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Cervical chondrocutaneous remnant: a case report

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Cervical chondrocutaneous branchial remnants are very rare congenital lesions of the lateral neck; thus, our knowledge of this condition derives almost entirely from occasional case reports in the literature. They are thought to originate from the branchial arches and, therefore, can be found anywhere on the pathway along which those branchial arches migrate during embryogenesis. We report the case of a 5-year-old girl presenting with a cervical chondrocutaneous branchial remnant on the right lateral neck that had existed since birth, with no other anomalies.

Keywords: Branchial region / Case reports / Congenital abnormalities / Ear cartilage / Embryonic development / Review literature as topic

Case Report

INTRODUCTION

Cervical chondrocutaneous branchial remnants are rare benign congenital masses. As they are widely believed to be derived from the second and third branchial arches, they can be seen in various locations on the cervical area. In 1858, Birkett first described them as "imperfectly developed auricles on the sides of the neck" [1]. Since then, 117 cases have been reported but described with many different names [2]. Cervical chondrocutaneous branchial remnants were first formally defined by Atlan et al., in 1997 [3]. The same author believed that the remnants were "left behind" during the embryologic migration of auricular hillocks from the second branchial arch [2,3]. This condition is sometimes described using the term "accessory tragus," which refers to a congenital mass most commonly located anterior to the auricle. However, in contrast to what the term accessory tragus implies, these remnants can occur at any site along the embryonic migration track in the facial and cervical area. This report presents a case of cervical chondrocutaneous branchial remnant, which appears similar to an accessory tragus. Indeed, it is likely to be misdiagnosed at the first encounter due to its rarity.

CASE REPORT

A 5-year-old girl was brought to the outpatient center of the plastic surgery department for a congenital cervical mass. A complete physical examination of the head and neck was performed. The girl had a prominent, sharp mass on the right lower neck, which was observed at the anterior border of the sternocleidomastoid muscle (Fig. 1). The mass was hard and palpable. An ultrasound scan revealed a protruding soft tissue mass with long tracts to a deep portion (muscle layer) in the anterior neck, suggesting the possibility of a leiomyoma or fibroma (Fig. 2). The mass was excised with the patient under general anesthesia. Initially, the mass was presumed to be thyroglossal remnants or another skin lesion, such as fibroma, and it was also thought to be part of the branchial anomalies. During surgery, the skin incision was performed cautiously due to the potential risk of damaging vessels and nerves near the trachea, as the ultrasound scan determined a small tract beneath the lesion. The skin and subcutaneous tissues of the mass were dissected from the neck, revealing a cartilage fragment. The

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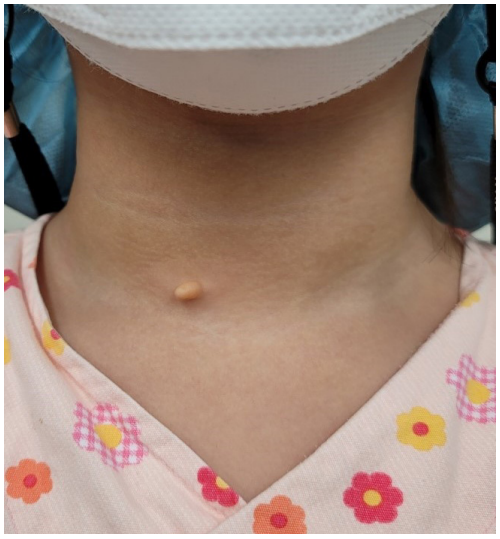

Fig. 1. A prominent sharp mass on the right lower neck at the anterior border of the sternocleidomastoid muscle.

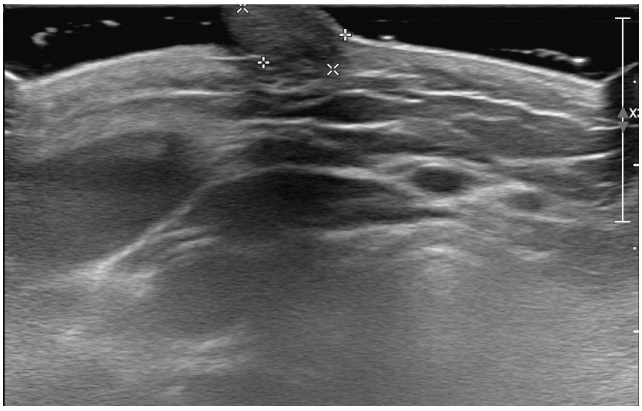

Fig. 2. Preoperative ultrasound scan. A protruding soft tissue mass with long tracts to a deep portion (muscle layer) is seen in the anterior neck, suggesting the possibility of leiomyoma, or fibroma.

root of the cartilage, which was deeply seated, was removed entirely from the fascia of the sternocleidomastoid muscle with a smooth cut. No remaining cartilage tissue was found on palpation after the mass excision (Fig. 3). The possibility of the mass on the neck being part of a thyroglossal duct cyst or involved with a branchial cyst was ruled out due to the following features: underlying cartilage base covered with normal skin; no signs of cysts.

An analysis of the gross features of the mass based on the surgical specimen showed that it had an ovoid shape and was composed of pediculate-patterned soft tissue with a cartilaginous base, resembling the features of an accessory tragus (Fig. 4). As soon as the 1.5-cm-long mass was removed, it was sent to the pathology laboratory for biopsy. Primary closure was carried out immediately using subcutaneous and skin sutures (Fig. 5).

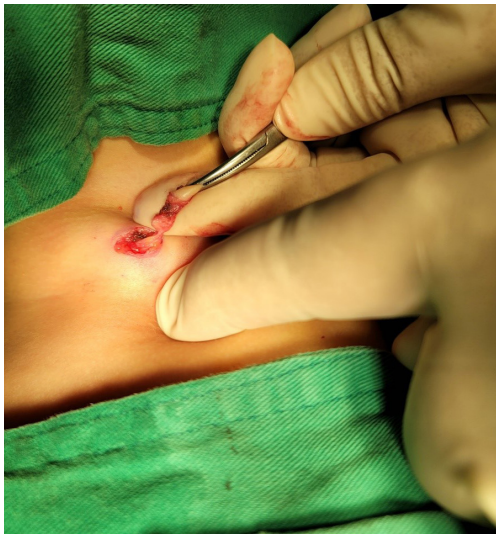

Fig. 3. No remaining tissue of the cartilage was found on palpation after excision of the mass.

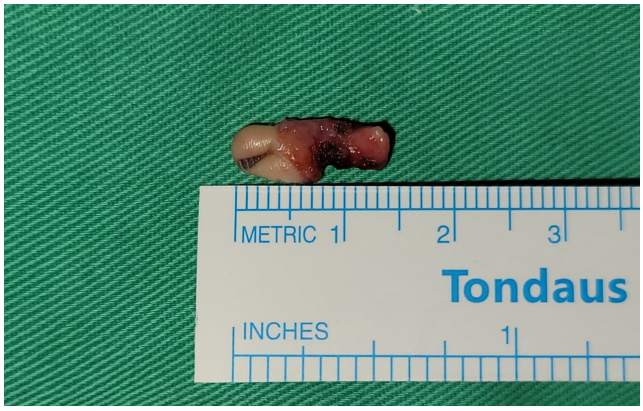

Fig. 4. A 1.5-cm-long mass (the surgical specimen), shown to be ovoid and sessile-patterned soft tissue with a cartilaginous base.

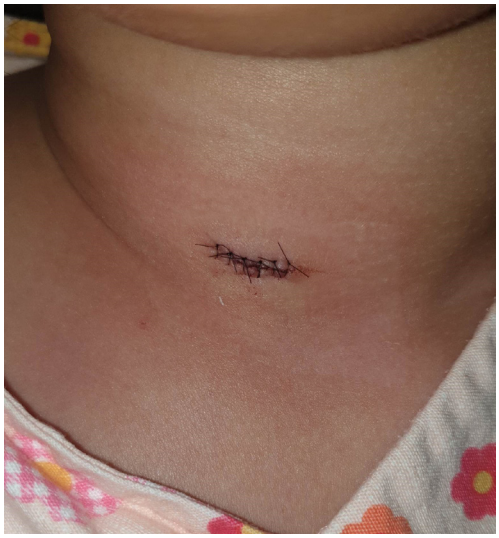

Fig. 5. Primary closure with subcutaneous and skin sutures.

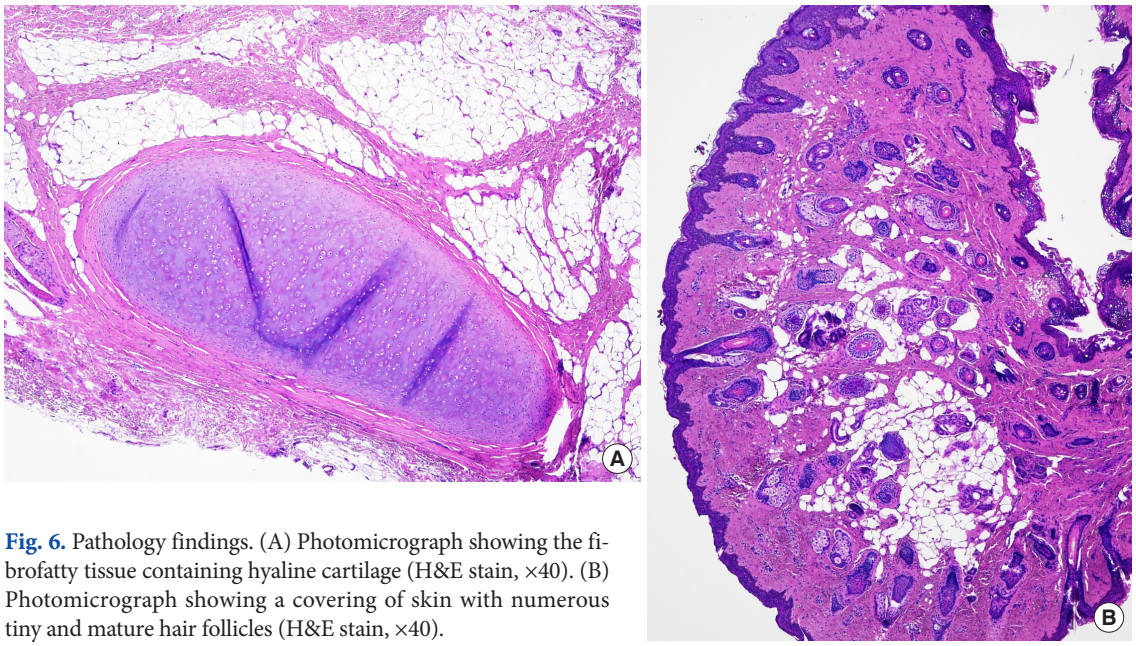

Fig. 6. Pathology findings. (A) Photomicrograph showing the fibrofatty tissue containing hyaline cartilage (H&E stain, ×40). (B) Photomicrograph showing a covering of skin with numerous tiny and mature hair follicles (H&E stain, ×40).

A photomicrograph of the biopsy showed that the underlying component of the mass comprised fibrofatty tissue containing hyaline cartilage, while the protruding portion consisted of a covering of skin with numerous tiny and mature hair follicles (Fig. 6). According to the pathology report, the possibility of fibroma or leiomyoma was ruled out [4]. Furthermore, the patient did not have any other anomalies according to her medical history and clinical manifestations.

DISCUSSION

A cervical chondrocutaneous branchial remnant consists of normal skin with a cartilage core. In this case, no connection to underlying deep structures was seen, but adherence to the fascia of the sternocleidomastoid muscle was observed intraoperatively. The clinical, ultrasonographic, and histological characteristics of cervical chondrocutaneous branchial remnants are well-known. However, their embryological origins are still debated. Therefore, the embryogenesis of the branchial apparatus is discussed below in relation to the specific features of this case.

In the fourth week of gestation, the neural crest cells migrate into the future head and neck, forming six branchial arches [5]. These are lined externally by ectoderm and internally by endoderm, with mesoderm in between [6]. In the fifth week, the primitive auricle develops around the first and second arch, and six auricular hillocks appear in the sixth week. During the seventh week, the six pre-cartilaginous hillocks on the first and second branchial arches start to enlarge and differentiate. This occurs during the migration from a ventral position on the lower side of the lateral neck to the lateral cranium. Those who believe cervical chondrocutaneous branchial remnants are of auricular origin suggest that any interruption in this migratory process can result in a remnant along the pathway [7]. Particularly for our patient, it was the remnant anterior to the sternocleidomastoid muscle in the lower third of the neck.

Meanwhile, the second and third branchial arches form the hyoid bone and adjacent neck areas: the dorsal parts of the first and second branchial arches give rise to the auricle, middle ear, inner ear, and facial nerve. The ventral parts of these structures develop into the mandible, maxilla, and the majority of the hyoid bone, and the third branchial arch is involved in forming the lower body of the hyoid bone. The third and fourth branchial clefts are mainly involved with forming the pharynx below the hyoid bone [6].

As the mandible and maxilla develop, the auricular areas are displaced dorsolaterally. The ectoderm overlying the dorsal parts of the first and second branchial arches induces the underlying mesenchyme toward chondrocyte differentiation and proliferation with collagen deposition [8]. The branchial clefts and pouches are gradually obliterated by the mesenchyme to form the mature head and neck structures. Branchial anomalies result from incomplete obliteration of the clefts and pouches during chondrogenic differentiation [6].

One suggests that cervical lesions with a cartilage core are ectopic auricular tissues. An accessory tragus also results from the failed fusion of the hillocks of the first and second branchial arches, and indeed, it occurs anywhere on the migration track of the head and neck [9-11]. Therefore, cervical chondrocuta-

neous branchial remnants are often confused with cervical accessory tragus [12,13]. Of note, they both have the same histological features. Even in our case, the lesion was once considered a cervical accessory tragus.

The other theory suggests that cervical chondrocutaneous branchial remnants originate from the second and lower branchial arches, which contribute to the formation of most cervical structures [14]. Even further, those who claim cervical chondrocutaneous branchial remnants are of a branchial origin hypothesize that they arise from the primordial laryngeal remnants of the second and third branchial arches [2].

The core of a cervical chondrocutaneous branchial remnant is either elastic cartilage or hyaline cartilage. We also agree with Atlan's suggestion for the origin of the cervical chondrocutaneous branchial remnant, but this still requires further investigation [3,6,13]. The histologic type of the cartilage is believed to be a clue to the embryological origin of cervical chondrocutaneous branchial remnants [2,3]. Since only the ear, epiglottis, corniculate cartilage, and part of the arytenoid cartilage are elastic in nature, the presence of elastic cartilage would suggest an auricular (first or second branchial arch) source [15]. In contrast, the presence of hyaline cartilage in the mass specimen of our patient excludes an auricular origin and suggests a cervical origin from the second or lower branchial arches [14]. Therefore, it is postulated that our patient's cervical chondrocutaneous branchial remnant might have resulted from incomplete obliteration of the second or lower branchial arches. This interpretation is also supported by the location of the remnant on the lower neck [2].

Nevertheless, the origin of chondrocutaneous branchial remnants on the neck remains a matter of debate since some authors have reported cervical masses with an underlying core of elastic cartilage. For instance, Clarke [16] suggested that cervical chondrocutaneous branchial remnants represent ectopic external ear cartilage. This proposal is based on the finding that they possess underlying elastic cartilage and numerous hair follicles containing vellus hair, as does normal auricular cartilage [15]. Further analysis of more cases of cervical chondrocutaneous branchial remnants is needed to understand more fully the correlation between the histologic type of cartilage and its origin.

Atlan [3] reported previously on the high complication rates of cervical chondrocutaneous branchial remnant, and also, it may be a marker of other malformation and abnormalities. However, in the study of Ishigaki et al. [13], the complication rate was lower than that reported in the Atlan. Small cervical chondrocutaneous branchial remnants might not be apparent; therefore, they may not be noticed and could be misdiagnosed. Even worse, most of them with no associated abnormalities can be dismissed. Since most studies were gathered through retrospective research, no standard method was available to investigate systemic complications and malformations [13]. We cautiously speculate that this may be one of the reasons for the discrepancy in the literature: the debate on the type of cartilage based on its origin and the inconsistent complication rates.

In our patient, the cervical chondrocutaneous branchial remnant could have been the sole manifestation of a congenital disorder associated with the development and differentiation of the branchial apparatus; however, it may be a sign related to genetic syndromes [17,18]. Some studies, such as those by Pham Dang et al. and Klockars et al., reported cases of familial presentations, indicating that this anomaly might have a hereditary tendency [6,19,20]. Thus, it is imperative to closely monitor patients for other abnormalities through a systemic examination [17,18]. A diagnosis should only be made after associated syndromes are ruled out [17]. It should also be kept in mind that the surgical removal of a cervical chondrocutaneous branchial remnant must include the entire underlying cartilage for a good prognosis [20].

NOTES

Conflict of interest

No potential conflict of interest relevant to this article was reported.

Ethical approval

The study was approved by the Institutional Review Board of Dongkang Medical Center (IRB No. DKMC 2022-06-03).

Patient consent

The patient's parents provided written informed consent for the publication and use of her images.

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Young Kim. Resources: Dae Hwan Park, Sun Young Kim. Software: Sun Young Kim. Supervision: Dae Hwan Park, Bong Soo Baik, Wan Suk Yang. Validation: Dae Hwan Park, Bong Soo Baik, Wan Suk Yang, Sun Young Kim.

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