Reference number to be mentioned by correspondence : **B-ENT/18126**

B-ENT, 2018, **14**, 311-313

Cervical chondrocutaneous branchial remnants: often only the tip of the iceberg

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Key-words. Benign neoplasms; neck; branchial arch

Abstract. *Cervical Chondrocutaneous Branchial Remnants: often only the Tip of the Iceberg.* This case study describes a cervical chondrocutaneous branchial remnant (CCBR) in a newborn. This rare, congenital disease manifested as a benign mass on the lateral side of the neck. These masses are typically located in the middle or lower third of the neck, anterior to or over the sternocleidomastoid muscle. The lesions consist of normal skin with a cartilage core. There is no connection to deep underlying structures, but they often adhere to the fascia of the sternocleidomastoid muscle.[1]
CCBRs are often visible markers of serious underlying anomalies. Up to two thirds of patients have associated anomalies, but prevalence varies widely.[2] These anomalies include auditory, gastrointestinal, genitourinary, cardiovascular, musculoskeletal, and visual anomalies, in addition to more complex syndromes.[1] The treatment of choice for a CCBR is surgical excision. A thorough physical examination and ultrasound study of the abdomen and heart are mandatory to rule out underlying serious anomalies.[3]

Introduction

Cervical chondrocutaneous branchial remnants (CCBRs) are benign neck tumours, first reported in 1858 by J. Birkett, who described them as "Congenital supernumerary and imperfectly developed auricles on the sides of the neck".[4] Previously, 118 cases of CCBR have been reported in the English medical literature.[5] The lesions are mostly unilateral (71%), with only 34 cases of bilateral lesions. The majority of unilateral cases are left-sided. Until recently, a marked preponderance of males has been reported, but in a 2015 review by Klockars et al., no gender predisposition was found.[2]

In 1997, Atlan et al. termed these lesions 'CCBRs', but numerous terms were used previously, including branchial cartilages, cervical auricles, cervical accessory tragi, cervical skin tags, cervical vestiges, choristomas, papillomas, fibromas, wattles, and congenital cartilaginous rests of the neck.[6] CCBR lesions are present at birth, and they show no, or very slow, growth. They are typically located in the middle or lower third of the neck, anterior to or over the sternocleidomastoid muscle. The lesions consist of normal skin, with no sign of inflammation, infection, or discharge, and they have a cartilage core. There have been no reports of attachments to deep cervical structures, but some studies have described adherence to the fascia of the sternocleidomastoid muscle.[3] CCBRs were often found to be visible markers of more serious underlying anomalies. Approximately one-third of patients with CCBRs have associated anomalies, although the prevalence varies, and could be as high as two-third of patients. The associated anomalies have involved auditory, cardiovascular, nervous, gastrointestinal, genitourinary, musculoskeletal, respiratory, visual, and endocrine systems.[7] Some studies even reported familial presentations of

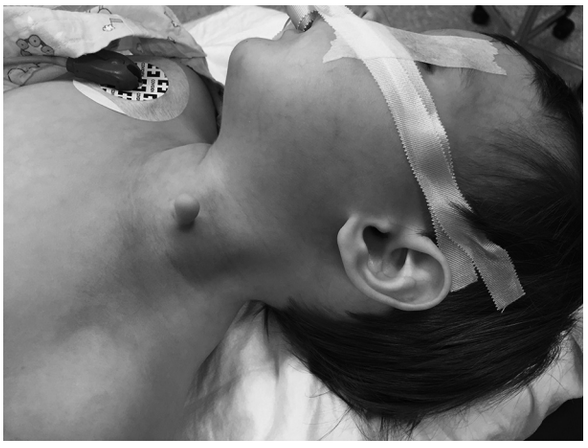

Figure 1
Cervical chondrocutaneous branchial remnant (CCBR) on the lower left side of the neck.

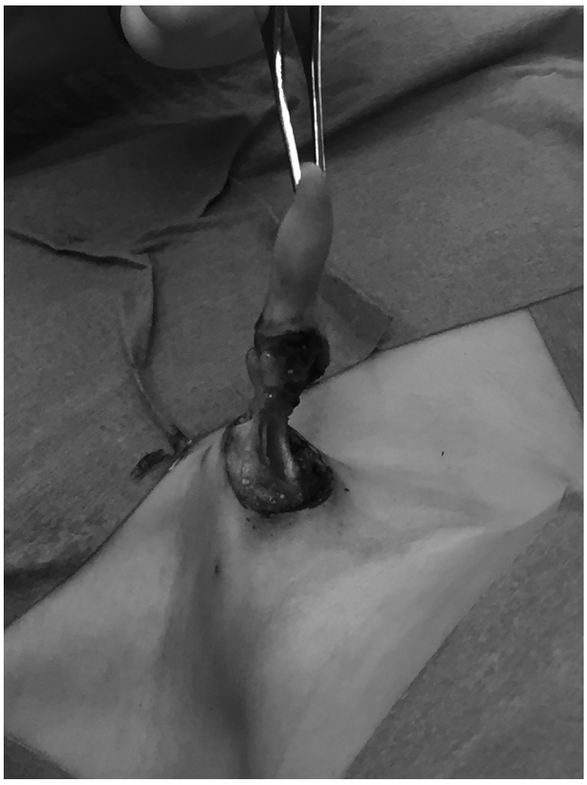

Figure 2
Adherence of the CCBR to the fascia of the sternocleidomastoid muscle.

Figure 3
The polypoid skin lesion after surgical excision (9 ´ 7 ´ 12 mm).

CCBRs, which suggested that there might be a heritable factor associated with CCBRs.[2,8]

Case Report

We describe an infant born in our hospital (AZ Delta, Roeselare, Belgium) with a skin lesion on the lower third of the left side of the neck, anterior to the sternocleidomastoid muscle. The lesion had an elastic consistency. There was no discharge, and the overlying skin appeared normal. The lesion was mobile relative to the underlying structures. A general examination showed normal findings. Cardiac and abdominal ultrasonography showed no anomalies. There were no similar presentations in any of the patient's family members. At 1 year of age, a complete surgical excision was performed under general anaesthesia. The lesion had extended into the neck and was connected to the fascia at the anterior part of sternocleidomastoid muscle. Macroscopic examination showed a polypoid skin lesion, measuring 9 ´ 7 ´ 12 mm. The specimen was fixed in formalin and stained with haematoxylin and eosin (H&E). A histopathological examination revealed that the lesion consisted of normal skin, subcutaneous tissue, and a large piece of hyaline cartilage in the centre. The surgical excision was complete.

Discussion

The pathogenesis of CCBRs is a controversial issue. There are two theories regarding the embryonic origin of CCBRs. One theory states that they arise from ectopic auricular tissue (and thus, from the first or second branchial arch). This theory is supported, in some cases, by the presence of elastic cartilage. The other theory states that they have a cervical origin (thus, from the second or lower branchial arches). This theory is supported, in other cases, by the presence of hyaline cartilage.

In the fourth week of embryogenesis, the neural crest cells migrate into the future head and neck to form 6 branchial arches. In the fifth week, the auricle develops around the first and second arches. Three hillocks develop from the first branchial arch, and three other caudal hillocks develop from the second branchial arch, which give rise to the

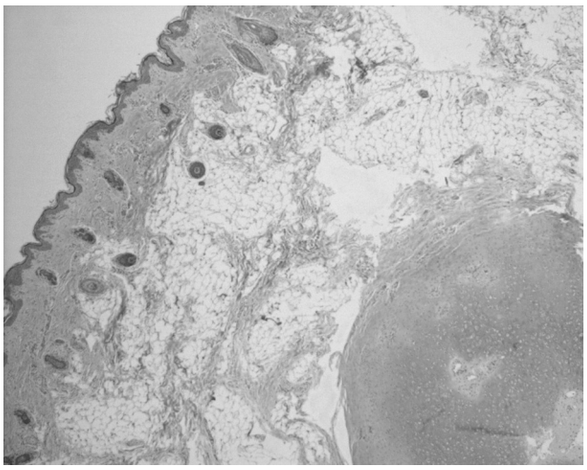

Figure 4
Histopathology shows normal skin, subcutaneous tissue, and a hyaline cartilage core, which suggests a cervical origin (of the second or lower branchial arches).

auricle during the sixth week of embryogenesis. During the seventh week, the auricular hillocks start to enlarge and differentiate. The auricle starts to translocate from its initial ventral position, on the lower side of the lateral neck, to its ultimate lateral cranial destination. During this migration, it follows the anterior border of the sternocleidomastoid muscle. Therefore, it is possible that CCBRs might be remnants of the auricular hillocks that are left behind during this migratory process. This hypothesis is compatible with their relatively superficial position and their location relative to the sternocleidomastoid muscle.

However, this explanation is unlikely to be correct for CCBRs of the ventral midline. The epiglottis and some laryngeal cartilages are of branchial origin and are normally composed of elastic cartilage. The epiglottis first distinctly appears as a prominence in the midventral area of the pharynx, between the third and fourth branchial arches. Embryologically, the epiglottis is a strictly ventral midline structure. Therefore, it is possible that a displaced portion of the epiglottis might be the source of CCBRs that occur in the ventral midline area. It remains unclear whether CCBRs originate from auricular cartilage or whether they are a remnant of branchial cartilage.[7]

Regardless of the exact origin of CCBRs, the treatment of choice is always surgical excision.[6] Treatment is recommended before school age, for social reasons, and for histopathological verification; however, treatment can be postponed to a later age for safety reasons. Neither recurrence nor malignant transformation has been reported.[3]

Conclusion

Chondrocutaneous branchial remnants are rare, benign, congenital lesions, typically found in the middle or lower third of the neck, over the sternocleidomastoid muscle. They consist of normal skin with a cartilage core. No malignant transformation has been reported. Treatment is simple surgical excision. It is very important to provide a correct diagnosis of CCBRs, because they are often a visible marker of serious underlying congenital anomalies. A thorough physical examination is recommended, with ultrasonography of the abdomen and the heart.

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