Original Research Paper

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CONGENITAL MIDLINE CERVICAL CLEFT

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ABSTRACT Congenital midline cervical cleft (CMCC) is a rare congenital anomaly of the ventral neck. Numerous malformations can affect the anterior part of the neck presenting at birth as a real diagnostic challenge. This can be wrongly diagnosed as a thyroglossal duct anomaly, dermoid cyst, brachial cleft anomaly or birthmark. A prompt clinical diagnosis and surgical treatment during early infancy are essential to ensure both functional and aesthetic outcome. It represents a failure of the brachial arches to fuse in the midline and presents at birth with a ventral midline defect of the skin of the neck. Inadequate treatment may cause secondary complications such as impaired neck extension, microgenia, exostosis, torticollis or infection.

KEYWORDS :

INTRODUCTION

Congenital midline cervical cleft(CMCC) represent a failure of the brachial arches in the midline and could include mandibular spurs, microgenia, thyroglossal cyst, brachiogenic cyst , cleft of the mandible ,lower lip, tongue, upper lip and sternum . There are hardly 200 cases reported in the literature so far. It represents a variant of the cleft category of the Teisser Classification system of craniofacial defects. Clinically there are six consistant findings.

1. Midline vertical atropic skin defect
2. Lack of adnexal element within the skin defect.
3. Superior skin tag .
4. An inferior blind sinus
5. A midline subcutaneous fibrous cord and
6. an increase in the size of the defect with an increase in the patient's age. Mucous could be expressed from almost all patients from inferior sinus.

The Fibrous cord becomes more prominent with increase in age with restriction of neck movements. These findings were clearly described by Ombardane in 1949.

Case report

3 years old boy was born at term to healthy, unrelated parents with normal milestones. On examination the child was noted to have nipple like projection in the anterior neck midline 2 cm inferior to chin. The hood contained a small sinus tract which ended blindly. Inferior and continuous with the hood was 2 cm wide pink mucosal surface. Inferiorly there was a blind sinus tract which was extending towards sternum. There was neck contracture due to fibrous cord like structure running along the lesion. His routine investigations were within normal limits.

He underwent excision of the lesion with sinus tract and closure with Z plasty. The excised specimen was sent for histopathology. It showed fibro muscular tissue lined by stratified squamous epithelium.

DISCUSSION

Congenital midline cervical cleft is caused by the absence of tissue in the ventral midline. (7) The usual presentation is a cephalic skin tag with a cordlike tract running inferiorly with a caudal sinus (7,10,11,12).In this case ,the caudal hood contained sinus .The tract is usually palpable and milking the tract may result in mucoid discharge (10,11,12,13).The tract may weep from the raw red surface.(10,13).A thin desquamating epithelium usually covers the atrophic cleft.(7,11,24)The cleft skin does not grow with the surrounding skin. If the subcutaneous fibrosis cord is severe enough ,it can act as tether and limit the movement of the neck.(7,15) causing webbing a condition known as pterygium collie medianum (13, 16).In addition the fibrous band becomes more prominent with age occasionally it is associated with bony spur of mandible and rarely with bronchogenic cyst or respiratory epithelium. Congenital midline cervical cleft can be differentiated from thyroglossal duct cyst. (TDC) as CMCC has no relation with hyoid bone (17). The constant histology of CMMS, can confirm the clinical diagnosis. It is covered with stratified squamous epithelium lacking skin appendages. The skin tag contains normal epithelium. The sinus tract is usually lined by pseudostratified columnar epithelium often demonstrates seromucous salivary gland. There are 5 cases report of respiratory epithelium in the sinus tract (15) as well as two reports of tuboalveolar glands of bronchial type adjacent to the tract. (11,12).

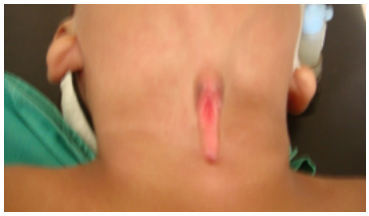
The exact pathogenesis of CMCC is still speculative ,though much have been known about embryogenesis of the area .Most of the investigators believe that the defect is a result of fusion failure of first and second brachial arches in the midline.(18,19,20,21)During normal embryology the brachial arches grow medially and then merge cephalic to caudal with the first arch closing before the second .Before the arches fuse , mesodermal tissue migrates between the arches and pushes the ectoderm outwards to fill and flatten the central furrow (13).Disruption of this process can lead to deformities .Congenital midline cervical cleft has been grouped into two entities based on the arch affected.(7,12)Decreased or deficient second arch migration results in midline cervical cleft. If the first arch is deficient the result can include hyperplasia of the mandible or more complicated presentations. There is evidence that Fibronectin and hyaluronate could play a role in the dysfunctional cell migration (7,23). The major tissue component of the brachial arches is the neural crest .However respiratory epithelium and salivary structures are endoderm ally derived. The presence of salivary gland tissue and ciliated respiratory epithelium suggest that It could be more complicated than simple failure of migration .Presence of salivary gland has been explained by abnormal development of 1st brachial arch .A delay in the merging of mandibular process could result in the deposition of ectodermal and mesodermal cells in the ventral neck, which could differentiate into skeletal muscle and salivary tissue (24,25).

Treatment involves excision of the skin tag and cleft. Surgery should be performed in early infancy. The purpose of early surgery is to prevent contractures and mandibular deformities. Many authors recommend 'Z' plasty with removal of the pathological tissue.

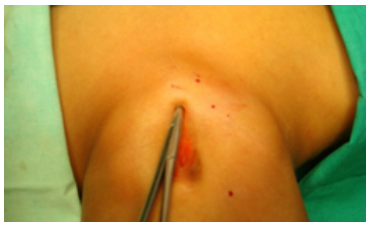
Congenital midline cervical cleft is an uncommon disorder which is evident at birth with around 200 cases reported in the literature. The first recorded case of CMMC was in 1848by Luschka ,under the description of 'Congenital Fistula of the neck'.(4) Tessier in 1973 presented classification of craniofacial cleft according to which CMMC was variant of No.30.Early reports were published by Baily in 1924,(5) and Brasky (6) in 1938.This abnormality was fully described in 1946 by Ombardane. The incidence of congenital midline cervical cleft is difficult to evaluate. In 1985 Gargan et al (7) reported 12 cases of midline cervical cleft as a part of their 30 yrs series of 672 cases of thyroglossal and other cleft sinuses. CMCC was reported to have incidence of 1.7 %. Gross in 1940 (8) called this entity as thyroglossal

fistula by the time he published on this subject again changed the term "Midline cervical cleft ". Sophie Achard (1) presented a series of 8 children where two cases had associated cervical midline cyst, three had significant micrognathia. Liana Puscas (2) presented series of eight male and two females with study of cases published all over the world including his 10 cases, total No. of cases 205. In 195 cases presented in world literature 77 were females and 58 males in 61 cases no gender was given.

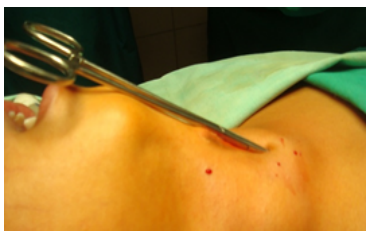
It is important to completely excise the lesion. Simply transecting the fibrous cord and performing incomplete excision leads to recurrence. Use of single or multiple Z plasties is recommended to break up the scar and improve the cosmetic and functional results. He was followed up for 5 years with no recurrence.



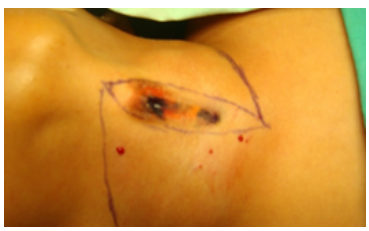
1. Congenital midline cervical cleft



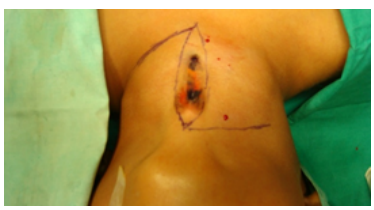
2. Blind track extending towards mandible.



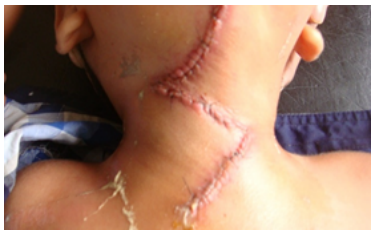
3. Blind track extending towards sternum



4. Methelene blue was injected in the track and marking of the tissue to be excised.



6. Flap raised



7. Final result

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