

## Case Report

# Congenital Midline Cervical Cleft

Francisco Aldo Rodrigues Júnior<sup>1\*</sup>, Isabel Marlúcia Lopes Moreira de Almeida<sup>2</sup>, Rogério de Araújo Medeiros<sup>3</sup>, Ivo Lima Viana<sup>3</sup>, Alexandre Gabriel Silva Rego<sup>4</sup>, Marina Nogueira Barbosa Rêgo<sup>4</sup>, and Bruno Pinheiro Falcão<sup>5</sup>

<sup>1</sup>Medical student from the State University of Piauí, Brazil

<sup>2</sup>Department of Neonatologist, Federal University of Piauí, Brazil

<sup>3</sup>Department of Pediatric Surgeon, Dona Evangelina Rosa Maternity, Brazil

<sup>4</sup>Medical student from the Federal University of Piauí, Brazil

<sup>5</sup>Department of Pediatric Surgeon, Federal University of Piauí, Brazil

**\*Corresponding author**

Francisco Aldo Rodrigues Júnior, Medical student from the State University of Piauí, Brazil, Tel: 5586981727986; Email: farj09@gmail.com

Submitted: 13 May 2018

Accepted: 27 June 2018

Published: 29 June 2018

ISSN: 2333-7095

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**OPEN ACCESS****Keywords**

- CMCC
- Diagnosis
- Imaging

**Abstract**

**Introduction:** Congenital Midline Cervical Cleft (CMCC) is a rare congenital malformation characterized by a classic presentation. A typical case of CMCC is reported, reiterating the importance of imaging for an accurate diagnosis and adequate surgical planning.

**Case presentation:** Newborn full term, at birth, a thin fusiform area was identified, located at the base of the anterior cervical region, in the midline, with a non-specific aspect, which may correspond to the congenital alteration. Doppler ultrasonography showed hypoechoic image, thin and apparently on blind bottom, measuring 0.8 x 0.2 cm. Nuclear magnetic resonance imaging identified a thin, low-signal area, located in the subcutaneous area, without areas of edema or associated fluid collections and no signs of extension to larynx / trachea, confirming the diagnosis of CMCC and excluding the presence of fistula.

**Conclusion:** The CMCC has its diagnosis classically established through physical examination, although - in some cases - it may be inconclusive. Ultrasound is already used as a diagnostic modality in relation to other cervical abnormalities, besides being fundamental for the surgical planning, since it shows the relation of the injury with the adjacent structures. Magnetic resonance imaging is a useful modality to demonstrate the extent of the cleft, to determine other associated anomalies, and to plan surgical treatment for repair. The relevance of imaging tests to define the extent of the congenital defect and exclusion of differential diagnoses, minimizing complications, is reiterated.

**ABBREVIATIONS**

CMCC: Congenital Midline Cervical Cleft

**INTRODUCTION**

The congenital Cervical Cleft of the Midline (CMCC) was first described in 1848 by Luschka [1], however it was Ombredanne in 1946 [2] who described it in detail in his book on pediatric surgery. The international literature reports at least 195 cases [3]. The researchers found that in a normal embryological development, the primary pharyngeal arches grow and close before the second arch, which closes before the third and soon. According to Maddalozzo, during the formation of the CMCC, before merging, the mesodermal tissue migrates between the arches and pushes the ectoderm outward to form the ventral sulcus [4]. Thus, several hypotheses have been postulated regarding the occurrence of CMCC from the embryological point of view: exteriorization of a remnant of the thyroglossal duct; increased pressure in the cervical area by the parietal pericardium in the developing embryo. However, the most widely accepted theory postulates a defective fusion of the pharyngeal arches, which impairs the fusion of the first or, more commonly, second pharyngeal arches in the midline, as well as an inappropriate interaction between the ectoderm and the mesoderm [5-7].

The typical presentation of the CMCC shows an erythematous skin band that occurs at any level between the sternum and the sternal manubrium, in addition to an ostium that can secrete mucous material [8,9]. A subcutaneous fibrous cord is always present and may represent the only clinical manifestation [10].

To avoid complications, such as limiting neck extension or impairment of mandibular growth, early intervention is recommended [11]. Image exams may be requested to differentiate CMCC from fistula lesions to the respiratory or digestive tract [5]. Magnetic resonance imaging is the best way to determine the extent of the lesion, its association with other abnormalities, and to define the pre-surgical planning. A CMCC is reported, whose diagnosis was suspected early by the physical examination and confirmed by the imaging tests, essential for the definition of the correct conduct.

**CASE PRESENTATION**

Full term newborn, cesarean delivery, obstetric indication for cephalo pelvic disproportion, gestational age of 38 weeks calculated by the Capurro method, weighing 3458 grams, with APGAR 9 and 10 in the first and fifth minutes, respectively. No maternal history of smoking, alcohol or illicit drugs. At birth, it was observed a thin fusiform area, located at the base of the anterior cervical region, in the midline, with a non-specific

aspect, which may correspond to a congenital alteration. Physical examination revealed a median cervical lesion, with an orifice close to the sterna furcula, hyperemic and secretive (Figure 1).

For diagnostic definition, investigation was initiated with imaging examinations, in order to exclude a communication between the external orifice and some cervical system. Doppler ultrasonography showed a thin, hypoechoic and apparently on a blind bottom, image, measuring 0.8 x 0.2 cm (Figure 2). Nuclear magnetic resonance imaging identified a thin, low-signal area, located in the subcutaneous, without areas of edema or associated fluid collections and no signs of extension to the larynx / trachea (Figure 3), confirming the diagnosis of CMCC and excluding the presence of fistula.

The patient progressed well, accepting oral diet and with good weight gain. No choking or episodes of cough. Hospital discharge without patient return and surgical correction scheduled for 4 months of life. Resection of them embranous mucosal área was programmed, followed by zeta plasty, in order to allow a better cervical extension and ensure a good aesthetic and functional result.

## DISCUSSION

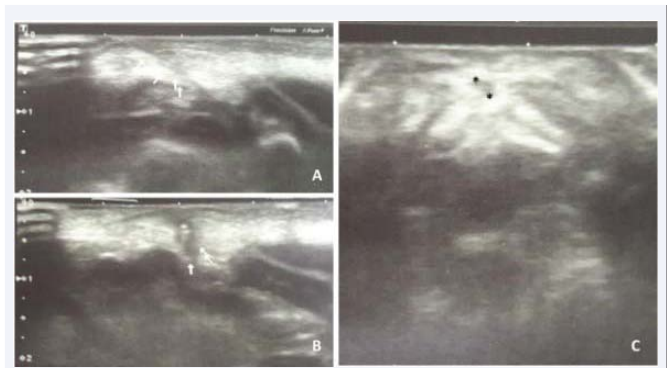
CMCC is a rare congenital anomaly, thought occur due to failure of fusion of the first and second pharyngeal arches during embryogenesis [5]. Its diagnosis has been established through physical examination [9], although the diagnosis of anomalies of the cervical region may escape this method [12].

Previous publications demonstrate consistent anatomical and pathological findings, yet few series with a greater number of patients and no complete review of the literature. One of the largest reported series includes 10 patients aged 4 to 27 years treated at the "Istituto di Chirurgia Plastica e Ricostruttiva" of the University of the United States of America Milano [14].

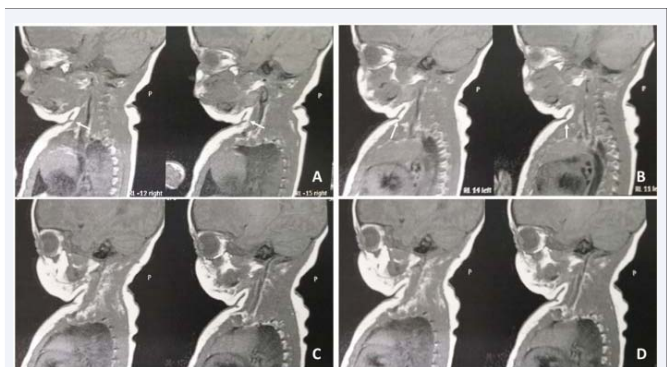
CMCC present sanincidence of 1-2% among congenital cervical malformations in several series, and it may be associated with absence of thyroid, midline hemangioma, ectopy cortices, cleft lip, absence of hypoid artery and additional cardiac abnormalities [13]. In the case reported, there were no other associated malformations.



**Figure 1** A thin fusiform area, located at the base of the anterior cervical region, in the midline, with or near the proximal surface of the sterile, hyperemic and secretory surface is observed in (A) and (B). The (\*) respond to the limits of the lesion, ending in a cranial fibrous cord.



**Figure 2** In (A) and (B), longitudinal section is observed and in (C) the transverse section of ultrasonography of the cervical region, showing a thin, hypoechoic image and apparently on a blind bottom, measuring 0.8 x 0.2 cm. The (white arrows) and (\*) identify the limits of the lesion to the method.



**Figure 3** In (A), (B), (C) and (D) sagittal T1-weighted sections of nuclear magnetic resonance are observed. One can clearly visualize thin, low signal area, measuring about 0.8 cm in extension, located in the subcutaneous on the anterior surface of the neck base, without areas of edema or associated net collections. Extension until near the is thmus of the thyroid and without signs of extension to the larynx / trachea. The (white arrows) point to the lesion in (A) and (B).

Ultrasound is already used as a diagnostic modality in relation to other cervical abnormalities. However, the total extent of the cervical tract and possible associated abnormalities are best demonstrated with nuclear magnetic resonance. It is also central to surgical planning as it demonstrates the relationship of the lesion to adjacent structures.

As the child grows or if the raisin in adequate repair of the lesion, the fibrous cord becomes more prominent and may lead to neck contractures, torticollis or limited extent of the cervical region [13]. Due to traction in them and able, a prominent bone maybe palpated and exostoses of the mandible or sternum can be seen on radiographs and / or resonances. Thus, an early diagnosis is essential to avoid such complications.

The treatment is always surgical. Although the exact timing and technique used are variable, most authors advocate the earliest possible intervention for best results. Z-incisions (orzetaplasty) are especially recommended in cases of late presentation in order to avoid the development of scarring and subsequent contractions of scarring [13]. Injury excision and

primary synthesis has been used with some success in patients with small fissures [7].

Thus, the congenital median cervical cleft is highlighted as a rare anomaly, with a small number of cases associated with other anomalies of the midline of the head and neck and the thorax. Magnetic resonance imaging is a useful modality to demonstrate the extent of the cleft, to determine other associated anomalies and to plan surgical treatment for repair.

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### Cite this article

Rodrigues Júnior FA, de Almeida IMLM, de Araújo Medeiros R, Viana IL, Silva Rego AG, et al. (2018) Congenital Midline Cervical Cleft. *J Radiol Radiat Ther* 6(1): 1080.