Unilateral Cervical Chondrocutaneous Branchial Remnants: A Case Report

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ABSTRACT:

Cervical chondrocutaneous branchial remnants (CCBRs) represent uncommon benign congenital anomalies located in the cervical region, resulting from developmental irregularities associated with the embryonic branchial arches. Typically presenting as asymptomatic, flesh-toned nodules located on the inferior aspect of the cervical region, anterior to the sternocleidomastoid muscle, these lesions are generally benign. Nonetheless, it is essential to assess for potential associated anomalies, particularly cardiovascular defects, in affected infants. Elective complete surgical excision is advised to address both cosmetic and psychological concerns. This report describes a case of an adult with pathologically confirmed CCBRs who exhibited asymptomatic, flesh-toned nodules on the left lateral aspect of her neck since the time of her birth.

Key words: Cervical chondrocutaneous branchial remnants, Congenital neck mass, Cervical nodule, Branchial anomalies

Introduction

Congenital cervical anomalies constitute a critical factor in the differential diagnosis of congenital cervical masses or nodules. These anomalies are infrequent and result from deviations in embryological development, specifically the persistence of structures that are otherwise expected to resolve with maturation. Branchial anomalies, often arising from the second branchial cleft, may be erroneously identified as other prevalent congenital cervical anomalies, including branchial or thyroglossal duct cysts. The partial failure of branchial cleft obliteration can result in the development of cystic formations, sinuses, fistulae, or residual cartilaginous structures. Cervical chondrocutaneous branchial remnants (CCBRs), resulting from developmental aberrations of the second brachial arch, represent the second most common congenital abnormalities observed in the pediatric head and neck region. These anomalies are generally located along the anterior margin of the sternocleidomastoid muscle, with a particular prevalence in the middle and lower thirds of the cervical region.

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Case report

A 24-year-old Thai female presented with a painless, flesh-toned nodule on the left lateral aspect of her cervical region since the time of birth. She had no notable medical history, and there was an absence of any familial occurrences of a comparable condition. Physical examination identified a unilateral, well-defined, flesh-toned nodule size 1.5x1.5 cm on the left side of the neck, without evidence of sinus drainage or fistula (Figure 1). No cardiac anomalies or other abnormalities were detected. An excisional biopsy of the left neck lesion was performed, revealing an exophytic, polypoid lesion with unremarkable epidermal coverage and a central cartilaginous core. The lesion contained remnants of salivary glands and ducts (Figure 2). The pathological diagnosis confirmed CCBRs.

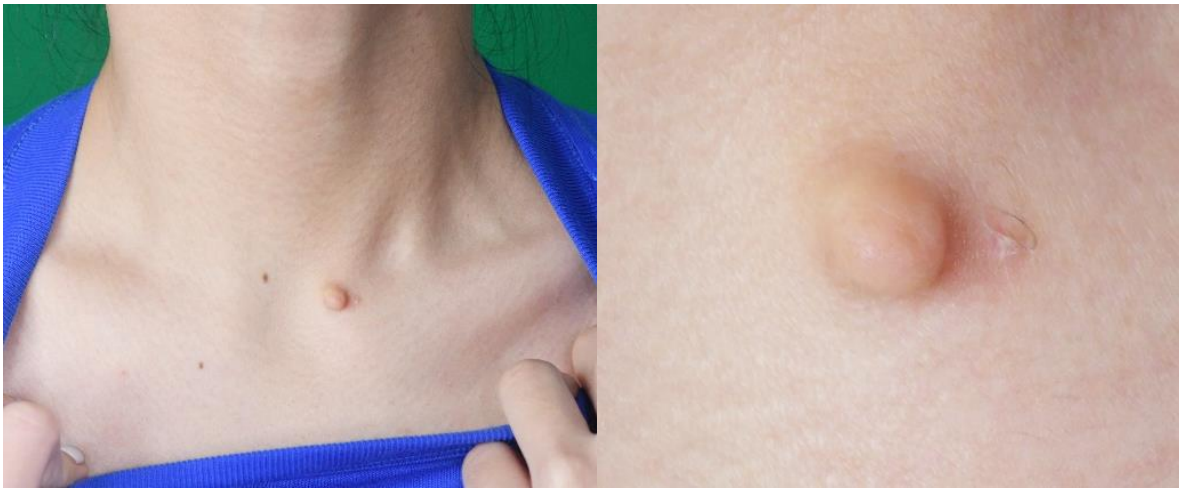

Figure 1 A unilateral, well-defined, flesh-toned nodule size 1.5x1.5 cm on the left side of the neck

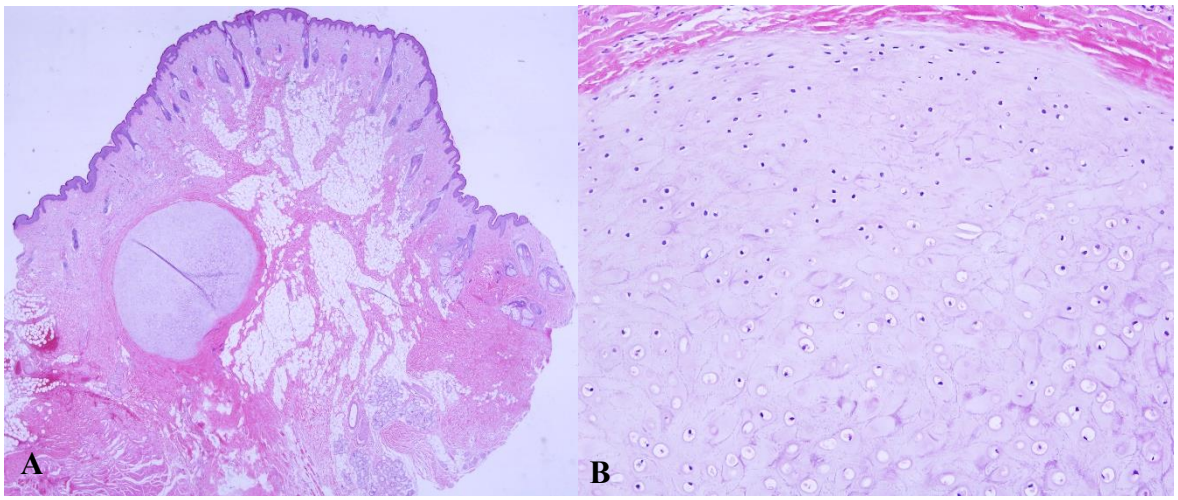

Figure 2 H&E (original magnification, X40 in A and X200 in B) shows an exophytic, polypoid lesion with unremarkable epidermal coverage and a central cartilaginous core. The lesion contained remnants of salivary glands and ducts

Discussion

CCBRs are atypical congenital benign neck masses resulting from developmental anomalies of the embryological branchial arches, specifically during the fourth week of gestation[1]. According to Woo et al., the 10-year incidence rate of branchial cleft anomalies is approximately 0.4% among 1,096 Korean patients[2]. During the fourth week of embryonic development, neural crest cells migrate into the nascent head and neck region, giving rise to six branchial arches. These arches are externally covered by ectoderm, internally lined by endoderm, with mesoderm situated in between. By the fifth week, the rudimentary auricle initiates its formation around the first and second arches, culminating in the emergence of six auricular hillocks by the sixth week. By the seventh week, these pre-cartilaginous hillocks undergo enlargement and differentiation as they migrate from the lower lateral neck to the lateral aspect of the cranium, resulting in fusion with the underlying integument and the creation of the cervical sinus. The persistence of the branchial cleft or pouch may lead to cervical anomalies along the anterior edge of the sternocleidomastoid muscle, stretching from the tragus to the clavicle. Such anomalies can manifest as cysts, sinuses, fistulae, or cartilaginous remnants, attributed to the failure of fusion and the disappearance of ectoderm[3].

The precise etiology of CCBRs remains unclear; however, the predominant theory attributes their formation to incomplete obliteration of the branchial apparatus during embryogenesis. Atlan et al. classify CCBRs into two subtypes based on their location and cartilage type: lesions containing elastic cartilage are believed to originate from the auricle, while those with hyaline cartilage situated below the hyoid bone are thought to derive from the second branchial arch[4].

Typically, CCBRs exhibit a male predominance and are characterized by painless, flesh-toned nodules with minimal or no growth. These nodules are usually approximately 15 mm in diameter, are positioned anteriorly to the sternocleidomastoid muscle, and may be situated in the anterior, middle, or posterior regions in relation to the sternocleidomastoid muscle, often observed in the lower third of the cervical region. Some cases may present bilaterally or originate from the midline of the neck[2,3,5,7]. CCBRs are generally not connected to deeper underlying structures, except for adherence to the lateral aspect of the sternocleidomastoid muscle. There is no documented association with underlying sinuses or cysts[5,8]. Differential diagnoses for CCBRs include thyroglossal duct cysts, thymic cysts, branchial cleft cysts, as well as congenital midline hamartomas.

Histological examination of CCBRs typically reveals hyaline or elastic cartilage, cutaneous appendages with hair follicles, and fibroadipose connective tissue covered by keratinizing squamous epithelium. This histological profile suggests a cervical origin associated with the second or third branchial arches[6,9]. The histological findings in this case are consistent with this pattern. Among reported cases, 76% of patients have presented with either single or multiple associated anomalies, ranging from minor conditions such as serous otitis media to more severe malformations including cardiac anomalies[5,6]. The most frequently reported associated anomalies are cardiovascular and auditory, followed by head and neck, and genitourinary anomalies. Notably, cardiac ventricular septal defects are the most common, followed by atrial septal defects and serous otitis media. There have also been reports of subependymal cysts and thyroid hemiagenesis linked to CCBRs[4,6]. Pediatric patients with CCBRs may require further diagnostic evaluation, such as abdominal ultrasound and echocardiography. In contrast, adult patients often do not present with associated anomalies, which may make screening for such anomalies unnecessary in

this population[2,7,8]. However, the majority of the literature supports screening for associated anomalies in all patients diagnosed with CCBRs[2,5,8,10]. Despite this, there is currently no consensus or large-scale studies providing clear guidelines for such investigations, and additional research is needed.

The utilization of diagnostic imaging modalities such as ultrasound or contrast-enhanced computed tomography is crucial for assessing various aspects of CCBRs, including the depth of the mass, invasiveness, cystic-like features, and fistula formation. Imaging techniques are also beneficial for evaluating additional branchial remnant anomalies. Prenatal diagnosis of CCBRs has been documented via routine transvaginal ultrasonography[1]. Currently, there is no established standard guideline for managing CCBRs. The prevailing recommendation is elective complete surgical excision in pediatric patients to improve both aesthetic and psychological effects, as well as to confirm the histopathological diagnosis[2,3,5,7]. Notably, literature reviews indicate no instances of recurrence or malignant transformation following surgical excision.

Conclusion

CCBRs represent rare benign congenital anomalies located in the cervical region, resulting from developmental anomalies of the embryonic branchial arches. Typically presenting as asymptomatic nodules since birth, they may be associated with other congenital anomalies. In the presented case, the patient underwent an excisional biopsy; however, the lesion was not fully excised, and the patient was lost to follow-up for further surgical intervention.

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