Case Report

Congenital Midline Cervical Cleft – A Case Report on a Rare Anomaly of the Neck

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INTRODUCTION

Congenital midline cervical cleft is a rare and usually an isolated sporadic anomaly and according to a literature search, only 205 cases have been reported till 2014.[1] Although the embryological origin is unknown, the most accepted theory is impaired midline fusion of branchial arches, commonly the second arch along with improper interaction of ectoderm and mesoderm.[2] Complete surgical excision at an early age is recommended because the length of the lesion and fibrosis increases with age.[1,3]

CASE REPORT

An 8-year-old girl presented with lesion in the midline of the neck, noticed at birth. The lesion was painless with no dyspnea or dysphagia. She came mainly due to the cosmetic aspect and also had mild difficulty in extending the neck. She did not have any other obvious congenital anomalies or family history. The lesion was linear extending from below hyoid bone to 1.5 cm above suprasternal notch with a length of 5 cm and width of 0.5 cm. The lesion had a spherical skin tag of 1 cm diameter superiorly, that was soft, nontender, strip of wet hypopigmented region about 4 cm long and 0.4 cm wide and inferiorly a sinus with opening extending toward the suprasternal notch. The sinus was about 1 cm long on probing. Fibrous cord-like tissue was palpable beneath the lesion [Figure 1]. The lesion was not moving with protrusion of the tongue or deglutition. Hyoid was palpated normally and thyroid gland was seen normally moving on deglutition.

Ultrasound of the neck revealed normal thyroid gland, blind ending 1.5 cm long sinus in the subcutaneous plane, and there was no regional lymph node enlargement.

Vertical elliptical incision was made and complete surgical excision was done including underlying fibrous tissue and sinus in toto [Figure 2a]. Simple vertical closure of wound was done in two layers. Histopathology revealed the presence of soft tissue with plenty of adipocytes and few smooth muscle cells in skin tag, the red strip was lined by nonkeratinized stratified squamous epithelium, and sinus was lined by endothelium. The child had uneventful postoperative recovery. After 2 weeks, suture removal was done, and after 4 weeks, physiotherapy was initiated along with soft cervical collar. Physiotherapy for 6 months (extension and flexion of neck, and turning the neck to right and left ten times twice daily) was started along with nighttime use of soft cervical collar for 1 year to prevent contractures. The child was on regular follow-up. After 1 year, the scar had healed well and the child had no difficulty in extension of the neck [Figure 2b].

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**DISCUSSION**

This is a rare congenital anomaly of the neck. Older studies showed a female preponderance, but with more number of cases, this difference is slowly disappearing.[1] Most cases reported are sporadic with no associated anomalies or family history.[2] The typical lesion has a superior nipple like skin tag, a linear pink moist strip of atrophic epidermis without adnexal structures and a sinus which is the end point of vertical moist strip of epidermis.[1] Excision with single Z plasty for <2cm lesions and multiple Z plasties for longer lesions are recommended to prevent scar contractures.[3] Early excision in infancy is preferred to minimize contractures and for better cosmesis.[1]

Our patient had the classical clinical presentation with the sinus directed toward the sternum. We did a simple vertical closure as the surrounding skin was lax and the wound could be freely approximated in the center. As the child was 8 years, past the age of maximum vertical growth of neck (3–6 years), we did simple vertical closure. For an infant, Z plasty is preferred as the neck is yet to lengthen and extension might become difficult with growth due to scar contracture. But for an older child, we observed that simple vertical approximation, with meticulous closure of the wound in two layers, with 5-0 prolene subcuticular sutures for skin, gave satisfactory cosmesis without contracture. Physiotherapy with soft cervical collar aided good healing of the wound.

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**Conflicts of interest**

There are no conflicts of interest.

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