

# Congenital Midline Cervical Cleft: A Case Report

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## Citation

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## Abstract

Congenital midline cervical cleft is an uncommon developmental anomaly. We report a six-year-old male who presented with a midline neck sinus with a typical appearance of congenital midline cervical cleft. The embryology, presentation and management are briefly discussed.

## INTRODUCTION

Congenital midline cervical cleft is an uncommon developmental anomaly with a wide spectrum of presentation. Less than 100 cases are reported in the literature. We present a case of congenital midline cervical cleft with its typical presentation.<sup>1</sup>

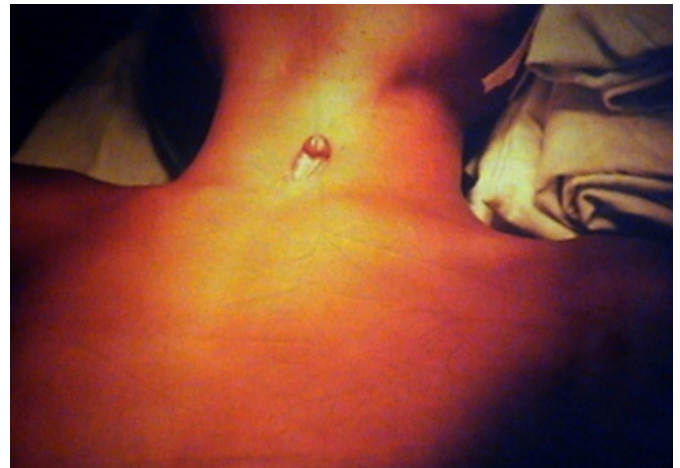
## CASE REPORT

A six-year-old male presented with a skin tag on the anterior aspect of the lower neck in the midline since birth. He had no history of discharge, swelling, change in the appearance of the lesion, trauma or surgery. The birth and the family history were insignificant. Examination revealed a skin tag of 1x1cm in size on the anterior aspect of the neck at the level of the cricoid cartilage in the midline. This tag was hooding an opening of 0.5x0.5cm lined by pink mucosa with a palpable subcutaneous cord running inferiorly for 4cm that could be probed (Fig.1, 2). The lesion did not move on deglutition or protrusion of the tongue and had no fixity to the underlying structures. Examination of the rest of the face, neck and oral cavity was normal; there was no other sinus or cleft.

The skiagrams of neck and chest were normal. At surgery, a sinus hooded by a skin tag with a subcutaneous tract superficial to platysma, surrounded by fibrous tissue and lined by mucosa, was excised and primary closure was achieved. Histopathological examination revealed a sinus lined by simple squamous epithelium with surrounding fibroplasia and few lymphocytes with no evidence of thyroid or thymic tissue.

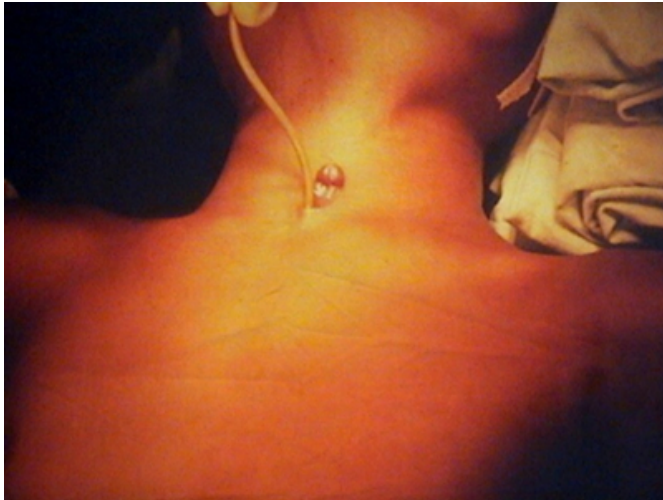
## Figure 1

Figure 1: Congenital midline cervical cleft: clinical photograph



### **Figure 2**

Figure 2: Congenital midline cervical cleft: tract running towards sternum in midline



### **DISCUSSION**

Congenital midline cervical cleft is an uncommon entity with less than 100 cases reported in literature and has been a descriptive exercise for the embryologists.<sup>1</sup> It is a sporadic condition, common in white females. It is characteristically seen in midline ventral neck with a cephalad skin tag, squamous epithelium lined mucosal surface and a caudal sinus with associated submucous fibrous cord and occasionally interwoven skeletal muscle, and has no relation with hyoid bone.<sup>3</sup> The spectrum of severity ranges from ventral cervical webs to mentosternal clefts leading to pterygium colli medianum with severe regional hypoplasia.<sup>2</sup> It is rarely associated with a bronchogenic cyst or with respiratory epithelium, hemangiomas and cardiac septal defects.<sup>2</sup> The differential diagnosis includes branchial, thyroglossal and chondrobranchial remnants but it is easily

differentiated by its typical presentation.

The embryological descriptions are discussed as failure of fusion of branchial arches in the midline considering its association with clefts of the lip, mandible and sternum, and rarely with thoracic ectopia cordis.<sup>4</sup> It may represent an ectopic first branchial arch derivative due to its common association with lower lip and tongue clefts and Tessier's type 30 craniofacial clefts.<sup>3</sup> Few reports propose the condition to be part of midline branchiogenic syndrome resulting from abnormal migration of cells derived from branchial arches.<sup>5</sup>

Management includes excision of cleft with reconstruction of the defects. The severity of regional hypoplasia decides the reconstructive armamentarium ranging from Z and V-Y plasty for simple webs to tissue expansion or myocutaneous flaps for severe regional hypoplasia and geniosternoplasty for mentosternal clefts.<sup>3,5</sup> This uncommon condition adds a page in the panacea of congenital neck lesions and is yet to be understood well.

### **References**

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