of inheritance. After the definition of BOR syndrome in 1975, all the reported cases of bilateral branchial anomalies have been associated with congenital syndromes, especially BOR syndrome.[5,6] In this report, we present a case of bilateral branchial cleft cysts that were not found to be associated with BOR syndrome.

CASE REPORT

A thirty-year-old man presented with a complaint of painless swelling on both sides of the neck. He stated that the lesions had appeared three months before and enlarged since then. There was nothing significant in his medical history. Nor did he have a history of alcohol consumption or smoking.

On otolaryngologic examination, a non-tender, semisolid, fluctuating mass, about 5 cm in size, was palpated in the upper jugular region on the left side of the neck. On the right side, there was another mass, about 3 cm in size, in the upper jugular region, with similar characteristics. Other findings of physical examination were normal.

Computed tomography (CT) revealed bilateral cystic masses located in the cervical chain in front of both carotid arteries (Fig. 1). Fine-needle aspiration biopsy was performed and a dark-yellowish non-viscous liquid was aspirated from the masses. No microorganisms were present on microscopic examination of the aspirate, and no bacterial growth was detected in culture. The initial diagnosis was made as a bilateral branchial cyst. Investigation into the presence of any congenital syndrome, in particular BOR through blood biochemistry and complete blood count, audiologic tests, temporal bone CT, and abdominal and renal ultrasonography yielded normal results.

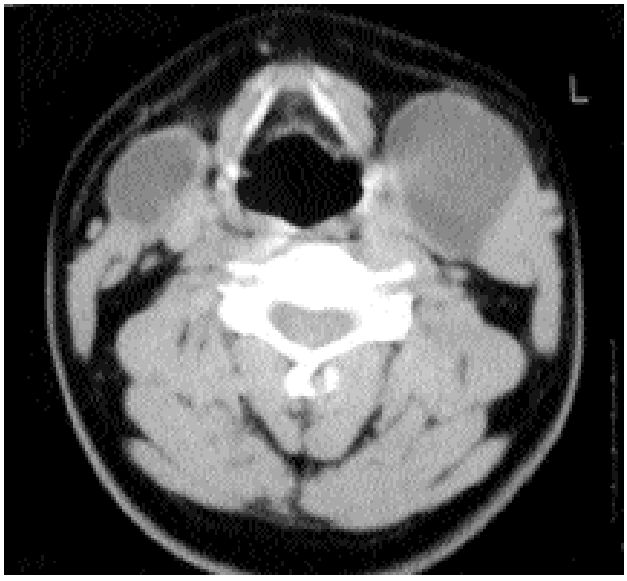

Fig. 1 - *Axial computed tomography image showing both cystic masses located in front of the carotid sheath.*

Both masses were surgically removed with a three-week interval (Fig. 2a, b). Histopathologic examination showed cyst walls covered by squamous epithelium, with dense lymphocytic infiltration forming follicles in some areas. These findings were compatible with a branchial cleft cyst (Fig. 3). The post-

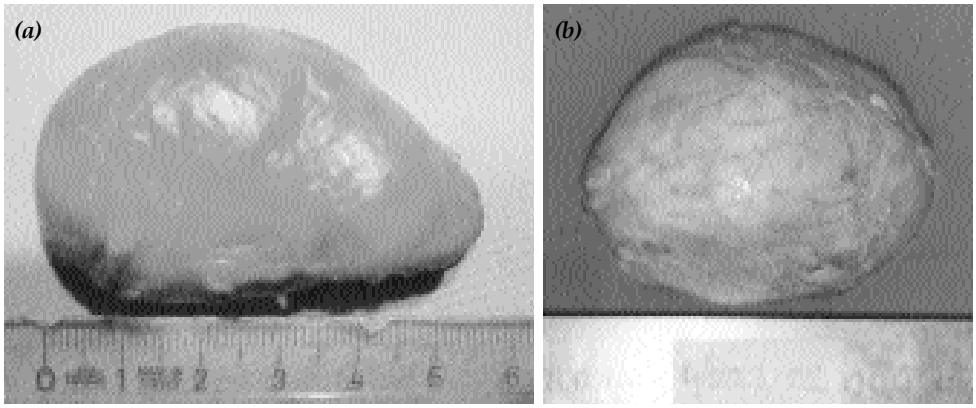

Fig. 2 - ***(a)*** *The left and* ***(b)*** *the right cystic masses after excision.*

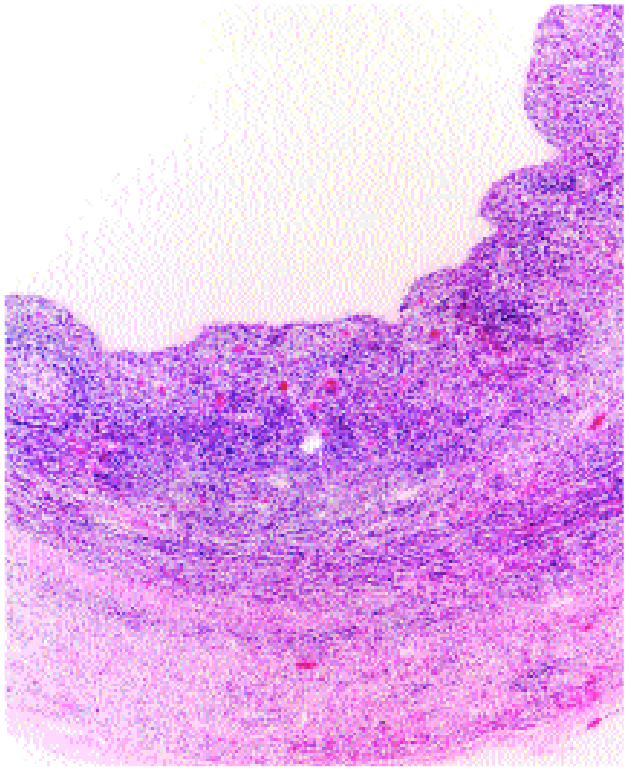

Fig. 3 - *Dense lymphocytic infiltration is seen on the cyst wall covered by squamous epithelium (H-E x 40).*

operative period was uneventful and no recurrence was seen during a follow-up period of 12 months.

DISCUSSION

The development of the branchial apparatus involves a complex series of events that are poorly understood. It begins during the third and fourth gestational weeks and is completed by the sixth and eighth weeks.[7]

Anomalies of the branchial system are divided into four groups depending on their anatomic location; namely, first, second, third, and fourth branchial anomalies. The course of a particular branchial anomaly is caudal to the structures derived from the corresponding arch, and dorsal to the structures that develop from the following arch. Branchial anomalies may be in the form of a cyst, sinus, or a fistula. A cyst is considered to be an entrapped remnant of a branchial cleft sinus, while a sinus is a remnant of a cleft or a pouch, and a fistula results from a pouch or a cleft.[8]

Branchio-oto-renal syndrome, first described by Melnick in 1975, is an autosomal dominant alteration with variable expressivity, including structural anomalies of the outer, middle, or inner ear, branchial fistulas, stenosis of the lacrimal duct, and renal anomalies.[9] Despite its recent description, it is not very unusual, its prevalence being about 1 in 40,000 births. Thus, it may be encountered in up to 2% of children with profound hearing loss.[10]

Bilateral first and second branchial fistulas (not cysts) are frequently seen in BOR syndrome. It was found that all the reported cases of bilateral branchial anomalies since the time of definition of this syndrome were associated with congenital syndromes, most commonly BOR syndrome.[5,6] Of note, no additional anomalies existed in our patient to lead us to the diagnosis of another congenital syndrome, in particular BOR syndrome.

A careful history and physical examination are essential parts of evaluation in branchial anomalies. Second branchial cleft cysts generally present as a non-tender mass anterior to the sternocleidomastoid muscle in the neck. Thus, the diagnosis may be quite straightforward.[8] In cases in which radiologic evaluation is needed, ultrasonography, CT, and magnetic resonance imaging can provide valuable information. Computed tomography is an accurate and noninvasive diagnostic tool to confirm diagnosis or suggest an alternative diagnosis, to define both the location and extent of the lesion, and to delineate an infectious process or malignant transformation. Ultrasonography may help distinguish cystic from solid masses.[4,11]

Branchial cleft cysts should be surgically removed because spontaneous regression is unlikely and there is a high risk for recurrent infections and some likelihood of malignant degeneration. In case of an acute infection, surgery should be delayed three to four weeks for medical treatment.[8]

In conclusion, despite bilateral branchial anomalies are almost always associated with a congenital syndrome, most likely BOR, there may be sporadic cases in which no any other congenital anomaly is encountered.

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