

Diagnosis and Treatment of Congenital Midline Cervical Cleft: 2 Case Reports

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Abstract

Congenital Midline Cervical Cleft is a rare congenital disorder of the ventral cervical area. The usual presentation is a characteristic superficial midline cervical lesion. Untreated, this anomaly may cause regional complications. The recommended treatment is surgical excision of the lesion.

We present 2 case reports of diagnosis and treatment of Congenital Midline Cervical Cleft in children including one case of prenatal diagnosis of the abnormality.

Keywords

Cervical cleft; Congenital; Surgical excision; Clinical presentation; Malformation; Surgical technique; Prenatal diagnosis

Introduction

Congenital Midline Cervical Cleft (CMCC) is a rare congenital disorder of the ventral cervical area. It is considered a midline fusion anomaly of the 1st and 2nd branchial arches that typically presents as a superficial characteristic lesion [1-4]. It is postulated that CMCC results from abnormal ventral fusion of these arches. Abnormal development of the first branchial arch, ectopically placed respiratory epithelium or mal-development of minor salivary glands are other less popular pathogenesis theories [2,10-11].

It is thought to be a sporadic disorder predominantly seen in white females [1] and is usually an isolated anomaly. However association with other malformations have been reported such as clefts of the tongue, lower lip, and mandible [1-7,10-13]. Usually, the patient is asymptomatic however there is a major cosmetic issue. The treatment is surgical excision of the lesion. Untreated CMCC may cause complications such as scarring, contractures and limitation of neck mobility.

Case Reports

We present two case reports that were diagnosed and surgically treated by our group between the years 2013-2014.

Case 1: Caucation Male

A routine prenatal Ultrasound (US) screen conducted on the 23rd gestational week (GW) revealed a midline cervical protuberance (figure 1). The lesion appeared superficial and no underlying fluid or cystic formations were visualized. No other abnormalities were present in this male fetus during the examination. A diagnosis of Midline cervical cleft was suggested.

The pregnancy was uneventful and the infant was born by normal spontaneous vaginal delivery during GW 38. An examination revealed a 1.5 cm midline skin tag in the shape of a nipple with approximately 1cm caudal diathesis of the skin. At the bottom of the projection was an opening of a sinus tract advancing subcutaneously towards the sternum in the shape of a fibrotic cord (figure 2). An additional subcutaneous cord was seen projecting from the cephalic end of the lesion towards the mandible. Upon extension pterygium colli was noted.

Post-natal US scan showed that the thyroid gland had a normal appearance and was not connected to the lesion.

The patient was otherwise healthy, had no respiratory distress or dysphagia and was gaining weight appropriately.

Surgery was performed at 16 months of age. An oval skin incision was made enclosing the lesion with inferior dissection and exposure of the inferior fibrotic cord. A transverse horizontal suprasternal skin incision was made in order to enable complete excision of the caudal cord (figure 3). Subsequently 2 delicate cephalic para-median cords were excised by proximal superior dissection (figure 4). Both incisions were closed by a vertically/horizontally linear suture.

Initial post-op examination showed good wound healing and cosmetic results. Histology showed fibroepithelial elements.

Figures

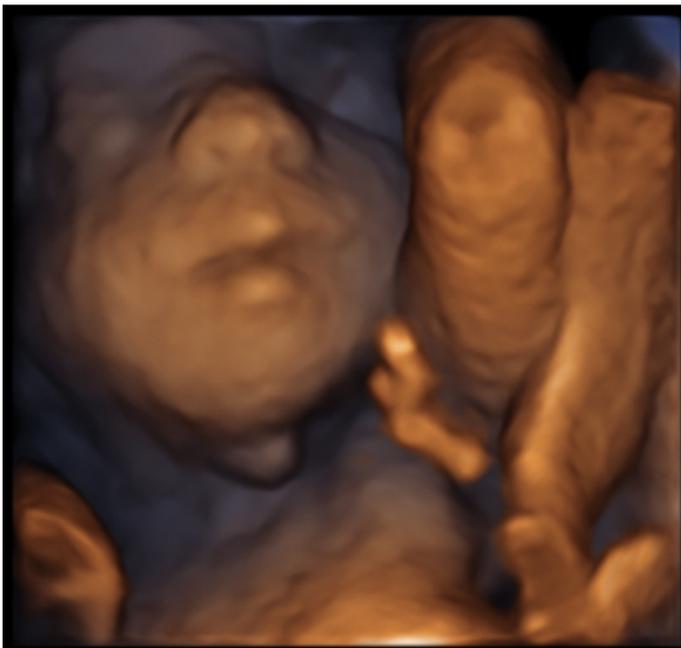


Figure 1: Routine prenatal US screen on GW 23 showing a midline cervical mass



Figure 2: The midline lesion: note the subcutaneous caudal cord (arrow)

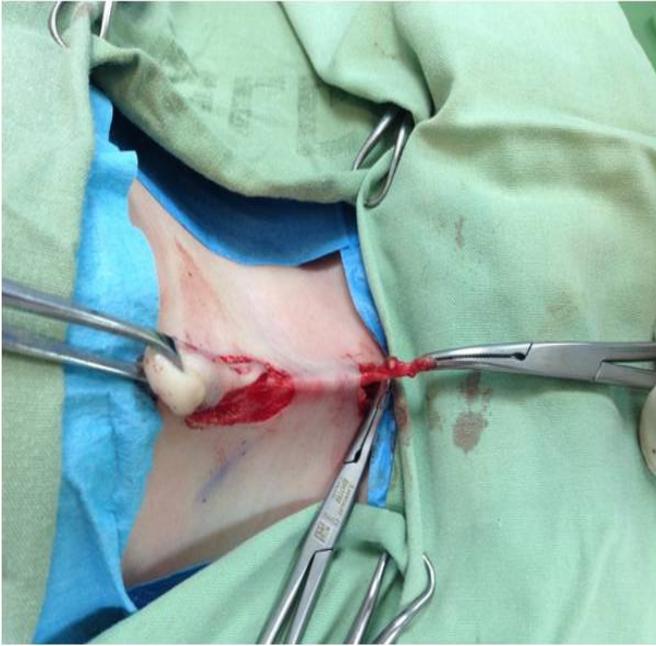


Figure 3: Excision of the lesion: an oval incision enclosing the main lesion followed by inferior dissection of the fibrotic cord and a stepladder suprasternal incision for the completion of the resection



Figure 4: The excised specimen

Case 2: Caucasian Female

A Caucasian female born at term via spontaneous vaginal delivery. A characteristic CMCC midline lesion was noted with a 1.5 cm skin tag and a caudal opening (pit) of a blind sinus projecting towards the sternum about 3 cm with overlying skin atrophy (figure 5). The patient was otherwise healthy.

Surgery was performed at 16 months of age. The lesion was completely excised (figure 6) using an enclosing oval incision and Z-plasty for closure.

Recovery was uneventful with good wound healing and cosmetic results.



Figure 5: The midline lesion with atrophic skin overlying the sinus



Figure 6: The excised specimen

Discussion

Congenital midline cervical cleft is an uncommon disorder of the ventral neck region that is clinically evident at birth. It is found predominantly in Caucasian females [1]. There have been fewer than 100 cases reported, with approximately 50 published in the English-language literature [1-2,11].

The usual clinical presentation is similar to that reported in our 2 patients: A midline cervical cleft with a nipple like projection at its cephalic end and a sinus tract opening at its caudal end which may produce mucous discharge [7-8,10-12]. Fibrotic cords projecting from the lesion's inferior end towards the sternum and from the superior end towards the mandible are usually seen [1-3]. These cords may result in neck webbing on extension known as ptergium colli medianum [12].

In severe cases or in untreated patients, CMCC may cause neck contractures and mandibular or sternal growth abnormalities. An exostosis from the midpoint of the mandible may form, resulting from persistent traction of the contracting fibrous cord underneath the cleft. CMCC can prevent full extension of the neck, resulting in micrognathia and torticollis [1-3]. This condition may predispose patients to infections, and can coexist with other clefting defects or cysts.

The usual histological features are: stratified squamous epithelium lacking skin appendages covering the cleft [2,7,11,13]. The skin tag consists of normal epidermis with occasional skeletal muscle bundles. The sinus tract is usually lined by pseudostratified columnar epithelium and often demonstrates seromucinous salivary glands. There are also several reports of respiratory epithelium in the sinus tract [2].

Careful distinction must be made to differentiate CMCC from the more common Thyroglossal duct cysts (TDC) or a branchial cleft deformity. Sinuses, fistulae, and cartilaginous rests are usually diagnosed in infancy or early childhood. Branchial cysts are usually diagnosed at a later time when they "fill up" or present with an infection. However the usual location of these lesions is the lateral neck as opposed to CMCC which is a midline lesion.

Thyroglossal duct cysts are also a midline lesion but are rarely clinically evident at birth. TDCs are not usually associated with a sinus unless an infection results in spontaneous drainage. These cysts most notably elevate when the tongue is protruded and can also be transilluminated. Histologically, unlike CMCC, TDC is often lined by thyroid epithelium. It is important to distinguish the two separate entities because treatment of TDC must involve removal of a central portion of the hyoid bone.

Treatment consists of early surgical intervention to avoid potential long-term complications, such as scarring, contractures and limitation of neck mobility. Complete surgical excision of the cleft including the underlying fibrous cord is the procedure of choice, preferably during infancy. The suggested advantages of Z-plasty are avoidance of a vertical scar and lower rate of contractures, [14-15]

In our opinion, in the usual mild presentation when the patient has lax tissue adjacent to the cleft and no anterior cervical contracture, it is possible to close the wound linearly with acceptable cosmetic results (case 1). This must be coupled with meticulous cord removal to prevent recurring contractures. In these cases an additional horizontal incision such as described in case 1 may minimize undesired cosmetic results.

In conclusion, CMCC deserves special and early attention followed by surgical treatment due to its potential to cause development of neck webbing and contractures if left untreated.

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