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Role of Imaging in the Diagnosis of an Enteric Duplication Cyst of the Esophagus: A Case Report

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Abstract **Case Report**

Aims: Show the importance of imaging in the diagnosis of enteric cysts of the esophagus and the main differences with other mediastinal cystic lesions. ***Presentation of case:*** We report a case of an enteric cyst of the esophagus, in a 25 year old patient, with dyspnea on exertion. X-ray shows an oval opacity in the posterior mediastinum and the CT scan and MRI show a cystic mass of the posterior mediastinum, presenting intimate contact with the esophagus, with discreet wall contrast enhancement. ***Discussion and Conclusion:*** Enteric duplication cysts are rare congenital malformations formed during the embryonic development of the digestive tract. They are usually detected prenatally or in the first years of life. Their size, location, type and presence of complications produce a varied clinical presentation and different imaging findings. They most frequently occur in the small intestine, particularly the ileum, but can occur anywhere along the gastrointestinal tract. In the case of an enteric cyst of the esophagus, the first imaging method for diagnosis is the chest x-ray because of the breathing difficulty patients can experience due to compression of the airway. Magnetic resonance (MR) and computed tomography (CT) can help the radiologist in the diagnosis and the surgeon by evaluating more precisely the mass measurements and describing its anatomical relationship with the vessels and surrounding organs. Confirmation remains histological.

Keywords: Enteric cyst, esophagus, mediastinum.

INTRODUCTION

Gastrointestinal tract duplication cysts are rare congenital gastrointestinal malformation in young patients and adults. They consist of foregut, small bowel, and large bowel duplication cysts.

Of note, 50-70% of foregut duplication cysts are enterogenous while 7-15% of them are bronchogenic. Foregut duplication cysts constitute 6-15% of primary mediastinal masses. Gastrointestinal tract duplication cysts most commonly occur in the ileum, esophagus, and colon. They may be contained within the gastrointestinal tract wall or extrinsic to it. Duplication cysts can also be cystic (80%) or tubular (20%).

Ultrasound is the method of choice to diagnose gastrointestinal enteric cysts. CT and MR imaging can be required in esophageal or rectal enteric duplication cysts (EDCs) for planning complicated surgical approach.

We report a case of an esophageal enteric cyst in a 25 year old male patient with respiratory symptoms, through which we will focus on the radiological aspect of this type of tumors and stress the importance of the CT scan and MRI in differential diagnosis with other mediastinal cystic masses.

PRESENTATION OF CASE

Our patient is a 25 year old male, with no pathological history. He presented with dyspnea on exertion. The clinical examination was without abnormality. His chest x-ray showed a mediastinal oval mass of hydric tonality, lateralized to the right (Figure 1). This motivated the realization of a chest CT scan that showed an oval well limited cystic mass in the posterior mediastinum, with thin septations. This mass presented intimate contact with the esophagus, with discreet wall enhancement, and measured 68x36x35 mm (Figure 2). A complementary MRI scan was perfomed on a Siemens 1.5 Tesla machine, and showed a well-limited ovoid mass, in T1-weighted hyposignal,

T2-weighted hypersignal, with the same septations and wall enhancement revealed by the CT scan. The MRI did not show any associated vertebral or medullary anomaly (Figure 3). The radiological images were in favour of an esophageal EDC, which was confirmed histologically after surgical treatment.

DISCUSSION

Duplication cysts, also known as alimentary tract duplications, are congenital lesions of the gastrointestinal tract. Although the exact cause still eludes us, multiple theories have been put forward to explain its occurrence.

The incidence of alimentary tract duplications is 1 in 4500 births. A slight male preponderance has been observed with these cysts. They are mainly detected in childhood, with the majority of duplications symptomatic within the first 2 years of life. However, late presentation in adulthood is also seen.

Endoscopic ultrasound (EUS) is the diagnostic tool of choice to investigate duplication cysts since it can distinguish between solid and cystic lesions. EUS can also establish cyst location relative to surrounding tissues. EUS shows duplication cysts as anechoic, homogenous lesions with regular margins arising from the submucosal layer or extrinsic to the gut wall, although a hypoechoic echo pattern can also be seen with a duplication cyst. On EUS, duplication cyst walls usually consists of 3-5 layers and the internal contents may be anechoic or hypoechoic. Duplication cysts may contain thick mucinous material, septations, fluid levels, debris and they may also contain detached ciliary tufts which could be diagnostic. In addition, duplication cysts can have peristalsis that appears as ring contractions with a concentric contraction of the cystic wall. Peristalsis in a juxta-enteric cyst is specific for a duplication cyst and can be a diagnostic feature.

Transesophageal ultrasound (TEE) is a great imaging method used for the diagnosis of these lesions, but it is not readily available at all centers, especially in developing countries like ours.

Ultrasound post-natal diagnosis of EDCs includes the same signs as postnatal cyst: the double–wall sign and the presence of peristalsis. However, on the prenatal US, the “double wall” is not always seen or can be partial, and it may require the differential diagnosis with other cystic lesions such as mesenteric, omental, ovarian and choledochal cysts. If it is possible to demonstrate the presence of peristalsis in the cyst wall, an intestinal origin is probed.

MR imaging is suggested to have a supplemental value in the assessment of fetal abdominal cysts.

CT is not typically performed for evaluation of EDCs due to radiation. CT may depict the location and extension of the cyst, as well as complications, the associated anomalies and anatomical relationship with surrounding structures. At CT, an EDC manifests as a cystic mass with a thin and slightly enhancing wall adjacent to the gastrointestinal wall. A high attenuation inside the cyst may be seen due to haemorrhage or proteinaceous material. A thick enhancing wall, air bubbles inside and cyst-surrounding inflammation may indicate an EDC complicated by infection.

Like CT, MR is not routinely used as a diagnostic method for EDCs, especially due to sedation requirement. On MR imaging, most duplications have low signal intensity on T1-weighed images and very high intensity on T2-weighted images. Both CT and MR play a major role prior to surgery in establishing the relationship between the cyst and its adjacent structures, and in locations where US presents a limited use, particularly in esophageal and rectal duplications.

Enteric cysts can be a problem of differential diagnosis with some bronchogenic cysts. However, bronchogenic cysts can be found in the middle mediastinum as well as in the posterior mediastinum, and there is generally no enhancement of their wall on imaging after injection of contrast product.

Other intrathoracic cysts, including the pericardial cyst and the thymic cyst, may be considered as close differentials of duplication cysts. However, these are located in the middle and the anterior mediastinum, respectively, unlike the duplication cyst, which is located in the posterior mediastinum.

The definitive treatment for a duplication cyst is surgery. The surgical approach can be open (laparotomy or thoracotomy) or minimally-invasive (laparoscopic or thoracoscopic) and depends on the expertise of the surgeon.

Early surgery is recommended in all the symptomatic cases. Intrathoracic esophageal duplication cysts: These require a posterolateral thoracotomy and can be removed by simple excision. In large cysts, decompression of the cyst may help in the better dissection.

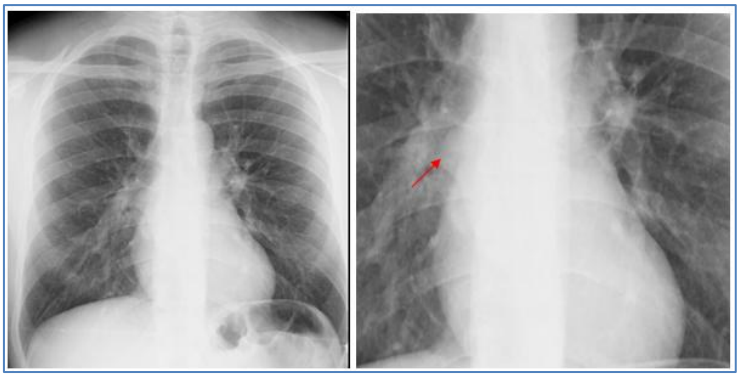

Fig-1: Chest x-ray showing a mediastinal oval mass of hydric tonality (arrow)

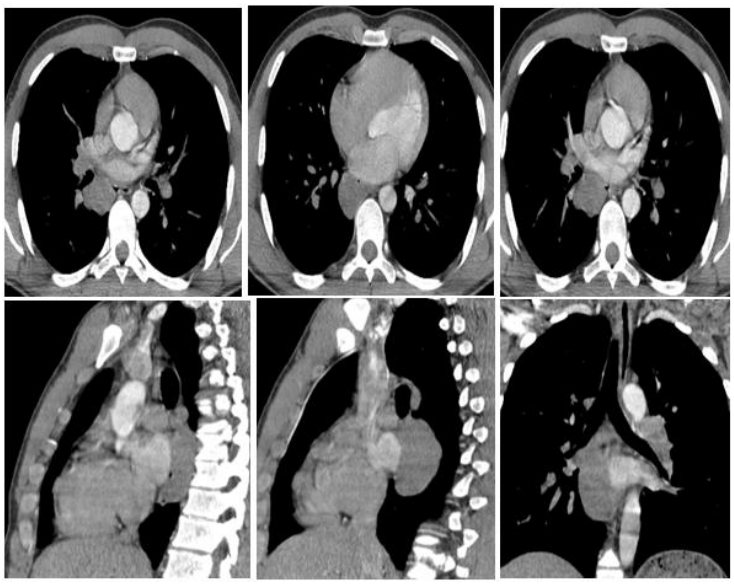

Fig-2 : Chest CT scan in axial, sagittal and coronal section showing an oval well limited cystic mass in the posterior mediastinum, with thin septations, presenting intimate contact with the esophagus, with discreet wall enhancement

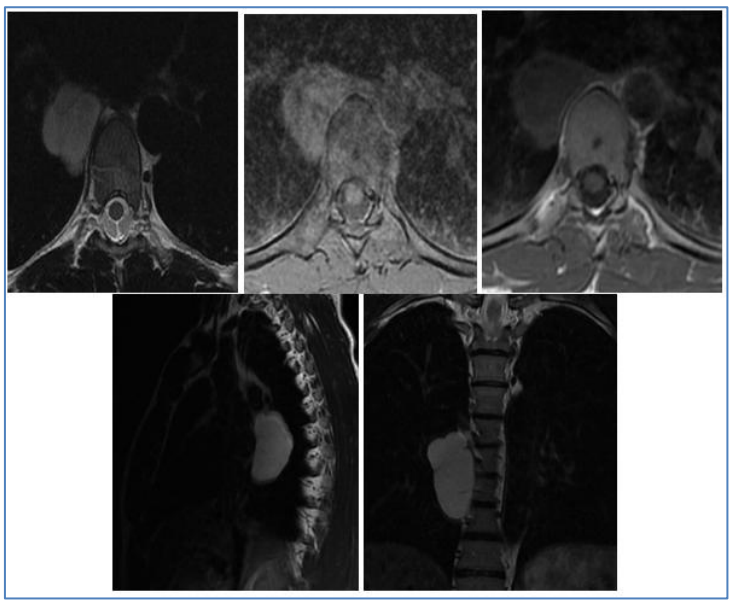

Fig-3: Thoracic spine MRI in axial, sagittal and coronal section showing the posterior mediastinum mass, with thin septations, discreet wall enhancement and no associated vertebral or medullary anomaly

CONCLUSION

Duplication cysts are rare congenital anomalies of the alimentary tract. Due to the varied clinical presentations, radiological evaluation is required for its diagnosis. The perfect knowledge of the characteristics of these cysts on ultrasound, CT scan and MRI should allow early diagnosis and better management.

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