Symptomatic calcifying fibrous tumor of the pleura in a teenager

Ahlam Mazia, Sherif Emil, Chantal Bernard, Anne-Marie Canakis

1. Introduction

Calcifying fibrous tumor (CFT) is a rare benign tumor that originates from subcutaneous and deep soft tissues, with the stomach, small intestine, pleura, mesentery, and peritoneum as the most common internal sites [1]. It is characterized histologically by a non-encapsulated, well-circumscribed mass which contains dense hyalinized collagen, bland spindle cells, lymphoplasmacytic infiltrates, and psammoma bodies or dystrophic calcifications. Its pathogenesis is unclear. It was first described by Rosenthal and Abdul-Karin in 1988 [2]. Multiple or disseminated calcifying fibrous tumors of the pleura are quite rare, with a total of 12 reported cases in the adult literature (Table 1). We are reporting the first case of multiple calcifying fibrous tumors of the pleura (CFTP) in the pediatric age group, in a 15 year old girl which presented with cyclical chest pain.

2. Case report

A 15-year-old girl presented with a one year history of intermittent pleuritic chest pain, which was relieved by non-steroidal anti-inflammatory medications. There were no inciting or associated factors. Past medical history and review of systems was completely unremarkable. The family history was significant for atopy, IgA nephropathy, and multiple sclerosis. Her physical examination was normal. An initial chest radiograph (CXR) (Fig. 1) was interpreted as a possible pneumonia. She was treated with a macrolide antibiotic for 5 days. Despite this treatment, her chest pain worsened. Follow-up CXR showed no resolution.

Given the unusual density of opacity, a chest ultrasound was done. It showed a dense soft tissue lesion in the posterior base of the left chest cavity. A computed tomography angiogram (CT) of the chest was done for further evaluation. (Fig. 1).

She underwent a video-assisted thoracoscopic VATS exploration of the left chest. Multiple small parietal pleural implants were seen on the chest wall, mediastinum and diaphragm. An excisional biopsy of the largest lesion, traversing the left posterior parietal pleura and the left lower lobe visceral pleura, was performed, including a wedge resection of the involved portion of the lung. The mass was a soft multi-lobulated, fleshy, brownish lesion, with thin broad based attachments to the chest wall, diaphragm and lung. It was tethering the left lower lobe to the diaphragm and lateral chest wall.

Histologically, the mass consisted of reactive fibrovascular proliferation with prominent adhesions, hemosiderophages, hemorrhagic and ischemic changes. There was no sign of malignancy or endometriosis. The wedge biopsy showed normal lung. No definitive diagnosis could be made.

Although her pain almost resolved for several months post-operatively, she had a recurrence of intermittent pleuritic left subcostal chest pain, radiating to the left shoulder, back, and epigastric region. Her physical exam, pulmonary function test (PFT), complete blood cell count (CBC), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and full rheumatologic work up remained normal. Her pain was worsening over time and a chart of her pain over one year showed a cyclic pattern in relation to her menstrual cycle. A CXR done during an acute episode ruled out catamenial pneumothoraces. A ten-month trial of a cyclic oral contraceptive pill (OCP) seemed to improve her pain.

Keywords:
Calcifying fibrous tumor of the pleura
CFTP
Cyclical chest pain

ABSTRACT
Calcifying fibrous tumor is a rare benign tumor that occurs in multiple locations, and originates from the subcutaneous and deep soft tissues. There are some reported adult cases of multiple or disseminated calcifying fibrous tumors of the pleura. We are reporting the first case of multiple calcifying fibrous tumors of the pleura in a 15 year old girl presenting with cyclical chest pain, with possible hormonal influence.
However, she presented with a severe, acute exacerbation of her pain, three weeks after stopping the OCP. A CXR showed recurrence of an opacity in the posterior aspect of the left chest. A second CT scan of the chest, 2.5 years after the first one, showed a persistent pleural-based lesions (Fig. 1).

She underwent a second VATS exploration, which revealed multiple nodular lesions of different sizes adjacent to the left lower lobe, along the posterior and lateral chest wall, and adherent to the diaphragm. A complete resection of all gross disease was completed, with cauterization of the smallest implants. Operative and pathologic findings are shown in (Fig. 2).

Pathological evaluation showed multiple irregular pieces of beige, relatively firm tissue, with hemorrhagic nodular areas. It was composed of low cellularity dense connective tissue with a variable amount of inflammation and psammoma bodies. It was positive for vimentin immunohistochemical (IHC) stain, negative for CD34 and ALK-1. There was no evidence of endometriosis and IHC for estrogen (ER) and progesterone (PgR) receptors were negative. These findings were consistent with calcifying fibrous tumor of the pleura. A retrospective review of the initial resection showed similar histopathological findings consistent with the same diagnosis. Post operatively she had no further episodes of pain, after a follow-up period of 2 years.

3. Discussion

Calcifying fibrous tumor (CFT) is an extremely rare benign slow growing tumor that usually occurs in the subcutaneous and deep soft tissues. Up to ten percent of all CFTs have been reported in the pleura [1]. When pleural-based, it is generally a solitary non-encapsulated, well-circumscribed, solid or firm mass although it may present with multiple lesions. Multiple or disseminated pleural CFTs have only been reported in adults. Histologically it is characterized by a bland spindled cell proliferation with dense bands of hyalinized collagen and variable numbers of psammoma bodies and chronic inflammatory cells; the tumor is positive for vimentin, and factor XIIIa with immunohistochemical staining, but negative for ALK-1. There was no evidence of endometriosis and IHC for estrogen (ER) and progesterone (PgR) receptors were negative. These findings were consistent with calcifying fibrous tumor of the pleura. A retrospective review of the initial resection showed similar histopathological findings consistent with the same diagnosis. Post operatively she had no further episodes of pain, after a follow-up period of 2 years.

<table>
<thead>
<tr>
<th>Case Report</th>
<th>Age (years)</th>
<th>Gender</th>
<th>Presentation</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>23</td>
<td>F</td>
<td>Chest pain</td>
<td>Pinkard et al. 1996 [3]</td>
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<tr>
<td>2</td>
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<td>Pinkard et al. 1996 [3]</td>
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<td>3</td>
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<td>F</td>
<td>Asymptomatic</td>
<td>Pinkard et al. 1996 [3]</td>
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<tr>
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<td>Hainaut et al. 1999 [5]</td>
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<tr>
<td>6</td>
<td>40</td>
<td>M</td>
<td>Uncomfortable feeling in the back</td>
<td>Isaka et al. 2011 [6]</td>
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<td>Jiang et al. 2011 [8]</td>
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<tr>
<td>10</td>
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<td>Mito et al. 2005 [10]</td>
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<td>12</td>
<td>35</td>
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<td>Suh et al. 2008 [12]</td>
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<td>Present case</td>
<td>15</td>
<td>F</td>
<td>Cyclical chest pain (onset of symptoms at 14 years of age)</td>
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![Fig. 1. (A), (B) Chest X-ray showed a dense opacity in the posterior left lower lobe. (C) A computed tomography (CT scan) of the chest with angiogram showed two dense opacities in the posterior base of the left chest cavity abutting the pleura. The larger lesion measuring 4 cm × 2 cm, and a smaller lesion measuring 1.5 cm × 1 cm. The provided image shows the larger of the two lesions. (D) A repeat computed tomography (CT scan) of the chest showed atelectasis of the posterior-basal segment of the left lower lobe, a moderate pleural effusion and a dense-homogenous pleural based lesion in the posterior aspect of the left hemithorax.](image-url)
common in adults than pediatrics such as solitary fibrous tumor, calcified granuloma, calcified pleural plaque, inflammatory myofibroblastic tumor (IMT), lipomas and chronic fibrous pleuritis.

4. Conclusion

In conclusion CFT of the pleura is a benign lesion with slightly more reported cases in females compared to males. There have been no reported cases of CFT of the pleura in the pediatric population, or of relation to hormonal influence, specifically to female reproductive hormones. CFTs are not found to express estrogen or progesterone receptors. However, our case may raise the possibility of a hormonal influence despite the absence of histologic confirmation of ER or PgR receptors. Awareness of this lesion dictates a complete excision. Incomplete excision in our case resulted in recurrence of symptoms one year later. In retrospect, the patient's symptoms may have been related to intermittent pleuritis due to hemorrhage within the tumors.

Our case represents the first case of multiple disseminated CFT of the pleura in the pediatric literature with possible hormonal influence, presenting with cyclic chest pain correlating to menstrual cycle.

Competing interests

All authors have no competing interest to declare.

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References