Case Report

ID: JCDR/2012/4671:2335

Surgery Section

Congenital Midline Cervical Cleft: A Case Report And An Embryological Review

SANGAPPA S.DODDAMANI, PRAKASH S HANDI, CHANDRASHEKHARAYYA S.H., SHASHIDHAR S SULIGAVI, SACHIN S.CHOUGULE

ABSTRACT

Congenital midline cervical cleft (CMCC) is a rare anterior neck anomaly with controversial theories / hypothesis regarding its embryogenesis. If it is not treated at an early age, it can result in complications like webbing of the neck, dental malocclusion and restricted neck movements.

We are describing here, one such case, who presented at an age of 19 years. The lesion was excised and closure was done by multiple Z- plasty, with satisfactory results. We have discussed the embryological theories / hypothesis with the review of literature.

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Key Words: Congenital Midline Cervical Cleft , Z - plasty

INTRODUCTION

Congenital midline cervical cleft is a rare midline anterior neck anomaly. Most of the cases which have been reported so far, have been detected and treated at an early age and there are very few cases who have presented at a later age with complications. This defect is common in females, with the ratio of 2:1, according to Eastlack et al [1]. The age of presentation ranges from birth to 23 years, [2]. There are reports of similar cases by Y Bajaj [3], Saha S [4], Fincher SG [5], A Mlynarek and others.

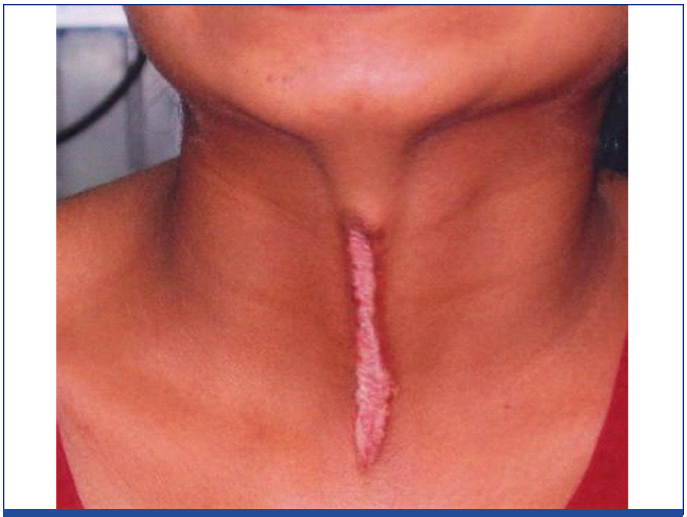
In most of the earlier reports, the authors considered surgery as the only option to cure this condition. Most of the authors have shown a preference for closure by multiple Z - plasty. The embryopathogenesis of the lesion remains unclear and several hypotheses have been proposed .

CASE REPORT

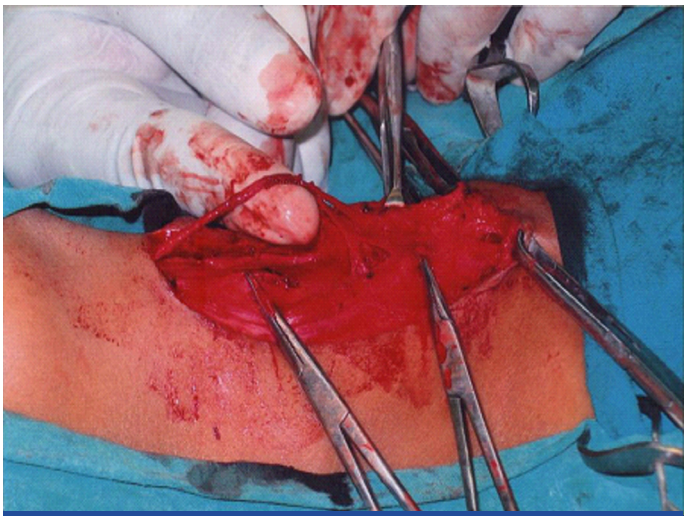
A 19 year old girl presented to the Outpatients Department with a history of a midline neck scar which had been present since birth and restricted neck movements since a few years. She was born after an uncomplicated delivery. On examination, in addition to the scar over the neck, she had micrognathia and dental malocclusion. The rest of the examination revealed nothing significantly abnormal and there was no family history of any congenital midline defects.

On detailed examination , it was found that there was a linear scar in the midline of the neck, which extended vertically between the mandible and the sternal notch [Table/Fig-1]. It was covered by atrophic skin and it measured around 7 - 8 cm in length and 4 - 6 mm in width. There was a sinus at the lower end of this lesion. A protuberance of skin was present at the superior aspect of the scar. A cord like structure was palpated underneath the skin lesion, which extended from the mental tubercle to the lower end of the lesion.

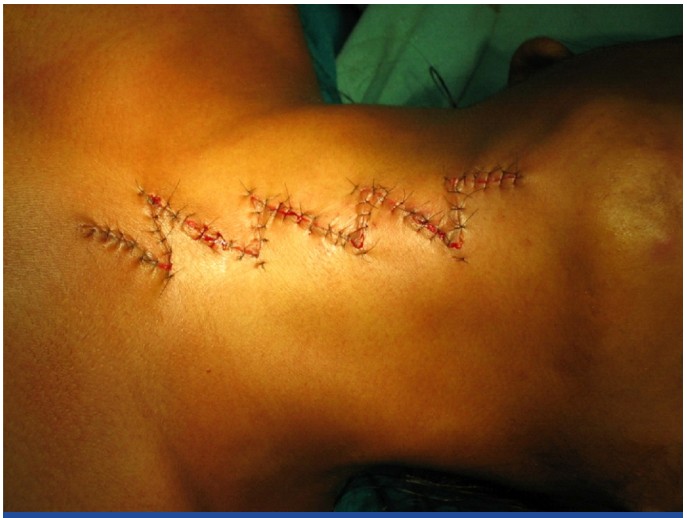
Ultrasound examination revealed the presence of a cord like structure which extended from the mental tubercle to the manubrium sterni and there was a cleft in the manubrium. The other cervical structures were normal.



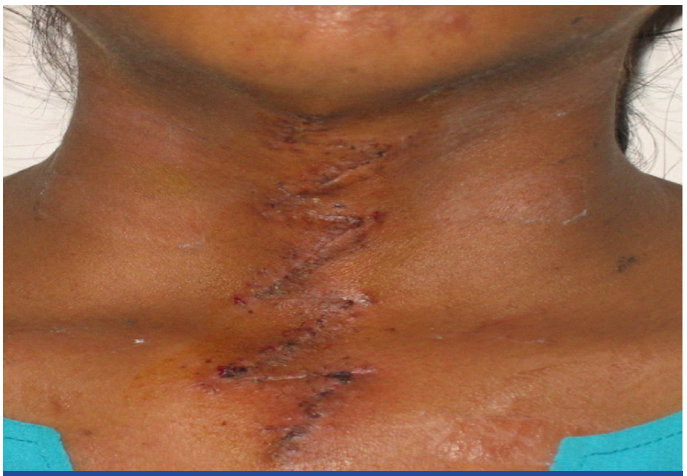
[Table/Fig-1]: Preoperative photograph



[Table/Fig-2]: Intraoperative photograph showing fibrous band



[Table/Fig-3]: Skin closure with multiple Z- plasty



[Table/Fig-4]: Postoperative Photograph

Under general anaesthesia, an incision was made, which incorporated the whole lesion within the incision margins and the lesion was dissected along with the fibrous band, which had web like strands which were extending and merging into the fasciae of the region [Table/Fig -2]. The fibrous band and the lesion were found in the subcutaneous plane mainly and they were anterior to the thyroid gland, with no extensions into the carotids or into the thyroid gland. The carotids and the thyroid were free of the fibrous band and they were normal. Then, the defect was closed by multiple Z - plasty. The post operative results as were observed on follow up at 6 months and 1 year, showed good healing and improved neck movements. [Table/Fig -3 and 4].

DISCUSSION

Congenital midline cervical cleft is a rare anomaly , but it may be overlooked or misdiagnosed. Eastlack et al. reported a female to male ratio of 2 : 1. The cases appear to be sporadic as no report has suggested a familial inheritance.

The literature has described a spectrum of clinical findings. The most consistently reported features are -

- A vertically oriented cleft of reddened tissue
- A protuberance of skin superiorly
- A blind epithelium lined sinus tract caudally
- A fibrous subcutaneous cord
- Mucoid material may be expressed from the orifice of the sinus [6].

The findings are however quite variable, occasionally involving only the midline webbing without the skin lesions. The fibrous cord may limit the neck movements and one or two bony prominences are frequently found at the base of the mandible, which are secondary to the traction of the cord.

These features tend to become more prominent with time, making an early intervention appropriate. Classically, there is some degree of retrognathism and various degrees of clefting of the lip and the mandible. The cleft may extend inferiorly to involve the sternum and superiorly the hyoid bone. The other associated abnormalities may be thyroglossal cysts and ectopic branchiogenic cysts. The less frequently associated features are ectopia cardis and midline haemangiomata [6].

Although the embryo pathogenesis of CMCC has not been fully elucidated, most of the authors regard CMCC as a part of a spectrum of branchial arch developmental anomalies. The most commonly accepted theory is impaired mesodermal fusion along the distal branchial arches in the midline [7]. An abnormal development of the first branchial arch on day 22 in the human embryo when the branchial arches are forming. The first arch is divided into the maxillary and the mandibular processes, with a horizontal cleft dividing each side in the midline. The mandibular process merges on day 26. A delay in this could result in the ectodermal cells with the underlying mesodermal cells being deposited in the ventral aspect of the neck. These would continue to differentiate and form the skeletal muscle [tongue derivative], the salivary glandular tissue and a mucosal surface, thus resulting in the CMCC defect [8].

Other theories include

[a] Rupture of a pathological adhesion between the epithelium of the cardiohepatic fold and that of the ventral part of the first branchial arch, causing tissue ischaemia again, with localized necrosis and scarring [8].

[b] The mesodermal deficiency theory states that the epithelium over the deficiency may have been intact, but pulled apart later due to the failure of the adequate mesenchymal ingrowth [9].

[c] Failure of the mesenchyme to penetrate the midline, thus causing a deficiency in its intersection with the ectoderm, which would explain the absence of the skin adnexae in the dermis [9]

[d] The pressure of the primitive heart on the branchial arches could generate adhesions and provoke a defect in the fusion between them [2,10].

[e] Vascular anomalies producing ischaemia and necrosis [11].

Gargan et al. proposed that CMCC occurs in a cluster into two major groups

[1] The midline cervical cleft and 2] Hypoplasia of the mandibular arch. If the migration along the second arch is deficient or delayed, an isolated CMCC results. The more complicated presentations such as inferior gnathoschiasis with a cleft mandible or tongue and the absence of the supporting structures in the neck are due to a deficiency of the first arch [9].

The diagnosis is made easily by simply observing the characteristic clinical findings. The treatment involves complete excision of the cleft and the sinus tract and then, Z - plasty. If needed, orthognathic procedures are undertaken, so as to achieve a correct dental occlusion. If the fibrous cord is not excised, then recurrence is common.5 An early treatment should be considered to reduce the risk of contractures, mandibular exostosis and dental malocclusion.

Congenital midline cervical cleft is a rare congenital anomaly of the neck, with many theories / hypotheses on its embryopathogenesis. The most accepted theory is impaired fusion of the distal branchial arches in the midline. An early diagnosis and surgical excision of the lesion and the underlying fibrous band with multiple Z - plasty closure, is necessary, to prevent the webbing of the neck and mandibular maldevelopment.

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AUTHOR(S):

1. Sangappa S.Doddamani
2. Prakash S Handi
3. Chandrashekharayya S.H.
4. Shashidhar S Suligavi
5. Sachin S.Chougule

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Otorhinolaryngology and Head Neck Surgery,
2. Assistant Professor, Department of Otorhinolaryngology and Head Neck Surgery,
3. Assistant Professor, Department of Otorhinolaryngology and Head Neck Surgery,
4. Assistant Professor, Department of Otorhinolaryngology and Head Neck Surgery,
5. Assistant Professor, Department of Otorhinolaryngology and Head Neck Surgery, S N Medical College, Navanagar, Bagalkot, -587102, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Prakash S Handi,
Associate Professor, Department of Otorhinolaryngology and Head Neck Surgery,
S N Medical College Navanagar Bagalkot -587102
Karnataka-India.
E-mail: prakashhandi@yahoo.com

FINANCIAL OR OTHER COMPETING INTERESTS:

None.

Date of Submission: **Jun 22, 2012**
Date of Peer Review: **Jun 30, 2012**
Date of Acceptance: **Jul 06, 2012**
Date of Publishing: **Aug 10, 2012**