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Congenital midline cervical cleft

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ABSTRACT

Congenital midline cervical cleft (CMCC) is a very rare congenital malformation of the neck. Although its embryologic origin is uncertain, the most widely accepted embryologic defect is impaired midline fusion of the distal branchial arches. We present a case of female child with this anomaly and review of relevant literature. In literature review, CMCC has been described almost exclusively in Caucasian Female, the present case documents the occurrence of this developmental anomaly in the Egypt populations.

11. Introduction

Congenital midline cervical cleft (CMCC) is a rare anomaly of the anterior midline of the neck identified at birth, but may be overlooked or misdiagnosed with delayed surgical repair. Impaired neck extension, microgenia, exostosis, torticollis or infection are the most feared complications. Although, its embryologic origin has not been clearly established, the most widely accepted embryologic defect is impaired midline fusion of the distal branchial arches. Although rare (with about 50 cases published in the English literature) this entity does have a reliable presentation that should lead to rapid and accurate diagnosis. Complete surgical excision at an early age is essential. We present a case of female child with CMCC and she was treated with multiple Z-plasty with excellent cosmetic and functional results.

22. Case report

A 9-years old female child from upper Egypt, presented to us with congenital neck abnormality since birth. The patient had no airway or respiratory difficulties, she complaining of obvious fibrous cord in the anterior midline of the neck causing limited and painful neck extension, with vertical mid line neck cleft, there is a cyst protrusion (nipple like) at the proximal end of the cleft and a fistula opening on the skin at the distal end of the cleft. Her parents were healthy and non-consanguineous.

Clinically, the patient was found to have (1) midline vertical skin cleft (2) superior cyst protrusion (nipple like) (3) an inferior blind sinus, (4) a midline subcutaneous fibrous cord, extending from the chain to the sternum. On examination, the active and passive neck extension was limited and painful and the fibrous cord became more prominent with neck extension. Mucous discharge expressed from the inferior sinus. The length of the skin defect was about 6 cm (Figs. 1 and 2).

Neck ultrasound revealed an anechoic tubular structure in the subcutaneous planes of the anterior cervical region located at the midline, about 7.8 cm in length, coursing toward the suprasternal notch and ending abruptly, where the fistula opening on the skin surface was observed. There was no connection with other neck structures.

The patient was treated surgically. After general endotracheal anesthesia. Small vascular cannula was inserted through the fistula opening at the lower end of the cleft and small amount of methylene blue was injected to help in tracing the whole sinus tract till its end. Vertical elliptical incision was planned around the skin cleft including the upper cyst protrusion with multiple Z-plasty flap planning (Figs. 3 and 4). The cleft with the cyst protrusion and all underlying fibrotic tissue and the sinus tract were completely excised (Figs. 5 and 6), and the vertical elliptical wound was closed by multiple Z-plasty (Fig. 7). Postoperatively, no wound infection and wound healed without sequelae with excellent cosmoses.

Histopathological examination of the excised tissue showed stratified squamous epidermal covering overlying fibrotic dermis, sebaceous glands and the sinus tract was formed of large lymphatic spaces with endothelial lining, closely arranged separated by fibrofatty tissue with sinus formation, the proximal cyst protrusion shows normal epidermis with occasional bundles of skeletal muscle. Cleft was lined by stratified squamous epithelium without skin appendages.

33. Discussion

Luschka published the first case of CMCC in1848 under the description of “Congenital Fistula of the Neck”. Heusinger in 1864 and

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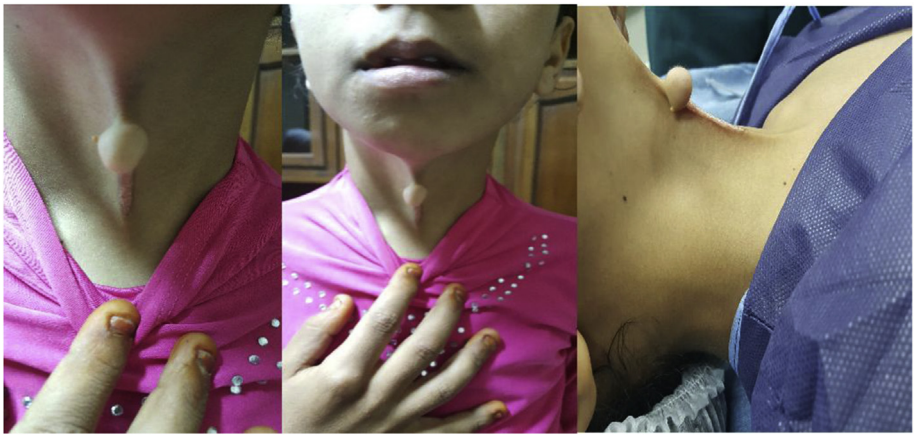

Fig. 1. Patient preoperative photo notes the tight fibrous cord at midline of the neck with cyst protrusion (nipple like).

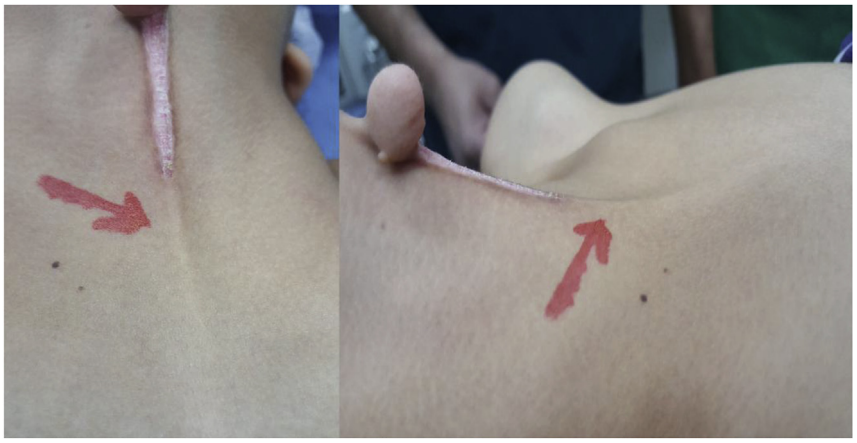

Fig. 2. The subcutaneous fibrous cord extending to the sternum (red arrow).

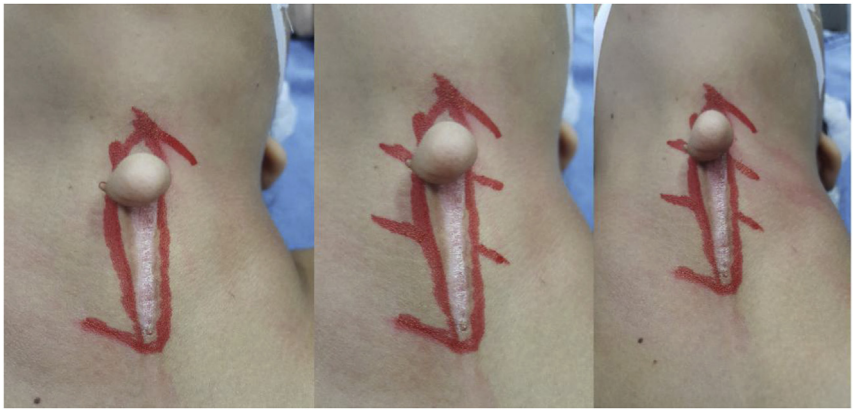

Fig. 3. Vertical elliptical skin incision around the skin cleft with planning of multiple Z- plasty flaps.

1865, published three reports of neck fistulas but one is more consistent with a bronchogenic cyst (barrel chest, cyanosis, and sinus tract that extends from the left anterior chest toward the lower neck) and the other two have fistula tracts/sinuses involving the lateral aspect of the neck. Barsky's in 1938 reported a case of CMCC in which the patient had a thick midline cord but no epithelial defect and Szenes' in 1922 reported a case in which the cleft in the neck extends inferiorly into the sternum [1–4]. The exact embryology of CMCC is unknown, however, it was explained as a midline fusion abnormality of the distal branchial arches during embryogenesis. It is commonly present as an isolated anomaly, however, two different entities were reported in the literature: midline cervical cleft with hypoplasia of the mandibular arch. Decreased or deficient cellular migration through the second arch (hyoid) results in isolated CMCC. More complicated anomaly presents with mandible or tongue cleft and absence of cervical support structures, are the result of defects in the first arch (mandibular). It occurs predominantly in Caucasian females. The genetic studies show mutation in the SIX 5 gene with detection of the pregnancy-associated plasma protein A, as potential candidates [5–8]. At birth, the external layer of the cleft may consist of a weeping, red membrane which then heals to produce cicatricial skin as the patient grows. The fibrous cord, which usually extends down to the pretracheal fascia, becomes more