Case report - Thoracic oncologic

Unusual primary pleural leiomyoma

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Abstract

Primary pleural leiomyoma is extremely rare and has only been described a few times. We present a case of a young woman with right pleuritic pain. A computed tomography confirmed the existence of a solid right pleural tumor which had compressed and displaced the lung, mediastinum and heart. Percutaneous biopsy showed a ‘proliferation of smooth muscle cells without evidence of malignancy’. Surgical excision was done and the tumor was not associated to vascular, broncho-pulmonary or mediastinal structures. The definitive diagnosis was primary pleural leiomyoma. Primary pleural leiomyoma should be included in a differential diagnosis of pleural tumors and suspected in asymptomatic patients with radiologically-apparent benign tumors and the presence of smooth muscle fibers in the biopsy. Complete resection and follow-up is advised because it can grow very large and has malignant potential.

Keywords: Pleural tumor; Leiomyoma

1. Introduction

Primary pleural tumors (PPTs) are a rare affection because 75% of the pleural tumors are metastasis. The most frequent benign PPTs are the solitary fibrotic tumors, lipomas, endotheliomas and angiomas. Primary pleural leiomyoma is an extremely rare benign tumor, its etiology unknown, and up to the present has only been described in a few cases in the literature.

2. Case report

A 48-year-old woman, a non-smoker, without antecedents of interest, presented with right pleuritic pain of a three-month evolution, accompanied by a sensation of dyspnea. A physical examination showed only a marked decrease of breath sounds in the lower right third and dullness to percussion. A simple thoracic X-ray showed a mass in the lower third of the right hemithorax. Thoracic computed tomography confirmed the existence of a solid right pleural tumor, with a benign appearance, vascularised, capsulated, 16×10×11 cm in size, without any signs of infiltration, which had compressed and displaced the lung parenchyma, mediastinum and heart (Fig. 1a). Routine blood biochemistry, cancer markers and fibrobronchoscopy were all normal. Percutaneous biopsy under radiologic control showed a ‘proliferation of smooth muscle cells without evidence of malignancy’.

Surgical intervention was decided for diagnosis and treatment (clinical symptoms, size and inconclusive diagnosis).

Right posterolateral thoracotomy was performed and a giant tumor was observed. It was well-circumscribed, capsulated, hypervascularized, occupied approximately 70% of the pleural cavity and compressed the lung and mediastinal structures (Fig. 1b). The tumor was not associated to vascular, broncho-pulmonary or mediastinal structures and no invasion with the lung, mediastinum and chest wall. No adenopathy was present.

So, a complete excision was done. Macroscopic study showed a well-circumscribed tumor weighing 1160 g measuring 18×14×11 cm. The section was solid white with cystic cavities. Microscopic findings revealed a proliferation of interlaced fascicles of monomorphic well-differentiated smooth muscle cells (cigar-shaped nuclei and eosinophilic cytoplasm with haematoxylin and eosin stain). Mitotic figures and atypia were absent (Fig. 2a). The immunohistochemical study was positive for α-smooth muscle actin and desmin and negative for estrogen and progesterone receptors (Fig. 2b). Because no organ of origin of leiomyoma, other than pleura, was detected, our patient was diagnosed as a primary pleural leiomyoma. The postoperative period passed without incidence and the patient was discharged after five days. An 18-month follow-up showed no signs of relapse.

3. Discussion

Leiomyoma is a benign tumor that derives from smooth muscle fiber, located mainly genitourinary, gastrointestinal and, very rarely, respiratory tract. Despite its benign histological appearance, leiomyoma has a low malignant potential [1–3]. At the thoracic level, this tumor can begin
in the oesophagus, bronchi or vascular structures; therefore, its location can be mediastinal, pulmonary parenchyma, thoracic wall, diaphragm and pleural cavity. Of all types, mediastinal leiomyoma is the most frequent, originating in the oesophagus, and some cases describe it originating in the aorta or vena cava [4].

Primary pleural leiomyoma is extremely rare, its etiology unknown, and up to the present has only been described in a few cases in the literature [2–5]. It is a well-delimited and capsulated tumor that grows in the pleural cavity, but is not related to thoracic structures that contain smooth muscle fiber, as in our case [5]. A differential diagnosis should always be established with both primary pleural (solitary fibrous tumor or mesothelioma) or metastatic tumors [6]. Clinically, they are usually silent tumors and the majority of patients are asymptomatic, and their presence is a discovery during an image study [2]. Symptoms, when present, are non-specific, and are frequently related to the tumor's size or location, manifesting as pleuritic pain, cough dyspnea or dysphagia. A physical examination usually has normal results or there is auscultatory hypophonia if the size is considerable. Routine blood biochemistry does not provide anything for the diagnosis and cannot be differentiated radiologically from other tumors of the pleura [2]. Radiological evaluation should be done with chest radiograph, computed tomography, angiography and magnetic resonance which can reveal the size, location, level of vascularity and anatomic relations [4]. A precise diagnosis requires histological confirmation and can be obtained by X-ray guided transthoracic biopsy in the preoperative phase. A definitive diagnosis is always histological and requires confirmation of smooth muscle fibers without signs of malignancy (mitosis and/or dysplasia) with the hematoxylin/eosin technique. Immunohistochemical techniques will show positive when faced with actin, vimentin, desmin and HH35 soft muscle protein and electronic microscope technique may help to provide a precise diagnosis [7]. The treatment of choice is excision to avoid symptoms and signs and confirm the diagnosis because this tumor is a well-defined entity with a low, but definite malignant potential [1–3, 8]. Therefore, the complete resection is recommended [1]. Surgery can be done by video-assisted thoracoscopic surgery for localized small lesions and thoracotomy should be reserved for large and highly vascularized tumors, as in our case [9]. Even though the histopathological malignant findings are not detected, the leiomyoma may increase with local infiltration if the tumor is not resected completely [2]. Preoperative embolization has been described to shrink the tumor and minimize the risk of intraoperative hemorrhage [10].

4. Conclusions

Primary pleural leiomyoma is an infrequent tumor but should be included in a differential diagnosis of pleural tumors. It should be suspected in asymptomatic patients with radiologically-apparent benign tumors and the presence of smooth muscle tissue in the biopsy. Surgical excision is always justified and recommended because these tumors can grow very large, cause serious symptoms or signs and degenerate to malignant tumor.

References