

Congenital Midline Cervical Cleft: Can It Be Treated in Newborn?

Roya Farhadi¹, MD; Alireza Alam Sahebpoor^{*2}, MD, and Maryam Ghasemi³, MD

1. Department of Neonatology, Department of Pediatrics, Mazandaran University of Medical Sciences, Sari, Iran
2. Department of Pediatric Surgery, Mazandaran University of Medical Sciences, Sari, Iran
3. Department of Pathology, Mazandaran University of Medical Sciences, Sari, Iran

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Abstract

Background: Congenital midline cervical cleft (CMCC) is a rare developmental anomaly of the anterior neck, mostly found in Caucasian females. Most authors consider it within the spectrum of branchial arch developmental abnormalities. There have been about 100 reported cases in the literature. However, there is no agreement on the age of surgery in this anomaly.

Case Presentation: In this report we present our experience of managing this anomaly in the early neonatal period in two neonates. One of them had concomitantly congenital heart disease.

Conclusion: It is important to recognize this rare congenital anomaly in neonate. Although the exact time of surgery for CMCC is not mentioned, based on our results the earliest time of intervention is recommended since then the operating result will be excellent.

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Introduction

Congenital midline cervical cleft is an uncommon congenital malformation^[1] which was completely described by Ombreadanne in 1946^[2]. Although it appears clinically in the first day of life and the diagnosis is typically made on the basis of the lesion's characteristic clinical presentation at birth, the cleft may not appear very striking in a newborn infant^[3,4]. When observed at birth the characteristic features are a defect at the ventral area of the neck with subcutaneous fibrous cord and a nipple like projection at the upper part and a sinus or fistulous tract at the lower end of the defect^[5]. The defect lies between the mental areas superiorly to the suprasternal notch, inferiorly

with variable length and width^[5]. Most cases are sporadic and it is more common in Caucasian girls^[6]. There have been less than 100 cases in the literature^[2].

Ultrasound is sometimes used to search for other cervical anomalies^[6]. Treatment is surgical excision and closure of the defect^[1]. Linear scars are much more noticeable than broken line, so closure with Z-plasty is done to prevent neck contracture^[1,7,8]. There is no agreement on the appropriate age for surgical correction of this anomaly. We here report our experience with two neonates operated in the early days of life because their parents preferred so. We evaluate the cosmetic and functional results of early surgery in this congenital anomaly.

* Corresponding Author;

Address: Department of Pediatric Surgery, Boooli Sina Hospital, Sari, Iran

E-mail: alireza_alam@yahoo.com

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Case Presentation

Case 1

A one-day-old boy was admitted in our neonatal unit with a midline cervical abnormality. The neonate was born by caesarean section after an uncomplicated pregnancy and weighed 3200 gram having a normal Apgar score. There was no family history of congenital defects and his parents were also in good health. On examination there was a midline vertical mucosal surface in the anterior neck extending from below the mental process of mandible to suprasternal area with no skin covering. The defect was 3 cm in vertical length and 0.5 cm in width. A nipple like projection was observed in the upper part of the defect and there was a sinus in the lower part of the defect above the manubrium. The sinus was examined by a small lacrimal probe and a narrow tract coursing toward the suprasternal notch was observed.

A fistulogram was not done, but ultrasound scan of the neck showed no evidence of any cervical abnormality (Fig. 1A). Auscultation revealed heart murmur caused by small ASD shown in echocardiogram. The chest X-ray was normal.

The infant was operated on the fifth day of life by the preference of the parents. A vertical elliptical incision was done around the cervical cleft and the mucosal defect, and then all the pathologic tissues with the underlying fibrotic cord were completely removed. The fibrous cord attached to the manubrium was removed. The skin defect was closed by 2 serial Z-plasties. The 4 resultant flaps were sutured first by vicryl 5-0 for dermal suture and by nylon 6-0 for skin closure (Fig. 1B).

Pathologic results showed that the skin cleft was covered by a thin epidermis with irregular papillomatosis, located on a fibrotic dermis devoid of skin appendages. A small skin projection was seen consisting of mature fibro fatty tissue, haphazardly arranged striated muscle bundles, and a normal epidermis as covering.

Also a sinus tract lined by parakeratotic stratified squamous epithelium as well as respiratory epithelium was observed. The sinus tract was surrounded by a mildly inflamed fibro vascular stroma enclosing a few striated muscle bundles and mucous secreting glands.

Post operative course was uneventful and there was no wound infection. In a follow-up examination, 1 and 6 months after operation, there was excellent wound healing of the Z-plasty and no wound contracture in the neck could be seen (Fig. 1C).

Case 2

A 2800 gram full-term newborn girl was born by caesarean section with an anterior midline neck defect that was 25 mm in length and 0.5 cm in width. There was a nipple like projection at the cephalic end and a sinus tract opening caudally (Fig. 2A). She had no stridor, and no discharge from the sinus was noticed. Other physical examination and routine laboratory data were normal. Ultrasound scan of the neck confirmed a normal thyroid gland, as well. No other associated malformations were observed. At 4 days of age, because of the request put forth by the parents and the good general health condition of the neonate, she underwent an operation for cervical defect correction. The cervical cleft and all the pathologic tissues with the underlying fibrotic cord were removed by surgical excision. The vertical defect was closed by 2 serial Z-plasties (Fig. 2B). Pathology showed thin layer of epidermis overlying skeletal muscle in sections of the cleft and superior portion. In sections of the sinus tract, respiratory epithelium with parakeratosis and seromusinous glands compatible with histological findings of CMCC was observed. In a 2 and 4 month post operative follow up, she had normal wound healing without any contracture and with good cosmetic result and the parents did not appear for follow up (Fig. 2C).

Discussion

CMCC with about 100 cases reported in the literature is a rare congenital anomaly of anterior neck region^[2,9]. It can be extended in the midline from mandible to manubrium and the length and width of the lesion can differ from one child to another. Although, the anomaly is mostly seen in females^[10], our first case was a male.

The anomaly presents a linear vertical area of



Fig. 1: Case 1, A. Congenital midline cervical cleft, B. Closure of the defect by Z-plasty technique, C. 6 months after operation



Fig. 1: Case 2, A. Congenital midline cervical cleft, B. Closure of the defect by Z-plasty technique, C. 4 months after operation

thin and erythematous mucosa at birth. There is often a nipple-like projection in upper part and a sinus or fistula at the lower end. Sometimes there is a fibrous band beneath the mucosal defect, too. If not treated in newborns, the midline cord begins to act as a tethered as the infant grows. Thus, surgical excision has both cosmetic and functional benefits.

CMCC may be associated with other anomalies like thyroglossal duct cyst^[11,12], ectopic bronchogenic cyst^[13,14], cleft lower lip, tongue and mandible^[15], cleft sternum, and ectopia cordis with intracardiac anomalies^[16].

Associated congenital heart diseases have been reported occasionally with cleft sternum or ectopia, too^[17]. ASD observed in our first case is similar to a case reported by Shinichiro et al; it can be due to broad clinical manifestations of midline anomalies^[17].

The common finding on histological examination of the specimen is a thin atrophic epidermis with absence of skin appendages in the dermis and fibrous connective tissue^[5].

The embryologic mechanism of this anomaly is not yet clear; however, an impaired fusion of the distal branchial arches in the midline is the most commonly accepted theory^[4].

Diagnosis is done by clinical examination of the lesion at birth and the treatment is surgical excision. Although there is no agreement on the appropriate age of treatment of CMCC, we recommend early intervention because of the disfiguring appearance of the lesion. Also, if not treated early, the lesion behaves like a cicatrix and, becoming a midline cervical cord, acts as a tether. This in turn will cause limitation in the extension of the neck as the child grows older. Furthermore, Ercocen et al suggested that early intervention avoids the disfiguring appearance of the malformation and also prevents subsequent limitation of neck motion^[18].

Derebz et al reported 5 cases of CMCC. All were treated in early life at the age ranging from 1 month to 2 years. They concluded that surgical repair should be done as soon as possible to reduce the risk of recurrence and avoid limitation of neck extension^[2].

Treatment consists of complete excision of the neck lesion and closure of the defect by Z-plasty technique. Linear closure will result in hypertrophic scarring and contracture. Spencer Cochran et al recommended single Z-plasty to be appropriate for lesions less than 2 cm, and serial Z-plasties for longer lesions^[2]. We used two serial

Z-plasties because the resultant defect after the excision of the lesion was more than 2cm in length.

Conclusion

It is important for neonatologists and pediatric surgeons to recognize this rare congenital anomaly. Although the exact time of surgery for CMCC is not clearly mentioned, based on our results the earliest possible time of intervention is recommended as the operating result consequently will be excellent. Our post operative follow up showed excellent midline cervical scar with no wound contracture. Also the parents were pleased to take the child home with their anomaly being corrected. Based on these results we suggest early operation for CMCC.

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