INTRODUCTION

The branchial apparatus anomalies are a heterogeneous group of disorders that arise from incomplete obliteration of branchial clefts and pouches during the end of the fourth week of gestation. Pharyngeal or brachial arches are first to appear at the fourth week of gestation. There are five branchial arches (I, II, III, IV and V) in humans; that later on lead to the formation of musculoskeletal and neurovascular structures of head and neck in humans. Branchial cleft cysts are congenital anomalies which usually express in the form of cystic mass in the neck. There are various manifestations secondary to abnormalities of branchial apparatus that can result in cyst e.g. sinus, fistula. Second branchial arch anomaly is the most common among all and accounts for almost all (95%) of the cases, followed by the first arch (1 - 2%). There are very few documented cases of the fourth branchial arch anomaly, especially along with mediastinal involvement.

The authors are reporting a girl child with posterior mediastinal branchial cleft.

CASE REPORT

A 2½ years old Kenyan girl presented at the Aga Khan University Hospital, Karachi with recurrent pneumonia and difficulty in swallowing. She had repeated episodes of coughing with fever since infancy and was treated as suspected pneumonia with reactive airway component inhaled bronchodilators and steroids and parenteral and oral antibiotics. Examination revealed vitally normal child with decreased left lower zone air entry with bilateral coarse crepitations. Septic workup including complete blood count and bacterial culture came normal, however, chest X-ray showed left lower lobe atelectasis (Figure 1a). Considering her deglutition history barium swallow was performed that showed mid-esophageal indentation pushing from the left side toward the right (Figure 1b). A possibility of mediastinal mass and/or vascular ring was suspected and CT chest was done with a nasogastric (NG) tube. The NG tube was placed to delineate the

POSTERIOR MEDIASTINAL BRANCHIAL CLEFT CYST: AN UNUSUAL SITE

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ABSTRACT

A two and a half year old Kenyan girl presented with recurrent chest infections and difficulty in swallowing. Her clinical and laboratory workup was suggestive of lower respiratory tract infection for which she received a course of antibiotics; however, she remains symptomatic after the management of her suspected diagnosis. Therefore, further radiological workup including a chest CT scan and barium study were performed. This showed a homogeneous mass surrounded by ascending and descending aorta in the posterior mediastinum that was compressing the middle esophagus. She had a posterolateral thoracotomy which revealed a cystic mass with smooth surfaces (5 x 5 cm) in the posterior mediastinum. Histopathology showed branchial cleft cyst predominately lined by stratified squamous epithelium, with lymphocytes predominance. A final diagnosis of posterior mediastinal branchial cleft cyst was made. She was discharged home and remained well at follow-up.


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Figure 1 (a,b,c,d): (a) Chest X-ray at the time of in-hospital admission showing left lower zone atelectasis (white arrow). (b) Barium swallow showing indentation of mid-part of esophagus (white arrow). (c) CT scan chest (mid sagittal view) showing a homogeneous mass surrounded by ascending and descending aorta in the posterior mediastinum (black arrow). (d) CT scan chest (coronal view) showing mass surrounded by major vessels with no calcification (white arrow).
esophagus from inside that showed a homogeneous mass at the level of carina surrounded by ascending and descending aorta in the posterior mediastinum with left lower lobe collapse with no intra-lesional calcification seen (Figures 1c and d).

A posterolateral thoracotomy was carried out which revealed a cystic mass with smooth surfaces (5 x 5 cm) in the posterior mediastinum attached with the esophagus, surrounded by ascending and descending aorta (Figure 2a). The mass was excised and sent for histopathology which revealed branchial cleft cyst predominately lined by stratified squamous epithelium, with lymphocytes predominance (Figure 2b).

Postsurgically, she was shifted to special care unit, recovered and discharged on seventh day of hospitalization with no sequel. She remained stable and without complications 6 months till last follow-up.

**DISCUSSION**

Pharyngeal arch anomalies arise from a failure of pharyngeal cleft obliteration during embryogenesis and can manifest themselves as cysts, sinus tracts, and fistulae. The type of lesion is determined by the presence of complete, partial, or no communication between endodermal (pouch) elements and ectodermal (cleft) elements. By the end of the fourth gestational week, there are four well-defined pairs of branchial arches and two additional rudimentary arches that are not visible on the surface of the embryo. These arches are covered externally by ectoderm and internally by endoderm with mesoderm in between. The mesoderm contains a dominant artery and nerve as well as cartilage rod and muscle. Arches are separated by clefts on the external endodermal side of the embryo and pouches on the internal endodermal side. Branchial anomalies are usually remnant of four main branchial arches and the related clefts fail to regress normally. Most branchial cleft anomalies involve the first and second cleft and pouch complexes. As far as third and fourth cleft anomalies are concerned only a few case reports are published. Branchial clefts give rise the structures of head and neck in humans.

This report describes a fourth branchial cleft anomaly with an unusual occurrence in the posterior mediastinum. Because of the rarity of fourth pharyngeal arch lesions, the diagnosis is often delayed. The remnant usually manifest in the form of cysts and sinuses tract formation at the region of neck and upper mediastinum. Historically, esophagogram was the diagnostic modality of choice for discovering sinuses arising from the pyriform region. The sensitivity of esophagogram was only 50% and may not lead to final conclusion about the disease; a possible explanation could be the occlusion of the tract opening by inflammation and tissue compression. Embryologically the cysts are located anterior to the aortic arch on either side. The tract hooks either the subclavian or the aortic arch, depending on the side, and ascends to loop over the hypoglossal nerve. Mostly it is on the left side i.e. 95 - 97%. In this case, it was also located on the left side surrounded by the ascending and descending aorta, with no communication in between.

Resection of fourth arch anomalies requires special understanding of the anatomy. Surgical excision is the mainstay of treatment as done in this case. Other modalities of treatment are chemo-cauterization of the tract with 40% tri-chloroacetic acid and primary mucosal coverage serves as a less invasive procedure although probably prone to recurrence. Newer methods involving electro-cauterization of the sinus tract have been used with good results.

Posterior mediastinum is a very unusual and unexpected presentation of branchial cleft cyst. One should consider branchial arch remnant presenting as cystic posterior mediastinal swelling once all common differentials are ruled out.

**REFERENCES**