The THAI Journal of SURGERY 2017;38:147-150.
Official Publication of the Royal College of Surgeons of Thailand

Case Report

Congenital Midline Cervical Cleft: A Case Report

Yaowalak Khomnuan, MD

Pediatric Surgery Unit, Department of Surgery, Buddhachinnaraj Hospital, Phitsanulok, Thailand

Abstract **Congenital midline cervical cleft (CMCC) is a rare anomaly of the ventral midline of neck. This anomaly is always present at birth with a midline vertical atrophic skin defect, superior skin-tag like a nipple and an inferior sinus tract. It can be diagnosed and surgically treated in an infancy period. The author reported one case of female neonate with CMCC and operated on one month of age, in order to share experience of this rare anomaly.**

Keywords: **Congenital anomaly, midline cervical cleft, branchial arch defect**

Introduction

Congenital midline cervical cleft (CMCC) is a rare anomaly of the ventral midline of neck and can be diagnosed at birth[1-4]. From the international literature review by Puscas[5], at least 195 cases reported in 2015. The embryological origin is not clear. Clinical presentations of CMCC can be identified since birth. The presenting signs appear a cleft defect at the anterior surface 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 of the defect[1-4]. It may develop secondary complications, such as impairment of neck extension, deformity of the jaw, torticollis and wound infection due to misdiagnosis and incorrect treatment[6,7]. In the present study, the author reported one case of CMCC which was diagnosed and surgically treated in the neonatal period.

Case Report

A 2-week-old female neonate was transferred from a rural hospital to Buddhachinnaraj Hospital because of a midline cervical lesion. She was conceived from a 31-year-old mother with 38-gestational age and delivered by cesarean section. Her birth weight was 2,505 grams and the APGAR scores were 9 and 10 at the 1 and 5 minutes, respectively.

On physical examination, her temperature and vital signs were normal. A midline cervical lesion revealed a linear cleft extending from the level of the hyoid bone to the suprasternal notch. Average size of the cleft was 2 cm in length and 0.5 cm in width. There was a nipple-like skin swelling at the cephalic end and a sinus opening with mucoid discharge at the caudal end (Figure 1). No associated anomaly was noted.

Correspondence address: *Yaowalak Khomnuan, MD, Pediatric Surgery Unit, Department of Surgery, Buddhachinnaraj Hospital, Phitsanulok 6500, Thailand; Telephone: +66 5527 0300; E-mail: yao_vy@hotmail.com*

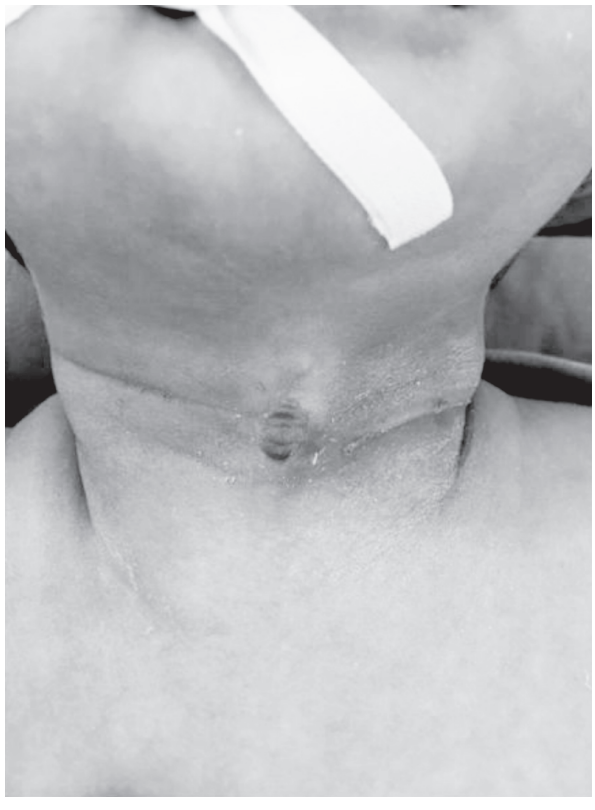

Figure 1 A one-month female neonate with a midline cervical cleft consisted of a nipple like skin tag at the superior end and a sinus tract at the inferior end

Figure 2 The excised specimen with a probe inserted in the inferior sinus tract

She underwent surgical treatment under general anesthesia at the age of one month old. An elliptical incision was performed around the edges of lesion through the healthy skin margin. Complete excision of all the pathologic tissue was done with primary skin closure by subcuticular suturing (No. 6-0 absorbable sutured material).

The specimen consisted of an elliptical shaped skin, measuring 2.0 × 0.6 × 0.5 cm. It showed an ulcerative lesion with papillary projection of yellow tissue, measuring 1.2 cm in length and 0.6 cm in width (Figure 2). Histological examination revealed presence of lumina structure like, with squamous and respiratory epithelium lining, striated muscle in dermis and focal parakeratosis.

The final clinical diagnosis of this patient was CMCC. Her post-operative course was uneventful. At 3-month postoperative follow-up, she was doing well without cervical scar contracture and other complication.

Discussion

The CMCC was first reported in 1848 by von Luschka[8]. It was published in the textbook of pediatric surgery by Ombredanne[9] in 1949. Female is more predominant than male[1,4,10]. The CMCC can be identified after birth, but it may be misdiagnosed that often caused inadequate treatment and complications[6,7].

The CMCC may be a solitary malformation, but it may have associated anomalies, such as thyroglossal duct cyst, ectopic bronchogenic cyst, branchial cleft cyst, midline hemangioma, cleft lip and other abnormalities of mandible, tongue, sternum, absence of hyoid bone and thyroid cartilage[11-14]. Hirokawa[15] reported CMCC associated with congenital heart diseases and the most serious anomaly with ectopia cordis.

Different theories have been proposed on the embryological origin of the CMCC. Most investigators believe that the defect is the result of fusion failure of

the first and second branchial arches in the midline[10,12,15-17]. Mechanisms with relation to incomplete branchial fusion are vascular anomalies (ischemia, necrosis and scaring), increased pressure in the cervical area and absence of mesenchymal tissue in the cervical midline. Typical three anatomic parts of CMCC consist of skin tag at the superior end, linear vertical cleft in the middle part and small sinus tract at the inferior end. The superior skin tag may present with normal skin or thin stratified squamous epithelium with parakeratosis. The inferior sinus tract consists of pseudostratified columnar epithelium and dense collagen. Secretions may be noted from accessory salivary glands draining into the cleft[4,10].

The diagnosis of CMCC can be done by physical examination[1-5]. The lesion locates in the anterior surface of neck, between the mandible and upper part of the sternum. It is a longitudinal cleft extending downwards. The superior end covered by red or pink moist surface of atrophic epidermis without adnexal structure. The inferior end of the cleft usually has a sinus tract with mucous discharge and it involves gradually in the first month of infancy period. Sometimes, the cleft spontaneously heals and has scar formation. It develops complications of neck contraction, limitation of neck extension and movement, or presence of torticollis in some cases[6,7].

Complete surgical removal of CMCC is suggested to perform before two years of age because of the cosmetic outcome and avoiding of secondary complications. The surgical technique has been reported using a series of Z-plasty, to avoid a contracting linear scar[1,7,11-13,17]. From the literature review, some investigators advocated early surgical intervention before three months of age in order to prevent cosmetic deformity and complications, such as impairment of neck extension, mandibular deformity, torticollis and infection[7,11]. This patient in the present study was diagnosed and surgically treated at the first month of life. Outcome of the surgical excision was satisfactory without any complication.

Conclusion

CMCC is a rare congenital malformation of the anterior midline of neck. It is important for neonatologists and pediatric surgeons to recognize this rare anomaly in neonate. Typical characteristics consist of a linear vertical area of thin and erythematous mucosa at birth with a nipple-like projection in the upper end and a sinus tract at the lower end. Early definite diagnosis and surgical excision in the infancy period should be done in order to prevent secondary complications. The patient in this report was diagnosed and surgically treated within one month after birth. Her post-operative follow-up was good without any complication.

References

1. Farhadi R, Sahebpour AA, Ghasemi M. Congenital midline cervical cleft: can it be treated in newborn? Iran J Pediatr 2012;22:547-50.
2. Franzese C, Hayes JD, Nichols K. Congenital midline cervical cleft: a report of two cases. Ear Nose Throat J 2008;87(3):166-8.
3. Maschka DA, Clemons JE, Janis JF. Congenital midline cervical cleft: case report and review. Ann Otol Rhino Laryngol 1995;104(10 Pt 1):808-11.
4. Sinopidis X, Kourea HP, Panagidis A, Alexopoulos V, Tzifas S, Dimitriou G, et al. Case report of congenital midline cervical cleft: diagnosis, pathologic finding, and early stage treatment. Case Report in Pediatrics 2012(2012), Article ID 951040 (http://dx.doi.org/10.1155/2012/951040).
5. Puscas L. midline cervical cleft: review of an uncommon entity. Int J Pediatr 2015, Article ID 209418 (http://dx.doi.org/10.1155/2015/209418).
6. King J,Patel RV, Huddart SN. Congenital midline cervical cleft. J Pediatr Surgery Case Report 2013;1:99-101.
7. Ayache D, Dueroz V, Roger G, Garabedian EN. Midline cervical cleft. Int J Pediatr Otorhinolaryngol 1997;40 :189-93.
8. von Luschka H. Fistula colli congenital. Arch Physiol Heil 1984; 7:24-7.
9. Ombredanne L. Precis clinique et operatoire de chirugie infantile. 5th edition. Masson, Paris, France; 1949.
10. 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.
11. Genc A, Taneli C, Arslen OA, DaGlar Z, Mir E. Congenital midline cervical cleft : a rare embryo-pathogenic disorder. Eur J Plast Surg 2002;25:29-31.
12. Bajaj B, Dunaway D, Harley DEJ. Surgical approach for congenital midline cervical cleft. J Laryngol Otol 2004;118: 566-9.
13. Mlynarek A, Hagr A, Tewfix TL, Nguyen IT. Congenital midline cervical cleft : case report and review of literature : Int J Pediatr Otorhinolaryngol of 2003;67:1243-9.
14. Bergerin M, Sheft S, Myer C. Congenital midline cervical cleft. Pediatr Pathol 1989;9:731-9.
15. Hirokawa H, 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.
16. Warden C, Millar AJW. A rare congenital midline cervical cleft. South Afr J Surg 2010;48:98-9.
17. McInnes CW, Benson AD, Verchere CG, Ludeman JP, Arneja JS. Management of congenital midline cervical cleft. J Craniofac Surg 2012;23:36-8.

บทคัดย่อ **ความผิดปรกติแต่กำเนิดที่พบคล้ายร่องตรงกลางคอ: รายงานผู้ป่วยหนึ่งราย**

เยาวลักษณ์ คำนวน, พบ.

หน่วยกุมารศัลยศาสตร์ กลุ่มงานศัลยศาสตร์ โรงพยาบาลพุทธชินราช พิษณุโลก

ความผิดปกติแต่กำเนิดที่พบคล้ายร่องตรงกลางคอ เป็นความผิดปกติที่พบได้น้อยมากชนิดหนึ่ง ความผิดปกติชนิดนี้มักพบได้ตั้งแต่แรกเกิดด้วยลักษณะอาการที่มีผิวหนังตรงกลางคอด้านหน้านูนขึ้น เป็นติ่งคล้ายหัวนมอยู่ทางส่วนหัวของรอยโรคและมีรูเล็ก ๆ อยู่ที่ด้านล่างของร่องตรงกลางคอ โรคนี้สามารถได้รับการวินิจฉัยและรักษาโดยการผ่าตัดได้ตั้งแต่วัยทารก ผู้เขียนขอรายงานผู้ป่วยทารกเพศหญิงหนึ่งรายที่มีความผิดปรกติดังกล่าว และผ่าตัดได้สำเร็จตั้งแต่ผู้ป่วยอายุหนึ่งเดือน เพื่อเป็นการแลกเปลี่ยนประสบการณ์ของโรคที่หายากนี้