

A rare congenital midline cervical cleft

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A rare case of congenital midline cervical cleft (CMCC) is presented to highlight the importance of correct recognition of the lesion and appropriate surgical management.

A 2-week-old female baby was referred to the paediatric outpatient department at Red Cross War Memorial Children's Hospital with an anterior midline neck lesion that had been diagnosed at birth. She had been born by vaginal delivery after an uncomplicated term pregnancy. Her birth weight was 3 280 g, and she was treated for neonatal jaundice. There was no family history of similar lesions.

On physical examination she had a vertical linear lesion that began 1.0 cm above the sternal notch and was 3.0 cm in length with a nipple-like protuberance at the cranial end (level of hyoid bone) (Fig. 1). A superficial midline band was palpable subcutaneously. A blind-ending sinus was noted at the caudal end of the cleft. No discharge from the sinus was noted.



Fig. 1. Congenital midline cervical cleft.

No bony spurs on the mandible or sternum were detected on clinical or radiological examination. An ultrasound scan of the thyroid gland showed it to be normal.

At the time of scheduled surgery, at 8 months of age, the CMCC was limiting the patient's neck extension. Elliptical surgical excision of the entire cleft and repair by Z-plasty was undertaken. A blind-ending sinus that ended on the manubrium sternum was dissected out. The lesion did not appear to extend up to the mandible or involve the hyoid bone.

Histological examination of the resected specimen revealed epidermis overlying prominent skeletal muscle in sections

of the superior portion. Respiratory-type epithelium and seromucinous glands were present along the invaginated surface more distally. The distal band showed dense collagen.

When the patient was seen at follow-up 6 months later there was a good cosmetic result and no residual contractures.

Discussion

CMCC is a rare condition, less than 100 cases having been reported in the English literature so far. The first recorded case of CMCC was in 1848 by Luschka. In a series of 672 thyroglossal and other branchial cleft sinuses over a 30-year period, Gargan *et al.*¹ reported 12 CMCCs (1.7% of their series).

This lesion is often misdiagnosed at birth by the primary care physician as a thyroglossal duct cyst² or, as in one reported case, referred to the dermatology service as an 'unusual birthmark'.³ Complete surgical excision is important to prevent neck contractures. Patients with CMCC seem to be referred to various disciplines, depending on the often incorrect diagnosis by the primary physician. The lesion has been reported in plastic and reconstructive,⁴ otorhinolaryngology,⁵ genetics and even dermatology journals.³

The lesion varies in size from patient to patient, but common features include a superior midline skin nipple, a midline linear area of atrophic skin extending from that nipple, and sometimes a caudal sinus tract. In some cases a bony spur is palpable on the mandible or a small cleft is felt on the manubrium sternum.

The embryological origin of CMCC has been disputed, but the most widely accepted explanation is failure of fusion of the branchial arches in the midline.⁶ It is not known which branchial arch failure is responsible.⁶ The postulated reasons for failure of fusion include pressure by the pericardial roof on the most distal branchial arches, and vascular anomalies that may result in necrosis and subsequent cleft formation.² Poor interaction between mesoderm and ectoderm may also prevent fusion.² CMCC is sporadic but does tend to have a female preponderance.

In the cases reported in the available literature, the Z-plasty technique has been the favoured and advocated technique of repair of CMCC.⁵ Z-plasty allows for lengthening of the anterior neck skin which assists in preventing contractures. In the recent literature there appear to be more reports of simple excision and closure^{2,3} or an alternative flap technique⁴ without adverse outcomes.

It is important for paediatricians and primary care physicians to recognise this disorder, counsel parents appropriately, and refer patients early and suitably for surgery.

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Melanoma in black South Africans

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