INTRODUCTION

Congenital midline cervical cleft (CMCC) is a rare developmental anomaly of the anterior neck. The etiology remains unclear, but is likely related to a failure of the branchial arches to fuse in the midline.\(^1\) The lesion presents at birth as a midline defect of the cervical skin occurring anywhere between the mandible and sternal notch. The diagnosis is clinical, and can be associated with defects of the mandible or sternum, thyroglossal cyst, defects of the hyoid bone, cleft lip, or bronchogenic cysts.\(^2,3,4\) There appears to be a predilection for Caucasian females.\(^5\) The defect is typically associated with a fibrous subcutaneous band which can tether the movements of the neck and, if severe enough, can lead to mandibular defects. Clinical characteristics of CMCC can vary in degree but typically include a midline defect of atrophic or raw appearing skin, a subcutaneous fibrous cord and a thickened nipple-like protuberance at the cephalad aspect of the lesion.\(^6\) Surgical excision of the defect including the fibrous band is advocated in order to avoid long term sequelae.

CASE PRESENTATION

An 8 month old girl presented to our clinic with a midline anterior neck lesion which was diagnosed as a CMCC. She was taken to the operating room for surgical excision with multiple z-plasty reconstruction. On 10 month followup she demonstrated good cosmetic results with no tethering of the anterior neck and no associated mandibular defect.

TREATMENT

Several surgical approaches have been described including primary closure and various local flap reconstruction.\(^1-6\) Simple primary closure has been reported to lead to hypertrophic scarring and neck contracture. The multiple z-plasty approach re-orient the scar, lengthens the defect, and creates a broken-line appearance which is better camouflaged and more aesthetically pleasing. Multiple z-plasty also allows closure of longer defects under minimal tension when compared to single z-plasty alone.

CONCLUSIONS

CMCC is an unusual congenital defect that may be encountered by the otolaryngologist. Surgical excision of the entire defect with multiple z-plasty reconstruction yields good cosmetic results while improving neck mobility and reducing the risk for long term cervical tethering and mandibular defects.

REFERENCES

1. Franzese C, Hayes JD, Nicholas K. Congenital midline cervical cleft: A report of two cases. ENT Journal 2008;87:166-8