The Journal of Laryngology & Otology
July 2004, Vol. **118,** pp. 566–569

Surgical approach for congenital midline cervical cleft

Y. Bajaj, M.R.C.S., D. Dunaway*, F.R.C.S., B. E. J. Hartley, F.R.C.S.

Abstract

This is a report of a case of a rare congenital midline cervical cleft and a description of the surgical approach.

Congenital midline cervical cleft is a very rare developmental anomaly. It represents a failure of the branchial arches to fuse in the midline and presents at birth with a ventral midline defect of the skin of the neck. Associated clinical features could include mandibular spurs, cleft mandible, microgenia, thyroglossal cyst, cleft lip or bronchogenic cysts.

The authors present a case of a midline cervical cleft that was diagnosed and managed at an early age. They discuss the clinical presentation and embryological development of this rare condition. The operative findings, surgical excision and repair of the long vertical defect by Z-plasty are discussed in detail.

The authors report a very satisfactory result following excision and Z-plasty closure of this rare congenital anomaly.

Key words: Branchial Region; Surgery, Plastic

Introduction

Congenital midline cervical cleft (CMCC) is a rare developmental anomaly of the ventral neck. It has been described with various terms such as medial cleft,[1] median fissure of the neck, congenital midline cervical cord,[2] midline cervical webbing[3] and pterygium colli medianum.[4] CMCC has not been widely reported in ENT or general practice literature. It may be misinterpreted as a branchial cleft deformity or thyroglossal cyst.[5] Although developmentally related to these disorders, CMCC represents a distinct anomaly that should be recognized at initial examination. The parents can then be appropriately counselled about the implications and management that is peculiar to this disorder.

Bailey[6] first described the anomaly in 1924. However, until now it has been a relatively unknown entity and there is much controversy over its aetiology. Several pathogenic mechanisms have been proposed to explain midline cervical clefting, but embryo pathogenesis of the anomaly has not been established clearly. CMCC is thought to be sporadic,[4] and is primarily seen in white females.[1,7,8] Although not common in the general population, it is more common among those with thyroglossal and branchial cleft anomalies.[8] Untreated the lesion affects neck movements and mandibular growth so surgery is recommended.

The authors present the case of a patient with CMCC to increase awareness amongst ENT surgeons, paediatricians and general practitioners about this midline anomaly. Surgical excision creates a sizeable vertical defect in the neck, which is likely to lead to significant scarring or webbing. A Z-plasty approach is advocated. The clinical findings, embryology, histopathology and surgical management are discussed.

Case report

A six-month-old boy was referred to the authors' department because of a midline cervical abnormality. The patient was born after an uncomplicated pregnancy and delivery. Apart from this lesion in the neck, the child was otherwise normal. Both the parents were in good health and there was no family history of any congenital defects. According to the history at birth the child had an ulcerated lesion in the midline ventral neck approximately 3 cm in length and 1 cm wide. This had healed to leave a small midline sinus when the child was first seen. The mother had noticed that movement of the head was partially restricted.

On examination, there was a linear cleft in the midline of the child's neck midway between the mandible and the sternal notch and covered by atrophic skin measuring 3 cm in length and approximately 5 to 10 mm wide (Figure 1). At the cephalic end of the cleft, there was a soft pink sessile papule with a diameter of 5 mm. There was a sinus in the upper part of this lesion. No secretions or discharge were noted. There was some tethering to the sternum and an underlying cord could be felt. When the patient's head was dorsiflexed, midline webbing appeared that extended from the submental region to the manubrium sternum. The chin appeared widened giving the impression of being bifid. There were no other head and neck abnormalities. Thorough laryngopharyngeal and paediatric examination showed no abnormalities in the laryngopharynx, the sternum and the rest of the thorax or the heart. An ultrasound scan was performed that confirmed a normal thyroid with no associated cysts. A diagnosis of midline cervical cleft was made and it was decided to excise the lesion.

From the Departments of Otolaryngology and Plastic Surgery*, Great Ormond Street Hospital, Great Ormond Street, London, UK.
Accepted for publication: 26 February 2004.

https://doi.org/10.1258/0022215041615074 Published online by Cambridge University Press

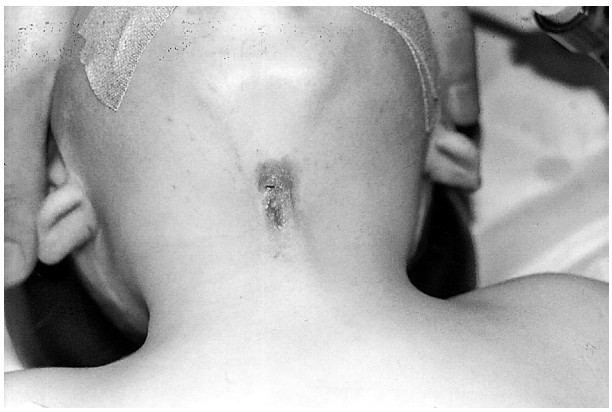

FIG. 1
Midline cervical cleft.

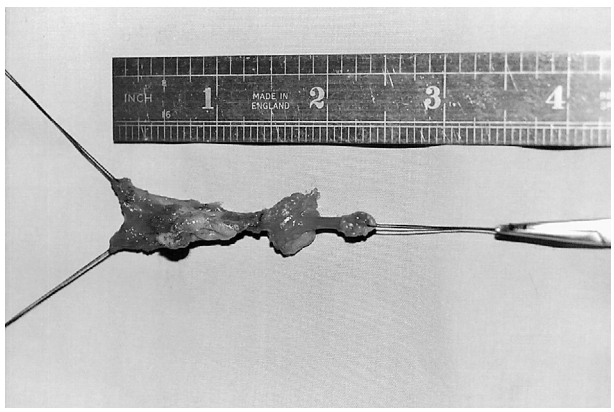

FIG.3
Excised specimen.

Operative procedure

Surgical excision using elliptical incision and repair by Z-plasty was planned. An elliptical incision was made enclosing the lesion (Figure 2). The lesion was dissected. There was a Y-shaped fibrous band attached to this lesion. Two limbs of the fibrous band extended superiorly and were attached to the mandible and one limb extended inferiorly to the sternum (Figure 3). A bony callus was felt at the inferior border of the mandible where these bands were attached, probably due to traction. Complete excision of the lesion was performed and the skin was closed with a Z-plasty approach. The two limbs of the Z were made at the ends of the spindle as shown in Figure 2. The flaps were elevated and rotated to close the defect (Figure 4). There were no complications and the post-operative recovery was uneventful. (The child was able to move his head around better on the same day.) The scar was healing very well three weeks post-operatively.

Histologically, the excised lesion was reported as skin that had a cleft lined by squamous epithelium in the centre. The cleft was surrounded by focal areas of mild chronic non-specific inflammation. The underlying tissue had focal areas of dermal fibrosis containing a small amount of striated muscle, fibrous tissue and fat. The histological findings were consistent with a diagnosis of CMCC.

Discussion

Congenital midline cervical cleft is a rare anomaly that is present at birth but may be overlooked or misdiagnosed.[9] The cleft occurs in the midline at the anterior aspect of the neck and extends over a variable length in a vertical direction from beneath the chin to the suprasternal notch. CMCC is reported predominantly in females.[8]

When observed at birth, the area was weeping, and covered by a thin, often desquamating epithelium, but within the next few months the epithelium toughened and dried up. It healed with scarring. The cephalic end of the cleft was distinguished by a nipple-like protuberance, whereas the caudal end of the cleft presented an opening to a sinus track from which mucoid secretions discharged.[10] This mucoid discharge can be explained by the presence of ectopic salivary glands in some cases.[11] Beneath the epithelium there may be a firm fibrous cord.[5]

Additional features associated with CMCC include a cephalad skin protuberance, a caudal blind mucosal tract and a subcutaneous fibrous cord that originates from the deep layer of the skin tag and ends in the subcutaneous tissue of the chin. This latter feature may be asymptomatic or it may cause webbing of the neck with extension, known as pterygium colli medianum.[5] In severe cases there is a contracture, limiting the extension of the neck or even depressing the lower lip during extension of the neck.[2] Such a contracture is thought to develop with growth of the neck and to be the result of cicatrisation of the cleft.[11] In some cases a bony prominence of the mandible could be palpated, the spur of which is visible on radiographic examination.[2] This spur can be due to traction of the fibrous cord on the bone.

In addition to its central features, CMCC may be associated with other anomalies of the head and neck, including clefts of the lower lip, mandible, chin and tongue, an underdeveloped mandible and hypoplasia/absence of supporting structures of the neck, such as hyoid bone.[5,9] It

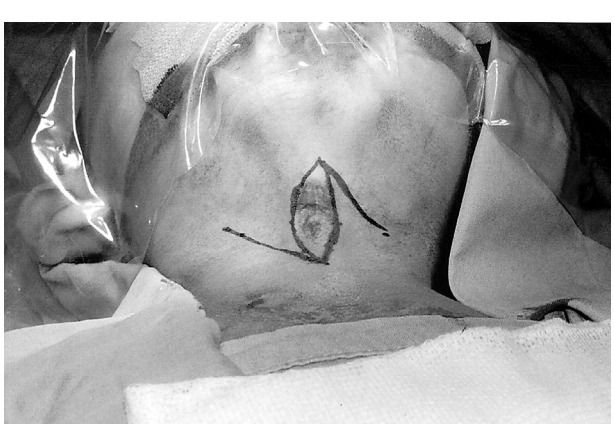

FIG. 2
Incisions planned for excision and Z-plasty.

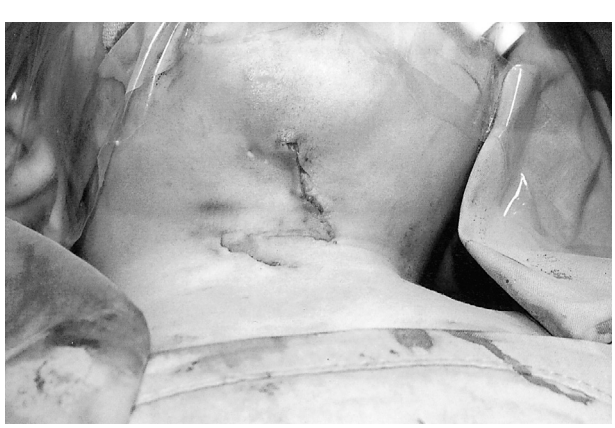

FIG. 4
Z-plasty closure.

https://doi.org/10.1258/0022215041615074 Published online by Cambridge University Press

may be related to both bronchogenic cysts[11] and thyroglossal duct abnormalities.[6,7] CMCC has also been associated with defects in other parts of the body, including cleft sternum, midline abdominal raphe, midline haemangioma as well as congenital heart lesions.[4,9]

- **Congenital midline cervical cleft is a rare developmental anomaly representing failure of fusion of the branchial order in the midline**
- **A case of Ludwig's angina of odontogenic origin is presented that progressed into mediastinitis and pericarditis**

Embryology

The embryological mechanism that accounts for CMCC is not firmly established; however, most investigators believe that it involves a failure of the branchial arches to fuse in the midline.[2,12-15] During normal embryological development, the branchial arches begin to merge in a cephalad to caudal fashion during the third and fourth weeks of gestation. The first (mandibular) arches fuse prior to the second (hyoid) arches, and the rest follow sequentially. Before the arches come together, mesodermal tissue must migrate between them and push the ectoderm outward, flattening the ventral furrow. Disruption of this process may result in various defects.[9]

A number of hypotheses have been set forth to explain the mechanism of faulty closure of the branchial arches. These include increased pressure on the cervical area from the pericardial roof in early stages of the developing embryo;[12] persistent remnants of the thyroglossal duct and sinus cysts;[7] rupture of a pathologic adhesion between the epithelium of the cardiohepatic fold with that of the ventral part of the first branchial arch; vascular anomalies, causing localized tissue ischaemia, necrosis and scarring[16] and absence of mesenchymal masses in the cervical midline.[2]

Although the exact arches involved and the manner of abnormal cell migration are still debated, morphologically CMCCs can be divided into two major groups depending on the involved arch or arches.[8,17] An isolated CMCC develops if the second (hyoid) arches did not fuse. If a bilateral defect of the first (mandibular) arches was present, then the defect would be more extensive, involving clefts of the lower lip, tongue, and mandible or the absence of the supportive structures of the neck.[4,18]

Some authors dispute the branchial arch origin of CMCC. A bronchogenic origin of CMCC has been proposed due to pathologic findings suggesting the presence of respiratory epithelium or bronchogenic cysts.[16] Also, it has been suggested that it occurs secondary to exteriorization of a thyroglossal remnant with subsequent exposure of the mucosa.[7]

On the basis of this data it is difficult to explain the exact embryologic mechanism of the anomaly, but the authors conclude that CMCC represents a developmental defect involving branchial arches, grooves and pouches to a more or less severe extent.[19]

Histopathology

Histologically, CMCC usually consists of parakeratotic stratified squamous epithelium without normal skin appendages.[5,11] The underlying dermis may, or may not, show areas of alternating hypertrophy and atrophy,[5] and the subcutis may contain dense fibrous connective tissue and a mild inflammatory infiltrate of primarily lymphocytes, plasma cells and neutrophils.[11] The skin tag generally consists of normal overlying skin, but may contain cartilage[20] and irregular skeletal muscle.[18] The sinus tract associated with CMCC usually consists of pseudo-stratified columnar or cylindrical epithelium and often demonstrates adjacent seromucinous glands.[11,21] In some cases, the histology of the fistula resembles bronchogenic epithelium[16,21,22] or contains part of the thyroglossal duct or its remnants.[6,7] A central fibrous cord can be seen beneath the squamous epithelium.[5,16] It has its own distinct microscopic structure and may include interfasciculated bundles of skeletal muscle.[2,18]

Surgical treatment

The treatment of CMCC is surgical removal. Early intervention is recommended because of the nonaesthetic appearance of the malformation, as well as to avoid subsequent limitation of the extension of the neck or impairment of mandibular growth. It is recommended that all of the pathologic tissue including the fibrous cord be completely removed. Simple excision or transaction of the fibrous band leads to recurrence of scarring and fibrosis.[21] The defect should be closed using Z-plasty technique. It has been reported that straight-line closures are not as cosmetically pleasing due to recurrent scarring[11] and they lead to a higher incidence of neck contracture recurrences.[21]

Conclusion

Congenital midline cervical cleft is a rare developmental anomaly of the neck that may be isolated or associated with a spectrum of midline defects. It is a vertical midline skin defect of the neck, with a nipple-like nodule on the superior portion of the defect. The cleft usually consists of a stratified squamous epithelium without any skin appendages and may contain cartilage and skeletal muscle. There are various theories about its origin. Most believe it is due to impaired fusion of the first and second branchial arches. The recommended treatment is complete surgical excision without which problems with mandibular growth and neck movements are anticipated. The sizeable neck defect can be closed very effectively by the Z-plasty procedure.

References

1 Davis AD. Medial cleft of the lower lip and mandible. *Plast Reconstr Surg* 1950;**6**:62–7
2 Van Duyn J. Congenital midline cervical cord with report of a case and note on the aetiology of congenital torticollis. *Plast Reconstr Surg* 1963;**31**:576–86
3 Wood GA, Deister E. Anterior midline neck webbing with microgenia and symphyseal exostosis. *Oral Surg* 1983;**56**: 128–31
4 Godbersen S, Heckel V, Wiedeman HR. Pterygium colli medianum and midline cervical cleft: Midline anomalies in the sense of a developmental field defect. *Am J Med Genet* 1987;**27**:719–23
5 Maddalozzo J, Frankel A, Holinger LD. Midline cervical cleft. *Paediatrics* 1993;**92**:286–7
6 Bailey H. Thyroglossal cysts and fistulae. *Br J Surg* 1924; **12**:579–89
7 Gross RE, Connerley ML. Thyroglossal cysts and sinuses. *N Engl J Med* 1940;**223**:616–24
8 Gargan TJ, McKinnon M, Mulliken JB. Midline cervical cleft. *Plast Reconstruct Surg* 1985;**56**:128–31
9 Eastlack JP, Howard RM, Frieden IJ. Congenital midline cervical cleft: Case report and review of the English language literature. *Pediatr Dermatol* 2000;**17**:118–22

https://doi.org/10.1258/0022215041615074 Published online by Cambridge University Press

10 Hirokawa S, Uotani H, Okami H, Tsukada K, Futatani T, Hashimoto I. A case of congenital midline cervical cleft with congenital heart disease. *J Pediatr Surg* 2003;**38**: 1099–101
11 Van der Staak FHJ, Pruszczynski M, Severijnen RSVM, Van de Kaa CA, Festen C. The midline cervical cleft. *J Pediatr Surg* 1991;**26**:1391–3
12 Morton CB, Jordan HE. Median cleft of lower lip and mandible, cleft sternum and absence of basihyoid: Report of a case. *Arch Surg* 1935;**30**:647–56
13 Wynn-Williams D. Congenital midline cervical cleft and web. *Br J Plast Surg* 1952;**5**:87–93
14 Gottlieb E, Lewin ML. Congenital midline cervical clefts of the neck. *N Y State Med J* 1966;**66**:712–8
15 French WE, Bale GF. Midline cervical cleft of the neck with associated branchial cyst. *Am J Surg* 1973;**125**:376–81
16 Braithwaite F, Watson JA. A report on three unusual cleft lips. *Br J Plast Surg* 1949;**2**:38–49
17 Nicklaus PJ, Forte V, Friedberg J. Congenital mid line cervical cleft. *J Otolaryngol* 1992;**21**:241–3
18 Bergevin MA, Sheft S, Myer C, McAdams AJ. Congenital midline cervical cleft. *Pediatr Pathol* 1989;**9**:731–9
19 Ercocen AR, Yilmaz S, Handan A. Congenital midline cervical cleft: Case report and review. *J Oral Maxillofac Surg* 2002;**60**:580–5
20 Ikuzawa M, Matsumoto K, Sakuda M. Midline cervical cleft: a case report. *Int J Oral Maxillofacial Surg* 1992;**21**: 258–9
21 Fincher SG, Fincher GG. Congenital midline cervical cleft with subcutaneous fibrous cord. *Otolaryngol Head Neck Surg* 1989;**101**:399–401
22 Ayache D, Ducroz V, Roger G, Garabedian EN. Midline cervical cleft. *Int J Pediatr Otorhinolaryngol* 1997;**40**: 189–93

Address for correspondence:
Mr Yogesh Bajaj,
64, Westwood Park Road,
Peterborough,
PE3 6JL, UK.

Fax: 01733 311041
E-mail: yogesh_bajaj@hotmail.com

Mr Y. Bajaj takes responsibility for the integrity of the content of the paper.
Competing interests: None declared

https://doi.org/10.1258/0022215041615074 Published online by Cambridge University Press