Midline Cervical Clefts: A Rare Anatomic Anomaly

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Summary

Purpose of the report: Midline cervical clefts (MCC) are rare congenital abnormalities thought to occur from abnormal fusion of the branchial arches at the midline. Closure types range from multiple z-plasties for larger defects to w-plasties to primary closure for mild soft tissue deficiencies. We present a 4 case series of midline cervical cleft repairs.

Design and method of study and analysis: Review of an ongoing case series of midline cervical clefts repaired by the chief of pediatric otolaryngology at our institution.

Summary of results: Three of the four cases received opposing z-plasties of the platysma in hopes of improving definition of the cervicomental angle as primary closure in the vertical or horizontal plane could result in unacceptable scarring or contracture. The fourth case had a mild soft tissue deficiency, and was closed primarily. He is currently being followed to determine if his cervicomental profile is as well defined as the other three patients in our series.

Conclusions: The overall lateral cervicomental profile were improved in all three patients who received the z-plasty. Scarring in the horizontal plane of the z-plasty is excellent, though hypertrophic scarring is more common at the oblique closures. Although no consensus exists regarding optimal surgical management of MCC, opposing z-plasties of the subcutaneous tissue and the platysma appear to improve the cervicomental contour of our patients, and should be considered when planning surgical reconstruction.

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CASE REPORT 1

A 16 day-old male infant presents with a small 7mm midline mass with a de-epithelialized undersurface, and a subcutaneous fibrotic band extending from the mentum to the manubrium (Figure 1). The patient was brought in for surgical excision at 6 months of age. A U-shaped incision along the inferior aspect of the skin tag was made, and the fibrous cord was completely dissected from the surrounding subcutaneous tissue from the manubrium. An elliptical excision rather than a z-plasty was performed due to sufficient space between the skin and the fibrous band that lay in the subcutaneous layer. The platysmal layer was closed vertically, and the subcutaneous layer was closed horizontally. This was the only case in our series where the incision was closed primarily.

CASE REPORT 2

A 1 month-old female was diagnosed with congenital midline cervical cleft. Physical exam reveals a vertical cleft dehiscence, dimpling at the submentum, and a fistulous tract extending to the manubrium. At 1-year of age, the patient underwent surgical excision with a double-opposing Z-plasty repair for closure. The entire cleft and sinus were carefully dissected away and sent as pathologic specimen. Incision through the skin and subcutaneous tissue was completed through the limbs of the planned z-plasty incision. Next an opposing z-plasty, approximately 60 degree angles from the vertical midline defect, was performed on the platysma in a fashion to improve the cervical mental angle. (Figure 2A-2B).

CASE REPORT 3

A 4 year-old female patient presents with a previously diagnosed MCC from birth. Palpation of the neck revealed a fibrous cord from mentum to manubrium and the presence of an open ventral cleft at the cervicomental junction 1.5 cm wide. The midline web extended superiorly with fibrous attachments to the mandible as well as the dermis. (Figure 3A, 3B). A cervical sinus tract was present, but no drainage was observed. Patient had the MCC surgically removed shortly thereafter. The skin was incised vertically down to the layer of the platysma, and the sinus tract was dissected out (Figure 3C). Deep portions of the dissection extended down to the level of the superficial layer of deep cervical fascia. A double Z-plasty, approximately 60 degree angle from the vertical midline, was incised through skin and subcutaneous tissues. Postoperative outcome revealed good healing (Figure 3D-3E).

CASE REPORT 4

A 19 month-old female patient presents with features of congenital midline cervical cleft anomaly of the neck with a darkened periphery of deformed skin inferior of the mentum. A sinus was found upon inspection with no drainage noted. The patient underwent surgical excision shortly after presenting to clinic. An elliptical incision was made, and the z-plasty limbs were incised. The cord was isolated and completely removed from the neck. The z-plasty limbs were then further developed. Initially the skin and subcutaneous tissue was dissection off the surface of the platysma bilaterally. The opposing z-plasty was then drawn out on the platysma and incised. The platysma was then elevated off the underlying fascia of the sternocleidomastoid muscle and the deep fascia of the neck and closed.

DISCUSSION

Midline cervical cleft is a rare congenital anomaly of the head and neck with the first case being documented in 1924 by Bailey. Since then, approximately 100 cases have been reported in the English language. The pathogenesis of MCC is still debatable with the following theories:

1.) MCC is the result of improper fusion of the paired first and second arches in the ventral midline during weeks 3 and 4 of embryogenesis. MCC may be caused by defective interactions between ectoderm and mesoderm during embryogenesis causing failure of the midline arch to close as well as possibly improper differentiation of mesenchymal tissue.

2.) MCC may have been due to vascular anomalies and over-exerted pressure in the cervical area by the pericardial roof during embryogenesis.

3.) MCC has also been proposed to be an external remnant of cervicomental sinus tract extending to the manubrium.

Treatment for MCC usually consists of complete excision of the pathological tissue, cord, and cervical cleft mass followed by closure. Z-plasty has been the most frequently documented treatment option.

Z-plasty is a common surgical technique that involves making two triangular flaps upon incision and alternating the original positions of the flaps to close the central incision. Z-plasty disrupts the linear formation of the incision, resulting in better healing for anatomical functionality as well as less vertical scar development compared to a linear closure.