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Case Report

Giant anterior mature mediastinal teratoma with gastrointestinal tract organoid differentiation: A rare presentation

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ABSTRACT

A teratoma is a tumour with tissue or organ components resembling normal derivatives of more than one germ layer.[1] Anterior mediastinal teratoma with organoid structures is a rare presentation of germ cell tumour. We report a case of such a rare giant tumour (30 × 15 cm) in a young adult male of 18 years who presented with right-sided chest pain of 3 years duration. Surgical removal is the definitive treatment of a mature anterior mediastinal teratoma. The patient was operated successfully with complete removal of the tumour. As superior vena cava (SVC) was encased in the tumour wall causing SVC syndrome, partial resection of the pericardium was required to free SVC from the tumour. The defect in the Pericardium was repaired to prevent herniation of heart. In the Post-operative period, patient developed recurrent pneumothorax of the left side, which was successfully treated by intercostal tube drainage and pleurodesis. Post resection histopathology examination confirmed mature teratoma with representation from all the three germ layers including organoid differentiation of gastrointestinal tract.

1. Introduction

Most common extra gonadal site of germ cell tumour is the anterior mediastinum. Benign teratoma of mediastinum accounts for 3–12% of mediastinal tumours with approximately equal incidence in both genders.[1] A mature teratoma is composed of several foci of mature tissue derived from at least two out of three embryonic germ layers (ectoderm, endoderm, mesoderm) and presence of all the three germ layers is rare.[2] The tumour is composed of tissues which are foreign to the organ or anatomic site in which they arise. Usually tissue arising from different germ layer has no correlation in a mature teratoma. The case reported has a very large mature teratoma (30 × 15 cm) with fully developed gastrointestinal organoid differentiation manifesting as multiple intestinal loops (cystic component of the tumour), which was confirmed by histopathology. Organoid differentiation of anterior mediastinal teratoma has been reported only rarely in English literature, although ovary and sacrococcygeal mature teratoma has been occasionally reported to contain different degree of organoid differentiation.[3–5] This teratoma was occupying most of the right hemithorax, anterior and middle mediastinum, extending up to left hemi-thorax. Due to its massive size, pressure effect was clearly visible on right lung, which was compressed; into a small non-functional tissue mass; all the great vessels were compressed, particularly superior vena cava, which was embedded in the tumour raising its pressure to 30 mmHg. The heart and mediastinum were shifted to the left. Surgical removal of teratoma is the only definitive treatment, with no role of chemoradiation.[1] Large lateral defect in the pericardium, which was required to isolate SVC from tumour mass in this case, was repaired to prevent potential herniation of the heart. Recurrent tension pneumothorax is an uncommon complication following large mediastinal teratoma removal.

2. Case report

An 18-year-old boy presented with complaints of right-sided chest heaviness, pain and exertional dyspnoea for last one year. There was no history of cough, fever, weight loss or haemoptysis. Physical examination revealed a bulge in the right hemi thorax with dilated neck veins. Jugular venous pressure was markedly raised. Trachea was shifted to left on palpation. On auscultation, there was no air entry on right side, middle and basal zone; however, breath sounds were diminished in the apex. Breath

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sounds were normal in the left hemithorax. Other system examination, including external genitalia was normal.

Chest X-ray revealed a well circumscribed homogenously opacified mass lesion occupying right hemithorax along with anterior mediastinum, extending up to left hemithorax with near total atelectasis of right lung. Trachea was deviated to left side with normal patency of left main bronchus with multiple area of calcification in the tumour mass (Fig. 1a). Computed tomography scan (CECT) chest revealed a large heterogeneous solid-cystic multiseptated and lobulated mass lesion (14.5 × 10.4 × 19.9 cm) containing areas of soft tissue and fatty attenuation, multiple coarse calcifications and calcified septal thickening in right hemithorax and anterior mediastinum (Fig. 2a–c). The giant mass caused significant contralateral mediastinal shift. Superior vena cava was encased by the mass in its upper portion causing partial SVC obstruction. Right pulmonary artery and their branches and pulmonary vein were compressed within the collapsed lung. Aorta was displaced leftward by the mass. Right lung appeared compressed and collapsed with some amount of aerated parenchyma noted in right apical region. Trachea and oesophagus were normal. Ultra sound examination of abdomen and external genitalia did not reveal any abnormality. All laboratory investigations along with tumour markers such as α-fetoprotein (AFP) and β-human chorionic gonadotropins (β-HCG) were normal. A CT guided fine needle aspiration cytology was suggestive of benign teratoma.

A primary median sternotomy was carried out, which provided excellent exposure of the tumour, and also allowed safe dissection of all mediastinal structures adherent to the tumour. The tumour was occupying most of the right thoracic cavity, anterior mediastinum and reaching up to medial one third of left thoracic cavity. A small incision was given on the tumour to allow the suction tip catheter to enter the cavity over a purse string suture. Copious pultaceous liquid material was sucked out from the tumour. The tumour became markedly decompressed thus facilitating easy dissection and removal. It was adherent to right hilar structures, SVC, pericardium, and diaphragm. Cheesy material along with bone, teeth and bunch of hairs and multiple intestinal loops like structures were removed from within and the tumour was resected out completely using sharp and blunt dissection. Tumour wall was dissected meticulously all around its attachment except pericardium. The plane of dissection between tumour and pericardium was not defined so a small portion of the right-sided pericardium was excised to complete the removal. The tumour had multiple septated cavities, and in the lower portion of the tumour, multiple cystic loops resembling gastrointestinal tract were seen (Fig. 3). Right lung, which was almost totally collapsed at the beginning, started to expand with positive pressure ventilation after surgery. Once the mass was removed, the heart herniated through the pericardial defect into the right hemithorax (large space due to collapsed lung) causing significant hypotension.

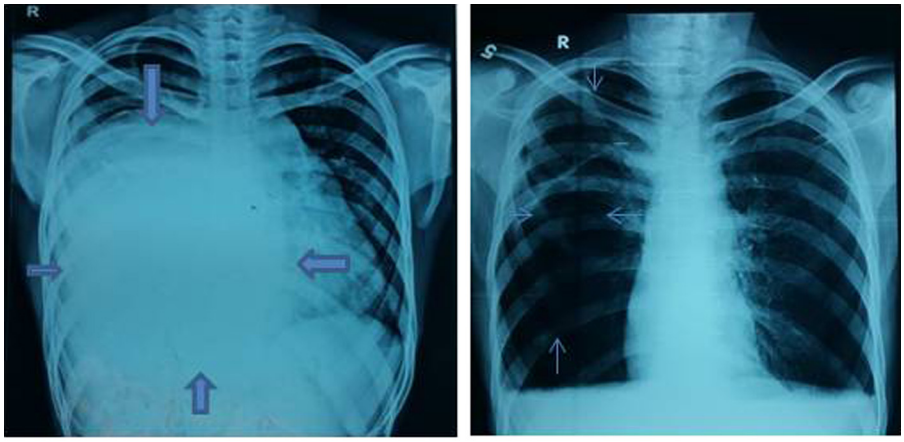

Fig. 1. (a) Pre op CXR showing large opacified lesion in anterior mediastinum, occupying most op the right hemithorax with contralateral mediastinal shift (block arrows). (b) Post op CXR showing complete removal of mass with expanded lung (arrow heads).

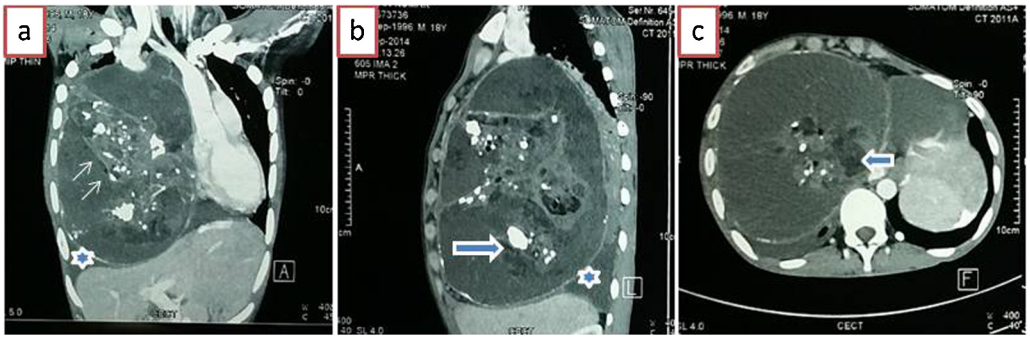

Fig. 2. (a) Coronal, (b) lateral and (c) horizontal cut section of contrast enhanced high resolution computed tomography chest. A large heterogenous solid cystic (black arrow) multiseptated (thin arrow head) lobulated mass lesion (14.5 × 10.4 × 19.9 cm) containing areas of soft tissues and fatty attenuation with multiple coarse calcified septal thickening (thin arrow head) is noted in right hemithorax extending from anterior mediastinum with significant contralateral mediastinal shift (a). Thin walled well-defined capsule with calcification (star) is also noted. Superior vena cava is encased in the mass but luminal patency seen. Final diagnosis anterior mediastinal mature teratoma.

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