Congenital Cartilaginous Rest Of The Neck: Case Report And Review Of The Literature

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Citation

Abstract
Objective: To help in the recognition of a rare developmental abnormality, warn of possible associated developmental abnormalities, and to provide an overview of the embryology.

Case Report: A 3-year-old boy presented with a history of a neck lesion evident since birth. The child was asymptomatic and the parent's only concern was its cosmetic appearance (Figure 1). The child was otherwise fit and well. He had been born at term without any complications and developmental milestones were within normal limits. There was no family history of similar lesions. Excisional biopsy was performed to ascertain the nature of the lesion.

Conclusion: Congenital cartilaginous rests of the neck (CCRN) are rare developmental abnormalities that appear as firm nodules in the lower anterior neck near the insertions of the sternocleidomastoid muscle. Treatment is usually excision for cosmesis. CCRN can be a marker for associated anomalies and patients should be considered for further investigations.

INTRODUCTION
Congenital cartilaginous rests of the neck (CCRN) is a rare developmental abnormality that appears as a firm nodule in the lower anterior neck near the insertions of the sternocleidomastoid muscle. CCRN has been variously described as wattle, cervicle tab, cervical auricle, Meckel's cartilaginous remnant and elastic cartilage choristoma of the neck to name a few (1,2,3,5,9).

The term ‘wattle’ means a fleshy appendage of the neck and is more commonly applied to the dewlaps of birds and skin tags of sheep, pigs and Egyptian goats. They were also a distinguishing feature of satyrs and fauns of Graeco-Roman mythology (5,8). The “Wattle sign” can have two interpretations. The first is a description of an enlarging facial mass when the head is in a dependent position. This is pathognomonic of a vascular malformation or haemangioma (10). The second describes the “jowls” of an ageing face.

We report on a recent case, and review the embryology and literature on the subject.

CASE REPORT
A 3-year-old boy presented with a history of a neck lesion evident since birth. The child was asymptomatic and the parent's only concern was its cosmetic appearance (Figure 1). The child was otherwise fit and well. He had been born at term without any complications and developmental milestones were within normal limits. There was no family history of similar lesions.

On examination there was a small but firm, non-cystic lesion overlying the posterior third of the left sternocleidomastoid muscle at the inferior and middle third junction. It was
painless, mobile and was neither fixed to deeper structures nor the skin. There were no associated skin changes or sinus tract formation. As a specific diagnosis could not be made on history and clinical examination, surgical removal was recommended for histological analysis.

The mass was superficial to platysma and not adherant to deeper structures (Figure 2). Histology revealed features consistent with a benign cartilaginous rest that could represent a variant of an accessory tragus (Figure 3).

DISCUSSION

Congenital cartilaginous rest of the neck (CCRN) is a rare anomaly with a reported male to female ratio of 2:1 (1). These lesions are always present at birth, are commonly unilateral and may be multiple (2,7,9). The prevalence as an isolated physical finding has been estimated as 1.7:1000 for unilateral lesions. Bilateral lesions are present in approximately 6% of cases, giving it a prevalence of 9-10:100,000 (6).

In a four-week embryo, the neural crest cells migrate into the future head and neck forming six branchial arches, five being rudimentary. Each arch has a mesenchymal core with an ectodermal external cover, and an endodermal internal cover. In the fifth week of embryological development, the auricle develops around the first and second branchial arches, with three hillocks appearing on each arch. Fusion of these hillocks occurs as they move dorsally along the line of the sternocleidomastoid muscle. It is along this migratory pathway that the cartilaginous rests may be deposited. By week seven the planned transformation of the auricle has occurred, and any defect in the process results in a structural anomaly. The locations of the remnants are consistent with their origins from the first or second branchial arch. In this case and in other similarly described neck lesions reported in the literature, the second arch would appear as the most likely candidate (1,2,4,5,6,7,8,9,10,11,12).

CCRN are rarely associated with congenital malformations involving the first and second branchial arch, namely Goldenhar, Treacher-Collins, Townes-Brocks, Wolf-Hischhorn and Dellemann syndromes. These syndromes are more commonly associated with multiple cartilaginous rests (7,9). One paper surprisingly showed a myriad of associated anomalies and congenital defects in patients with rests, involving all systems (2). ENT disorders known to be associated with CCRN include neurosensory deafness, otitis media, external auditory canal stenosis, tracheomalacia, cleft palate and oronasal reflux (2,7). Other anomalies are septal defects, hand and foot malformations, congenital hip dislocation, genitourinary malformations and Duane syndrome (2). A familial preponderance of CCRN has been reported in the literature (6).

CONCLUSION

Congenital cartilaginous rests is a benign condition. The lesions are usually excised for cosmetic reason. However CCRN can be a marker for associated anomalies and patients should be considered for further investigations.

References

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