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Case Report

Mature cystic teratoma of mediastinum compressing the right atrium in a child: A rare case report

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الملخص

الأورام المسخية (التيراتومات) أو أورام الخلايا الجنسية هي أورام مثيرة للاهتمام بسبب أصلها الغامض، ومظهرها المجهري الغريب وسلوكها غير المتوقع في بعض الأحيان. والأورام المنصفية المسخية أورام غير شائعة لدى الأطفال. وهي أورام بطيئة النمو يتم تشخيصها صدفة عند مرضى لا تظهر عليهم أعراض. ومعظم الأعراض إن وجدت، تكون ذات علاقة بأثر ضغط الكتلة على الأنسجة المجاورة مثل ألم الصدر، والسعال، وضيق التنفس وصعوبة البلع. نقدم في هذه الورقة تقرير حالة طفل ذكر يبلغ من العمر ٥ سنوات، قَدِم بألم في منطقة القص، والربع العلوي الأيسر مع تقيّؤ بدون ضيقة تنفسية. تبين أن المريض لديه كتلة في المنصف الأمامي ضاغطة على الأذين الأيمن، وتم تشخيصها بالفحص النسيجي على أنها ورم مسخي كيسي ناضج في المنصف. وتم علاج المريض بنجاح بالاستئصال الجراحي الكامل للورم.

الكلمات المفتاحية: تيراتوما؛ الأورام المسخية؛ المنصف الأمامي؛ الأطفال؛ الفحص النسيجي؛ العلاج الجراحي

Abstract

Teratomas or Germ cell tumours (GCTs) are interesting because of their obscure origin, bizarre microscopic appearance and unpredictable behaviour. Mediastinal teratoma is a slowly growing and rare tumour found in children that is diagnosed incidentally in asymptomatic patients. Most of the symptoms are related to mass compression effects such as chest pain, cough, respiratory distress and dysphagia. We report a 5-year-old male child who presented with a history of foreign body ingestion, sternal and left upper quadrant pain and vomiting without respiratory distress. The patient was incidentally found to have an anterior mediastinal mass compressing the right atrium and was diagnosed by histopathological examination as having a mature cystic teratoma of the mediastinum. The patient was successfully treated by the surgical resection of the tumour.

Keywords: Anterior mediastinum; Children; Histopathology; Surgical treatment; Teratoma

Introduction

The word teratoma is derived from Greek words "terato," meaning monster, and "onkoma," meaning swelling. A teratoma is a germ cell tumour derived from pluripotent cells and made up of elements of different types of tissue from one or more of the three germ cell layers.[1] The incidence of teratoma worldwide is approximately 1 in 4000 live births.[2] The most common reported sites are sacrococcygeal (40%), ovary (25%), testicle (12%), brain (5%) and others including neck and mediastinum (18%).[2]

Though teratomas are benign tumours, they can turn malignant.[3] A paediatrician should always remember the significance of diagnosing such teratomas and be vigilant about treating the condition promptly.[4] A delay of the

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diagnosis and treatment can lead to worsening of the symptoms of the patient and may be fatal.[4]

In this case, we share our experience with the diagnosis and management of a 5-year-old boy who was found incidentally to have a mediastinal mass that turned out to be a mature teratoma.

Case report

A 5-year-old male child presented to the emergency department with foreign body ingestion and had a history of sternum and left upper quadrant pain and vomiting. There was no history of cough, dyspnea, dysphagia or weight loss.

A chest X-ray showed a foreign body at the middle of the oesophagus (Figure 1).

A foreign body in the form of a battery was removed by an upper GI endoscopy, which revealed burn and ulceration of the oesophagus (Figure 2A, B).

A chest X-ray post endoscopy showed an irregular right cardiac shadow with a bulging right side of the heart (Figure 3).

All chest X-rays since admission were reviewed and revealed the presence of an abnormal cardiac shadow.

On examination, the patient had a café au lait spot on his trunk (Figure 4A–C). His complete blood count revealed a white blood cell count of 11.100/mm^3, a platelet count of 691.000/mm^3, and a haemoglobin level of 10.9/mm^3, and a peripheral smear showed lymphocytosis and monocytosis with few reactive and atypical lymphocytes.

Lactate dehydrogenase (LDH) 200 U/L, beta-human chorionic gonadotropin (B-HCG) 0.1 mIU/ml, alpha fetoprotein (AFP) 0.99 IU/ml, Erythrocyte sedimentation rate (ESR) 44 mm/h, Serum ferritin 90.69 ng/ml.

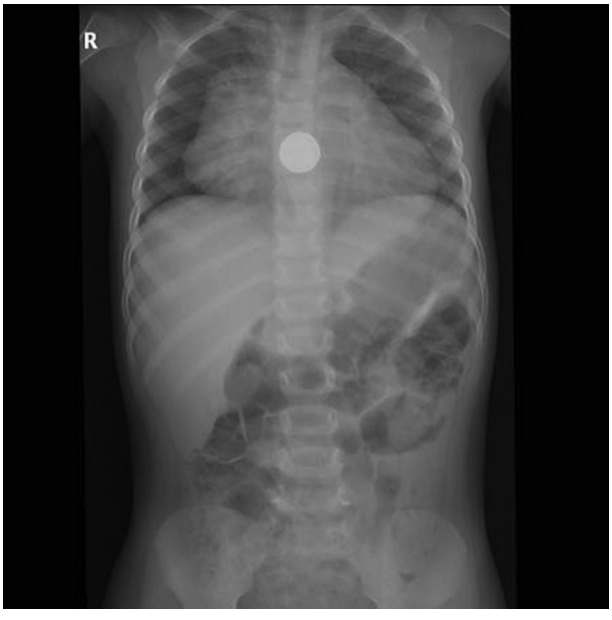

Figure 1: X-ray chest AP view showed a foreign body at the middle of the oesophagus.

Transthoracic echocardiography was performed, showing an echogenic mass collected at the right side of the heart extended to the anterior side and slightly compressing the right atrium (Figure 5).

A Computed Tomography of the chest (CT) was advised, and it revealed a well-defined, large cystic mass lesion noted at the right anterior mediastinum. An anterior cystic mediastinal mass with a fatty component likely arising from the right thymic lobe extended downward, reaching the right cardiophrenic angle and measuring 5.2 × 4.2 × 6.2 cm in AP, transverse and craniocaudal dimensions, respectively. The mass showed some septations with a large lobulated fat density structure within the cyst measuring 1.1 × 1.9 × 1.6 cm with adjustments for a small enhancing soft tissue component near the base of this fatty mass along with tiny multiple wall classifications. The mass is causing mass effect on the right side of the heart and mediastinal structures. No abnormal lymphadenopathy was present (Figure 6A–D). The patient was operated via right thoracotomy incision, and the mass was mobile on palpation but adherent with the lung thymus and the right side of the pericardium with adhesions. The vagus nerve was adherent on the surface of the tumour, and it was separated and secured. The mass does not seem to arise from the thymus. The tumour was totally excised. Haemostasis was complete. A chest tube was inserted and the patient was admitted to the paediatric intensive care unit (Figure 7).

The histopathology final report revealed a gross description: teratoma consists of previously opened cystic structure with sebaceous material measuring 5.0 × 4.0 × 2.0 cm. The wall thickness is 0.5 cm. There is sebaceous white to yellow nodular soft tissue mass arising from the inner surface measuring 2.0 cm in diameter. The remaining glandular wall lining is yellow to white in color (Figure 8).

Microscopic description revealed a mature cystic teratoma. In PICU, the patient tolerated extubation and chest tube removal. The patient was transferred to the general ward in good condition. Follow-up chest X-rays were obtained 13 days, 21 days and one month (Figure 9) from the operation. Finally, the patient was discharged from the hospital in good condition.

Discussion

Teratoma or Germ cell tumours (GCTs) are predominantly found in the gonads and are classified as extragonadal if there is no primary tumour in testes or ovaries.[6] They typically arise in a midline location generated from all three germinal layers, with sites varying with age.[5]

The mediastinum is the second most common site for teratomas in the paediatric age group[6] after thymomas, accounting for 8% of all mediastinal tumours. More than 80% of them are located in the anterior mediastinum.[1] Teratomas are equally common in boys and girls and seem to be more prevalent in adult males. Our case is a rare presentation that involved a 5-year-old male patient. However, the lesion was located in the anterior mediastinum, as in most cases.

In addition, mediastinal GCTs are classified into mature teratomas and immature teratomas. Mature teratomas occur

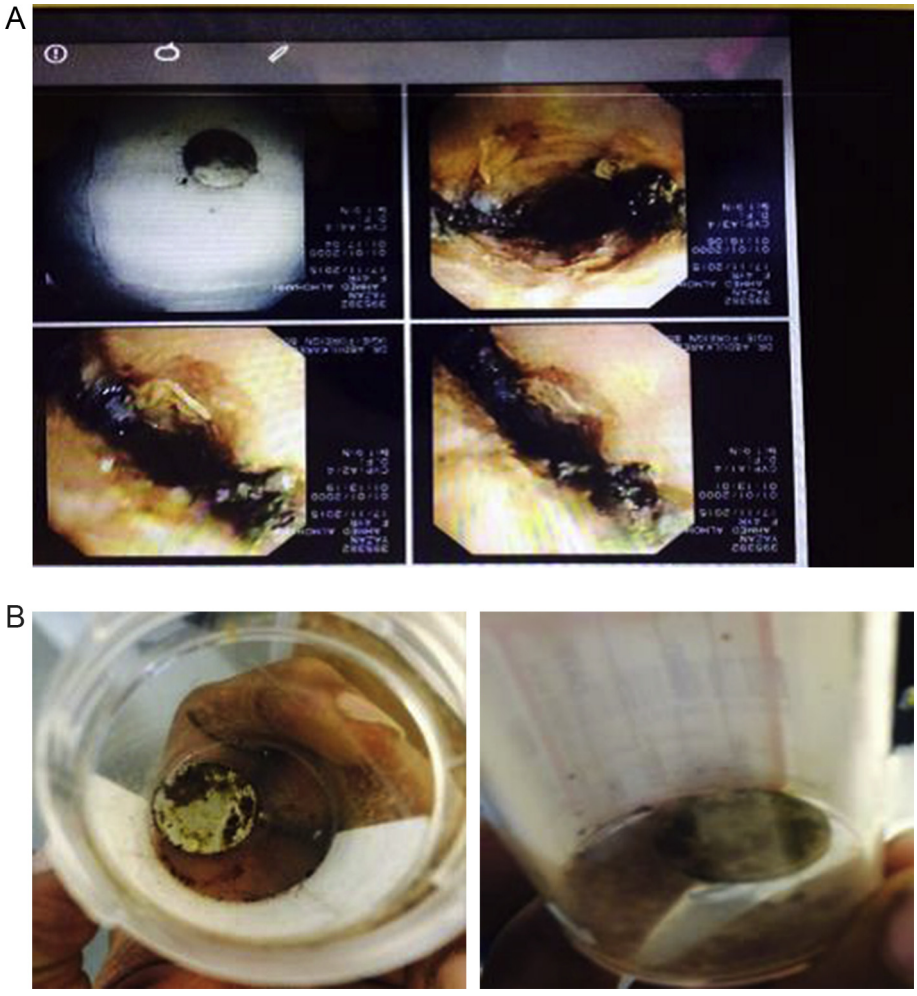

Figure 2: Upper GI endoscopy showed burn and ulceration of the oesophagus (A). The foreign body in the form of a battery (B).

in all age groups but are more common in adolescents. They are generally benign and well-differentiated, usually grow slowly, and are more likely than other GCTs to be diagnosed incidentally while they are still asymptomatic[2] like our patient in this case.

The symptoms of mediastinal teratoma result mainly from compression of the mass on adjacent structures, causing chest pain, cough, respiratory distress and dysphagia.[7] In our case, the patient had a history of sternum and left upper quadrant pain and vomiting,

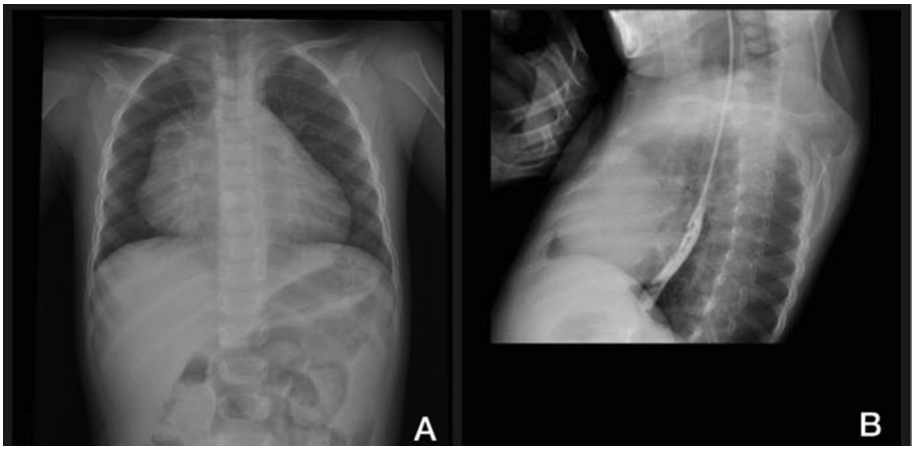

Figure 3: X-ray chest AP view post upper GI endoscopy showed an irregular right cardiac shadow with a bulging right side of the heart (A). X-ray chest lateral view.

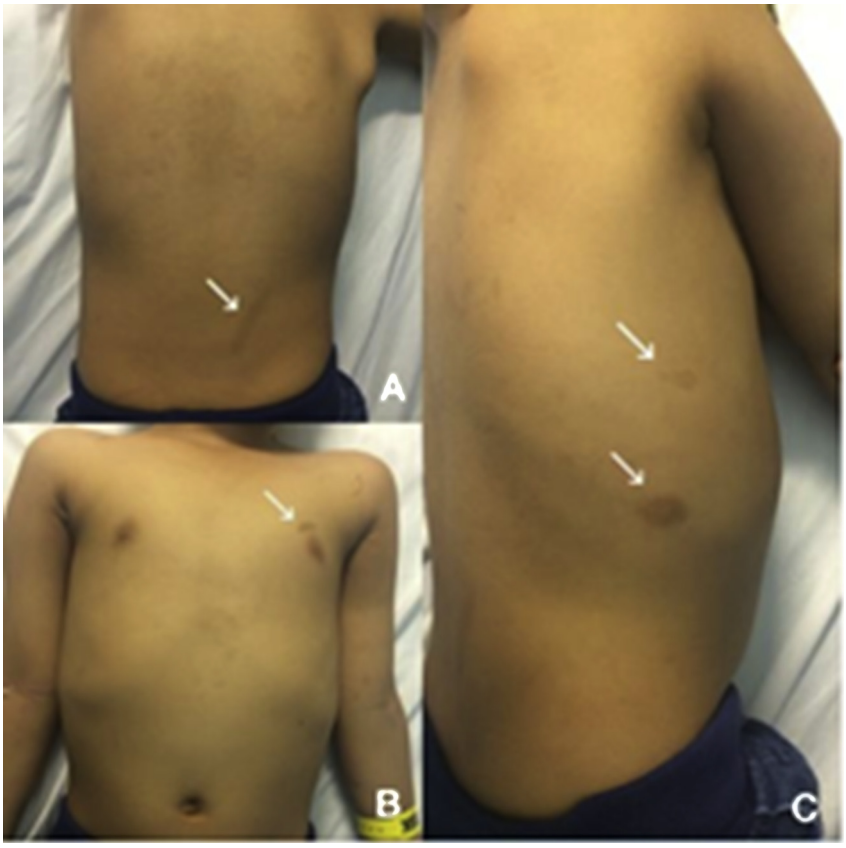

Figure 4: Café au lait spot (A, B, C).

without cough, dysphagia or dyspnea. This is different from other reports in the literature that reported respiratory distress.[8] Our patient had café au lait spots on his trunk, but no relation to mature cystic teratoma was found in the literature.

The most important prognostic factors affecting the mediastinal mass management are the disease, the patient's age, the symptoms at presentation and the mass location.[2] Nearly all masses in the anterior mediastinum consist of lymph nodes, germ cell tumours, thymic masses or the thyroid. Therefore, the differential diagnosis of the present case includes thymoma, lymphoma, and bronchogenic cyst. Chest X-ray and CT scan findings are consistent with the diagnosis of teratoma, which was proved by histopathological examination.

The surgical approach to these tumours has recently changed dramatically. In the past, death was the usual

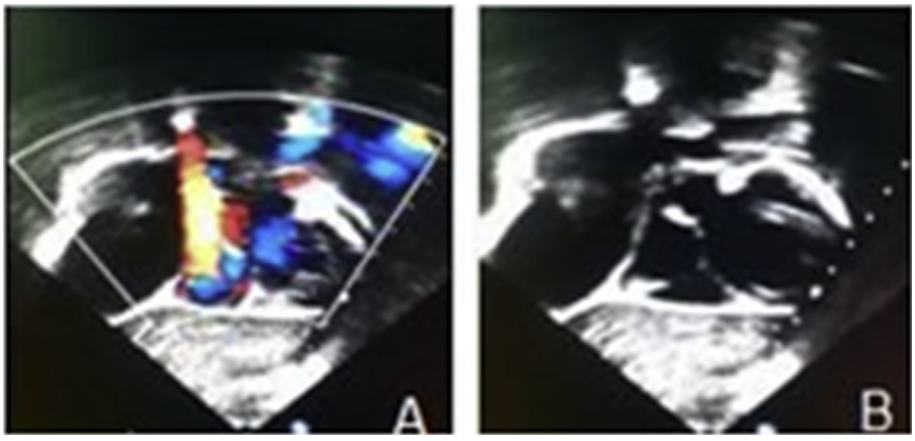

Figure 5: Transthoracic echocardiography showing echogenic mass collected at the right side of the heart (A, B).

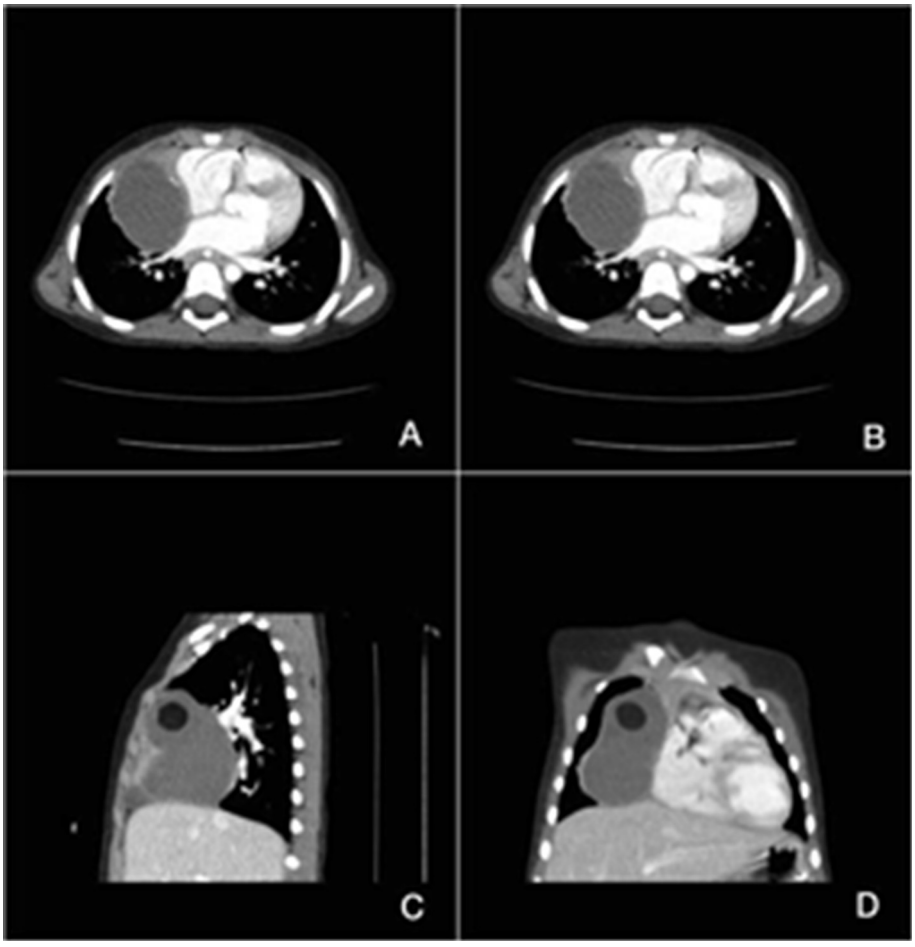

Figure 6: CT scan showed a well-defined large cystic mass lesion noted at the right anterior mediastinum (A) and an anterior cystic mediastinal mass with a fatty component (B, C, D).

outcome either due to the pressure of the tumour on vital structures or the complications of general anaesthesia. With advances in surgical interventions and anaesthetic management, the death rate has significantly declined.[2] Mediastinal teratomas are benign and can be resected completely with good prognosis,[9] as in our case. In a review of 153 children with non-testicular mature teratomas, the six year relapse-free survival rate for completely resected teratoma was 96%.[10]

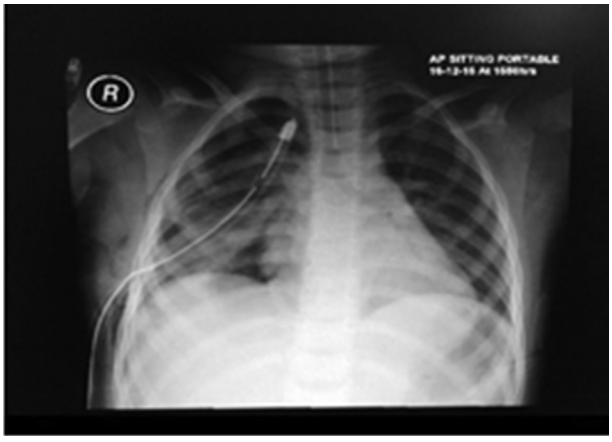

Figure 7: X-ray chest AP view, post-operative with chest tube insertion, showing right pneumomediastinum.

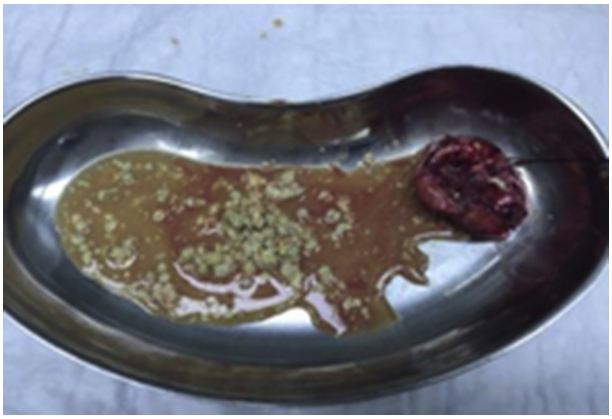

Figure 8: Teratoma cyst rupture shows fluid and fat component.

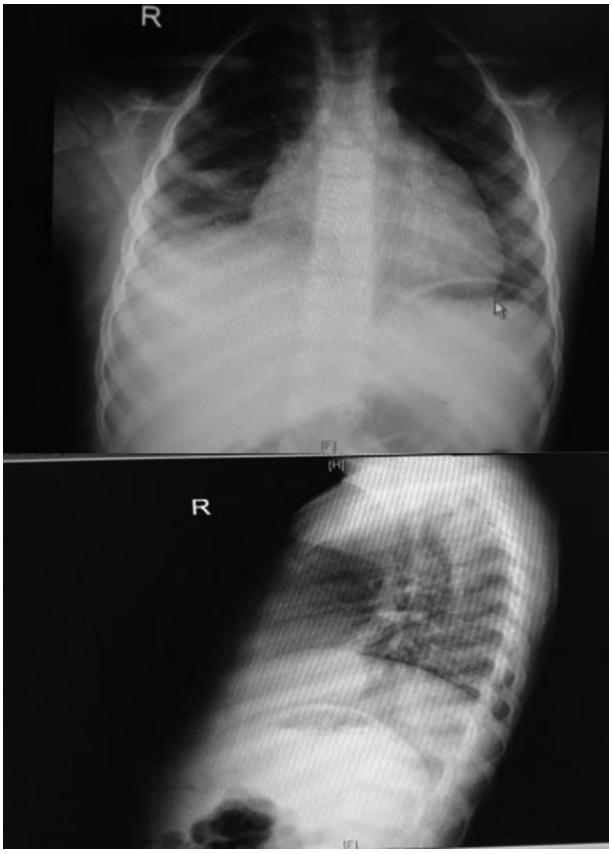

Figure 9: X-ray chest AP and lateral view showed persistent faint patchy opacity identified at the right lower zone with associated pleural thickening with possible effusion. Also noted is an irregularity along the right cardiac border likely related to right middle lobe opacity.

Conclusion

In children, mediastinal teratoma could be presented with a history of sternum and left upper quadrant pain and vomiting without respiratory distress. Therefore, it should be considered in the differential diagnosis of such cases.

Authors' contribution

KMA wrote the abstract, introduction, discussion and provided the references, prepared, edited, and revised the article; and approved the final draft for publication. SAA wrote the patient presentation, collect the pictures and wrote their description, MHS is the main treating consultant for the patient and she supervised reviewed and approved the final version of the article.

Conflict of interest

The authors have no conflict of interest to declare.

Acknowledgements

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