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Congenital midline cervical cleft: A retrospective case series of 8 children

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ABSTRACT

Objectives: Congenital midline cervical cleft is a rare developmental abnormality of the ventral neck of unclear etiology. It consists of a midline skin defect. This study reports a case series of 8 patients with congenital midline cervical cleft.
Methods: Retrospective review chart including all children referred with congenital midline cervical cleft over 5 years in tertiary center. The study was conducted to determine the presence of associated malformations, to specify the cleft pathology, to analyze the nature of associated cysts, and to discuss surgical procedure.
Results: Eight patients ranged from 3 days to 5 years. Two had an associated cervical midline cyst, 3 had a significant micrognatia. Pathological observations were in favor of a branchial origin. There was no recurrence of cervical contraction after a mean follow-up of 20 months.
Conclusion: Congenital midline cervical cleft is a rare and generally isolated congenital malformation. It does not require either extensive assessment or specific genetic. Described associated cysts might be part of the cleft and not bronchogenic or thyroglossal cysts. Early surgical excision reduces cervical contracture, but linear or *Z*-plasty closure is still debated.

1. Introduction

Congenital midline cervical cleft (CMCC) is a rare developmental abnormality of the ventral neck that consists in red atrophic skin at the ventral midline part of the neck, associated with a subcutaneous fibrous cord and a caudal sinus tract. The cleft can extend from the sub mental region to the suprasternal notch. It was first described by the German anatomist Luschka [1] in 1848. It remains an uncommon malformation with approximately 200 cases reported in literature and not much case series [2]. Embryologic development is not clearly established, but most authors suggest an impaired fusion of distal branchial arches on the median line of the neck [3].

The purpose of this study is to report a case series of patients with CMCC in order to discuss its clinical manifestation, malformative assessment, histopathology and surgical management. We also want to discuss the real origin of associated cyst that have been sometimes described.

2. Material and methods

After Institutional Review Board approval, a retrospective review chart including 8 consecutive patients referred for CMCC in a tertiary pediatric care center, from January 2007 to October 2013 was performed. Studied parameters included: age, sex, genetic background, size and characteristics of the cleft, associated malformations, type of surgery, pathology and scar evolution.

A complete cervico-facial examination was performed with clinical examination, flexible pharyngo-laryngoscopy, cervical ultrasound or cervical magnetic resonance imaging (MRI). General malformative assessment was not standardized.

Surgical procedure was performed under general anesthesia. A preoperative endoscopic examination of the pharynx, larynx and trachea was performed. Methylene blue was injected through a catheter into the sinus tract to guide the dissection. A vertical elliptic skin incision was then performed circumscribing the area of impaired atrophic skin. All pathologic tissues including skin, fibrous cord, nipple-like structure, sinus tract and associated cyst if

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Table 1
Patient characteristics.

Child	Sex	Genetic background	Age*	Cleft size†	Cyst	Malformation	Closure	Follow-up‡
1	M	Caucasian	70	14	Absent	Absent	*Z*-plasty	7
2	F	Caucasian	6	17	Absent	Absent	*V*-plasty	1
3	F	Caucasian	1	50	Present	Micrognatia	Straight	55
4	M	Caucasian	6	9	Present	Absent	Straight	45
5	F	Caucasian	6	21	Absent	Absent	*Z*-plasty	20
6	F	African	6	38	Absent	Absent	Double *Z*	13
7	M	Caucasian	7	50	Absent	Micrognatia/Atrophic kidney	Double *Z*	14
8	M	Caucasian	12	70	Absent	Micrognatia	Double *Z*	1

Abbreviations: M, male; F, female.
* Age at surgery in month.
† Cleft size related to mento sternal distance in %.
‡ Time of follow-up in month.

present were removed. Simple or double *Z*-plasty closure was proposed according to the length of the cleft. All removed tissues were sent for pathology. They were reviewed by a senior pathologist, looking for proof of thyroglossal or bronchogenic origin, researching specifically smooth muscle, cartilage of thyroid cells.

Follow-up was based on clinical assessment after ten days, at one month, six months and finally once a year after surgery to evaluate scar evolution and neck contracture.

3. Results

3.1. Patient characteristics (Table 1)

There were 4 boys and 4 girls. Seven of them were Caucasians and one patient was an African girl. Seven infants have been referred before 16 days after birth, only one at 5 years-old. All patients were born after uncomplicated pregnancy and delivery. There was no family history of congenital defects, and no consanguinity. One child had a family history of isolated micrognatia in both parents.

The length of skin defect was 34% of the mento sternal distance on average (SD ± 22%) of the distance sterno-mental distance in extension. None of them had a cleft extended to the mandible, inferior lip or tongue. All of the children presented a cephalic nipple-like structure, but only seven among them had a caudal sinus tract (Fig. 1).

No other facial cleft was found during physical examination. Three children presented a significant micrognatia at the time of the diagnosis, including the child known for having a family history.

Pharyngo-laryngo-tracheal examination was normal in all patients. Cervical ultrasound was performed on 5 children and cervical MRI on 2 children. Thyroid gland, hyoid bone, thyroid and cricoid cartilages were normal in all patients. A median cervical cyst (Fig. 2) was observed in 2 cases, located under the thyroid gland in both of them. Renal ultrasound was performed on three patients and one case of atrophic kidney was observed. Two patients have a heart ultrasound exam which was normal. A genetic study was performed and normal for the patient who presented atrophic kidney, micrognatia and familial history of micrognatia.

3.2. Treatment

Surgical excision was performed in all patients by the same surgeon under general anesthesia. Patients with micrognatia had no problem during or after intubation. Patients were 14 months old on average (SD ± 22 months) at the time of the surgery. Linear closure was performed in three cases and *Z*-plasty closure was performed in 5 cases including 3 double *Z*-plasties (Fig. 3). No complication was reported during procedure.

3.3. Follow-up

One patient had a delayed healing with minor necrosis at the angles of the scar. The infant of African origin had developed a

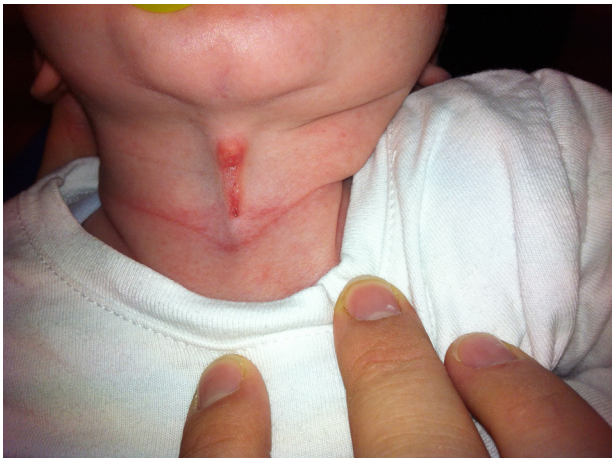

Fig. 1. Congenital midline cervical cleft with vertical atrophic skin defect, cephalic nipple-like protuberance, and caudal sinus tract.

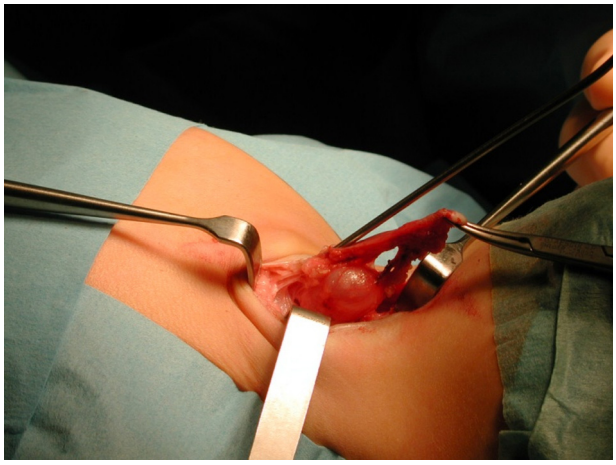

Fig. 2. Peroperative view of the excised cleft associated with a large cyst.