Duplication of cervical oesophagus: A case report and review of literatures

M. Nazem[1], A. B. Amouee[1,2], M. Eidy[1], Ishfaq Abass Khan[1], H. A. Javed[1]

ABSTRACT

Foregut duplication is commonly found in the posterior mediastinum. 10-20% of these anomalies are associated with oesophageal duplication. It can occur in all parts of oesophageal length. Although duplication of cervical oesophagus has been previously reported, but a majority of them were found in thoracic oesophagus. Infants with oesophageal duplication usually manifested by respiratory distress or asymptomatic thoracic mass, casually, detected in X-ray. A 7-month-old infant weighing about 7.5 kg, with the signboard of respiratory distress, fever and nutritional intolerance was admitted to our hospital. Physical examination showed dehydration, stridor, tachypnoea, intercostal retraction and neck stiffness. Plain chest radiogram showed dubious cystic mass in the distal neck. Cervical and chest computed tomography scan showed neck cystic lesion disseminated to posterior mediastinum, probably propagated to the respiratory system. Cystic lesion connected to oesophagus was partially resected, oesophagus was repaired and remaining mucous of a cyst was removed, then gastrostomy tube was applied.

Although cervical oesophageal duplication cysts are rare, but they must be considered as one of the differential diagnoses of cervical mass with respiratory distress in infants.

Key words: Cyst, duplication of foregut, oesophageal duplication

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INTRODUCTION

Duplications can occur in all parts of the oesophageal length. Most of them are located in right thoracic and lower half segment of oesophagus.[1,2] Cysts of oesophageal duplication are rare and at least 10 cases are reported till now.[3] Almost all these cases were detected in early days of life, manifested with respiratory distress and can be life threatening. If in case, clinical signs revealed before mass detection, tracheal intubation and urgent surgical interventions are needed.[3,4]

Oesophageal duplication is divided into three types: cystic, tubular and diverticular form. Most of them are cystic. Tubular kinds in contrast with cystics belong to normal oesophagus.[1,5] The importance of this case in addition to rarity is unusual sign and symptoms manifested by fever, neck stiffness and respiratory distress. These clinical features can be misleading and treated as meningitis.

CASE REPORT

A 7 month infant weighing 7.5 kg, presented with the respiratory distress, fever (38.5 C^o or 101.3^{o}F), and nutritional intolerance. Physical examination revealed dehydration, feebleness, stridor, tachypnoea, intercostal retraction and neck stiffness. O_2 saturation was found to be 91% by pulse oximetry. She was previously hospitalized for 3 days due to seizure in another centre and treated by antibiotics and anticonvulsant without lumbar puncture.

At our hospital, patient's radiogram showed dubious cystic mass in the distal neck. Cervical and chest computed tomography scan showed cystic neck lesion disseminated to posterior mediastinum, probably propagated to the respiratory system. On physical exam no signs of swelling and tenderness were present in the neck.

Plain chest radiogram showed dubious cystic mass about 3 cm × 1 cm. Lateral cervical X-ray showed a cystic lesion with detached air-fluid level which caused anterior displacement of tracheal and oesophagus [Figure 1].

According to cervical and chest computed tomography, lesion disseminated to posterior mediastinum, probably propagated to the respiratory system [Figure 2]. The duplication was resected and the oesophagus was

[1]Isfahan University of Medical Sciences, Al-Zahra Hospital, Isfahan, Iran, [2]Rafsanjan University of Medical Sciences, Rafsanjan, Kerman Province, Iran.

Address for correspondence:
Ishfaq Abass Khan, Isfahan University of Medical Sciences, Isfahan, Iran.
E-mail: Ishfaqkhan38@gmail.com

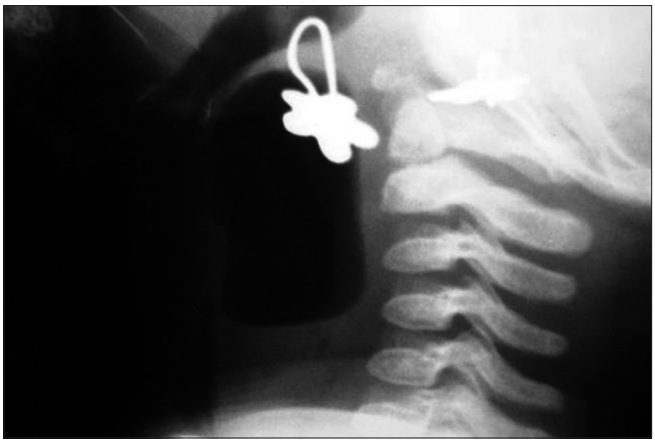


Figure 1: Lateral cervical X-ray which shows the cystic lesion with air fluid level in the posterior of oesophagus and trachea.

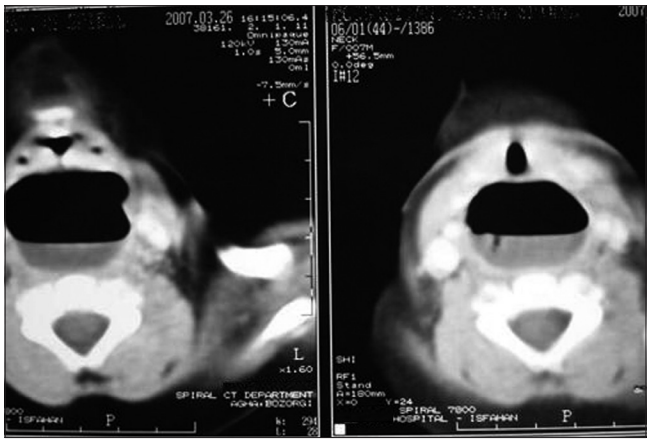


Figure 2: Cervical CT scan which shows the cystic lesion with air fluid level that seems connected to trachea.

repaired with interrupted absorbable sutures. A right thoracotomy was opened for mediastinal drainage and distal oesophageal duplication evaluation. A Stamm's gastrostomy was performed to allow the patient to eat. The excised oesophageal duplication was communicating with the oesophagus and consisted of a squamous cell epithelium and inflammatory cells infiltrated into the mucous membrane. The contents of the cyst were mucoid. Postoperatively, 3 months later the patient was doing well and able to eat a regular diet. The oesophagus-duplicated cyst was confirmed by histopathology.

DISCUSSION

Embryologically, duplications result from a defect in the tubulation (vacuolization) of the oesophagus, normally occurring in the sixth gestational week.[2,3] As the foregut epithelium develops, it elongates, develops a lumen and undergoes dextro-rotations. Thus, the majority of oesophageal duplications occur distally and on the right. In the present case the oesophageal duplication was proximal to the cervical part and on the left.

The duplication of the oesophagus may be of three forms: (1) a cystic form[6] that may or may not communicate with the oesophageal lumen; (2) a tubular form[7,8] or (3) a diverticular form.[9,10] In one study,[11] only six tubular duplications of 44 oesophageal duplications were identified. In another review of alimentary tract duplications,[12] 22% were intrathoracic, and all were cystic rather than tubular.

The duplication of the hollow structures of the digestive tract manifest three essential points: (1) they are in intimate contact with the alimentary tract; (2) they are lined by a mucous membrane similar to other such membranes at the same level of the digestive tract and (3) they are provided with a smooth muscle coat, though an oesophageal duplication without muscle fibres has been reported.[13] In the present case, the wall between the cyst and oesophagus was complete with a smooth muscle layer.

Most tubular duplications were reported in late childhood or adults, comparatively asymptomatic and commonly connected to oesophagus,[5] but in our patient symptoms appeared during the infantile period.

It often appears in form of soft tissue mass in posterior mediastinum and exerts pressure on oesophagus. Chest and abdominal CT-scan or MRI is indicated, because some of mediastinal duplication maybe continuous under the diaphragm ultrasonography is recommended as a screening method for such cases.[2] Oesophagogram also helps in diagnosis.[2] Mentionable paraclinic tests used in our patient were: chest roentgenogram, lateral cervical X-Ray, cervical and chest CT-scan.

Oesophageal duplication may be present with some anomalies such as small intestinal duplication, oesophageal atresia distal to duplication site, tracheoesophageal fistula, scoliosis, hemivertebra, adhesion of spine and anterior spina bifida.[14,15]

Because of caudal foregut and notochord cephalic development, spine defects basically are above the mediastinal cyst.[15] These are cysts found incidentally by chest roentgenogram in asymptomatic patients.[14] Finally, symptom appears in $\frac{2}{3}$ of cases when complications like infection, mass effects, bleeding, neoplasm and oesophageal or respiratory obstruction causing difficulties for patient.[14,15] Infants affected by a cervical oesophageal duplication cyst,

predominately had respiratory distress before detection of mass complications. These patients might confess malnutrition due to rapid growth of the cyst secondary to infection, bleeding and discharge to the cyst.[3]

Differential diagnoses for cervical cysts include lymphatic malformation, airway cyst, bronchial cyst or thyroglossal duct cyst sometimes present in the lateral of the neck.[3] Preferred treatment is complete resection of the cyst. In this way prognosis will be excellent.[3,15] If complete excision is not possible, mucosa should be removed and subsequently the cyst gets dissolve.[3,15]

CONCLUSIONS

Although cervical oesophageal duplication cysts are rare, but they must be considered as one of the differential diagnoses of cervical mass with respiratory distress in infants.

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