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Photo Vignette

Congenital cartilaginous rest of the neck in a boy

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Abstract

Congenital cartilaginous rest of the neck is a rarely encountered entity that requires surgical excision. In this case report, we describe a 12-year-old boy with asymptomatic congenital cartilaginous rest of the neck. We also discuss the associated congenital malformations that dermatologists must be aware of when caring for patients with this disease.

Introduction

Congenital cartilaginous rest of the neck (CCRN) is a rare developmental abnormality that appears at birth as a firm cervical papule or nodule. CCRN has been referred to as a wattle, cervical tab, cervical auricle, Meckel's cartilaginous remnant, and elastic cartilage choristoma of the neck [1-3]. Historically, congenital anomalies of the head and neck remained unexplained until Ranke first described the embryologic origins of the branchial system in 1825. Since that time, CCRN has been characterized as a branchial arch remnant that is considered to be a cervical variant of accessory tragus. We report a recent case of a 12-year-old male with CCRN.

Case synopsis

A 12-year-old boy with no known congenital abnormalities was referred by his primary care physician for evaluation of an asymptomatic enlarging "skin tag" on the anterior neck. The lesion was noted when the patient presented to another physician for a sports physical. The patient's pediatrician had noticed the lesion at birth and ultrasound of the lesion failed to reveal a fistula or cyst. Physical exam revealed an exophytic, firm, rubbery, non-cystic 1.0 x 0.8 centimeter papule overlying the inferior portion of the anterior triangle of the neck at the insertion of the sternocleidomastoid (Figure 1).



Figure 1. rubbery non-cystic papule over lower anterior triangle of neck

Differential diagnosis included CCRN, thyroglossal duct cyst, cystic hygroma, branchial sinus, acrochordon, and benign papilloma. The lesion was mobile, painless, and not fixed to any underlying structure. Shave biopsy was performed and histologic examination confirmed ectopic cartilaginous tissue under the epidermis with normal adnexal structures (Figure 2,3). These findings confirmed the diagnosis of CCRN.

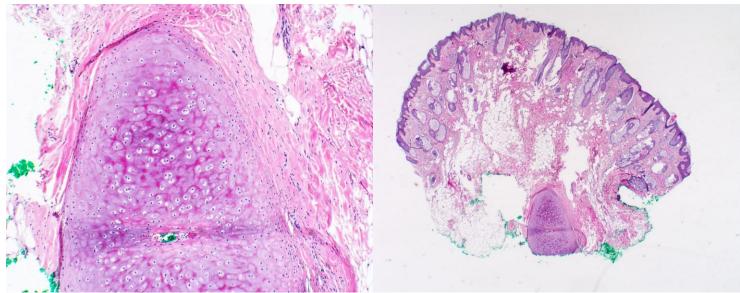


Figure 2 (a and b). Ectopic cartilaginous tissue with normal adnexal structures

Discussion

CCRN is a congenital malformation resulting from abnormal development of the branchial apparatus. The incidence of both unilateral and bilateral lesions is estimated to be 9-10:100,000 and is twice is common in males [4]. Clinically, CCRN presents as a firm subcutaneous nodule or papule overlying the anterior neck. CCRN is evident at birth and is generally stable in size but has been reported to grow up to 20 cm in diameter. It has also been associated with other malformations of the first and second branchial arches. These malformations include microtia, stenosis of the external ear canal, and branchiogenic fistulae. In addition Goldenhar, Treacher-Collins, Townes-Brocks, Wolf-Hischhorn and Delleman syndromes can be associated [4-9]. Diagnosis is confirmed by histological examination.

CCRN has a distinctive histologic appearance with discrete lobules of hyaline cartilage in collagenous tissue. The skin that covers the lesion often contains multiple vellous hair follicles, eccrine glands, and pilosebaceous units. Excision is usually the preferred form of treatment for diagnostic and cosmetic purposes. Shave excision is not recommended because it may leave the cartilage exposed and cause subsequent complications [7-9]. The lesion may be attached to underlying fascia by a fibrous band but can be easily excised since it is neither fistulous nor cystic [7-9]. Lesions that do not have fistulous tracts or connections with deeper structures can be carefully excised at the superficial musculature.

In conclusion, CCRN is a congenital malformation resulting from abnormal development of the branchial apparatus. Children presenting with CCRN should undergo a thorough evaluation for any developmental anomalies. An ultrasound of the neck is indicated before surgical intervention to determine whether fistulae or sinuses are present.

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