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Case report

A rare clinical case – Solitary fibrous tumor of the pleura

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ABSTRACT

We present a case of a 58 years old man with a large heterogeneous and well circumscribed soft tissue mass arising from the right pleural surface, found at a computer tomography of his chest. This mass after complete resection through a right lateral open thoracotomy, proved to be a Solitary Fibrous Tumor, previously known as 'benign mesothelioma'. This tumor is usually discovered at routine chest X-rays since patients are either asymptomatic or report atypical symptoms. Only 10–20% of the published cases report a malignant solitary fibrous tumor, however, definite diagnosis can only be made after complete resection which is the proposed diagnostic algorithm for these cases.

1. Introduction

Solitary Fibrous Tumor of the Pleura (SFTP) is a pleural neoplasm initially thought to be of fibroblastic origin and historically named 'benign mesothelioma, localized mesothelioma or submesothelial fibroma' [1]. SFTP is rare and accounts for less than 5% of the primary pleural tumors [2] and patients have a mean age of 58–66 years at the time of diagnosis according to large retrospective studies [3,4]. While approximately one third of the patients are asymptomatic, the majority report atypical symptoms like cough, shortness of breath or chest pain at presentation [3,4]. We present a case of a male patient who underwent a right lateral open thoracotomy for total resection of the pleural tumor which proved to be a SFTP by the histopathological and immunochemical examination.

2. Case report

A 58 years old Caucasian male appeared at the outpatient clinic complaining for dull aching right sided chest pain, moderate dyspnea on exertion over the last 2 weeks and dry chronic cough. His past medical history was unremarkable, apart from being a heavy smoker for more than 30 years. The clinical examination revealed decreased auscultatory sounds and dull note over the middle and lower right hemithorax, while electrocardiography, transthoracic echocardiogram and routine blood laboratory results were all normal. Because of a rounded mass appearing at the lower right side of his chest radiograph (Fig. 1A), the patient underwent a chest CT scan. The CT scan displayed a large right-sided heterogeneous but well circumscribed soft tissue mass arising from the pleural surface, which contained central areas of low density, with absence of local invasion (no chest wall or heart infiltration) (Fig. 1B). Diagnosis was obtained by a transthoracic cutting needle biopsy, and confirmatory diagnosis was made with the pathological examination of the specimen which was completely resected during a right lateral open thoracotomy (Fig. 2A). Histopathological examination of the mass (Fig. 2B) demonstrate hypocellular and hypercellular areas simultaneously, separated by fibrous stroma; hypercellular areas were composed of bland spindle-cells arranged in short fascicules. Immunohistochemical staining showed that the tumor cells were strongly positive for CD34 (Fig. 3), bcl-2 and vimentin which confirmed the diagnosis of SFTP.

3. Discussion

SFTP is a rare localized mesenchymal tumor which was initially thought to be a mesothelial pleural lesion; nowadays, is acceptable that it can originate from visceral organs (thyroid, lungs, etc) or mesothelial tissues (pericardium, peritoneum, pleura) [5]. Patients may be asymptomatic and SFTP is usually discovered on routine chest X-rays; other patients report atypical pulmonary symptoms like dry cough, chest pain and shortness of breath [6]. Interestingly, Pierre-Marie-Bamberg (hypertrophic pulmonary osteoarthropathy) and Doege-Potter (refractory hypoglycemia) syndromes have been also reported in patients with SFTP [5,7].

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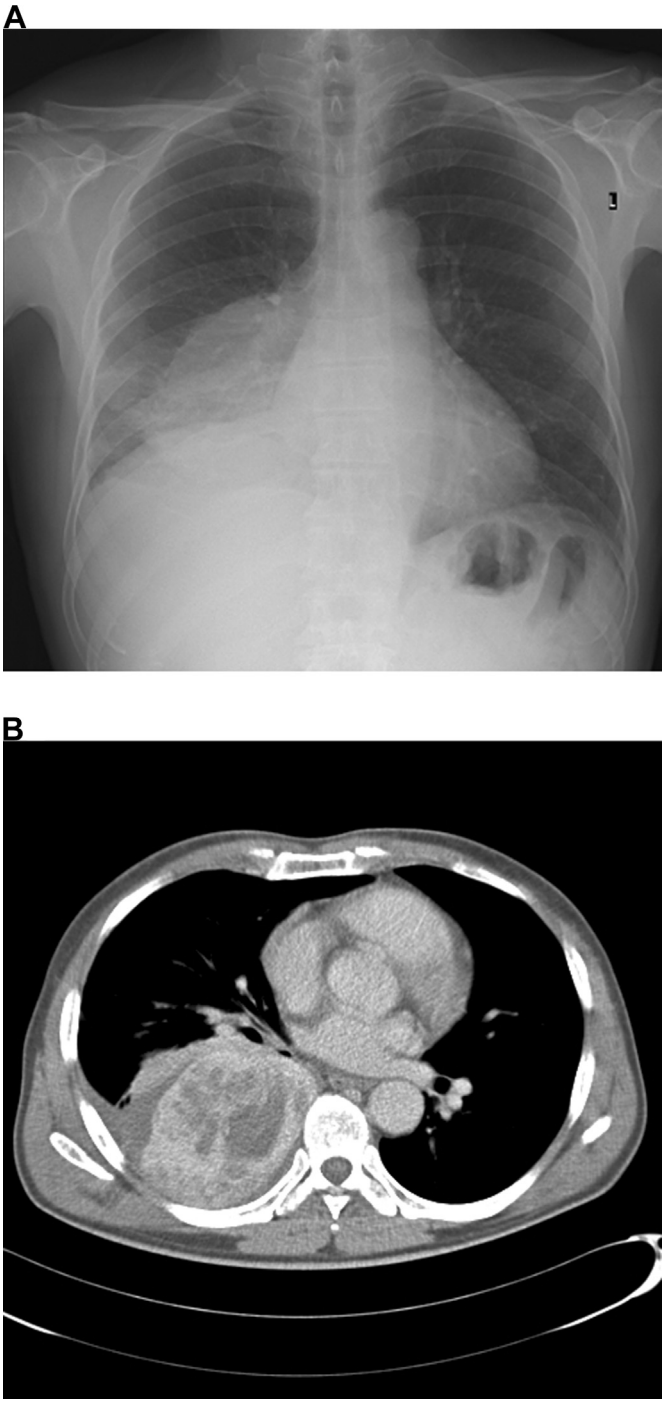

Fig. 1. A. is the chest-X-ray, B. the CT scan of the thorax.

CT of the chest commonly displays a well demarcated soft tissue mass heterogenous in attenuation, next to the chest wall or within a fissure, creating a characteristic obtuse angle with the surface [7].

Since benign and malignant SFTPs share the same radiological appearance and transthoracic needle biopsy has poor diagnostic value [8], their preoperative distinction is difficult and not encouraged. Therefore, complete resection of the tumor is the standard surgical approach to confirm the diagnosis.

Macroscopically, the tumor is whorled, encapsulated and lobular, and may vary in size and weight [7,9] (in our current case

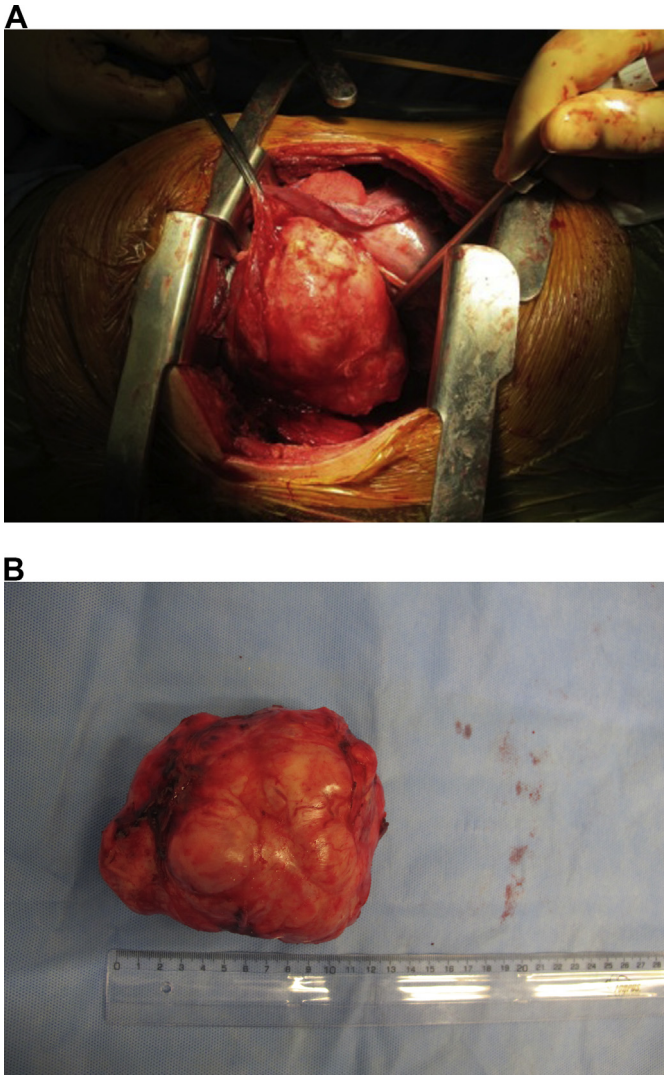

Fig. 2. A. the picture during the operation, B. the picture of the mass with the ruler.

the encapsulated mass was measured 13 × 8 × 2.5 cm with a weight of 480gr). The majority of these tumors are benign, but malignant histological aspects like infiltration of adjacent structures, presence of necrosis or hemorrhage, high mitotic index and nuclear atypia can be found [8]. Large SFTPs have been associated with intralesional calcifications and high likelihood for malignancy [7]. Immunochemistry is quite characteristic for SFTPs since these tumors are positive for CD34-, CD99-, bcl-2- and vimentin, but negative for cytokeratin [5,6].

SFTPs can recur locally and distant metastases can occur particularly in tumors that have histologic features of malignancy, therefore close follow-up is essential [9]. Approximately 10%–20% of the cases reported in literature are malignant [5]. Typically, resection of the tumor is correlated with favorable outcome and patients have a 5-year disease-free survival rate of 84% [10]. However, if the tumor cannot be removed surgically or when metastases develop, chemotherapy and/or local radiotherapy have been proposed as palliative care [7].

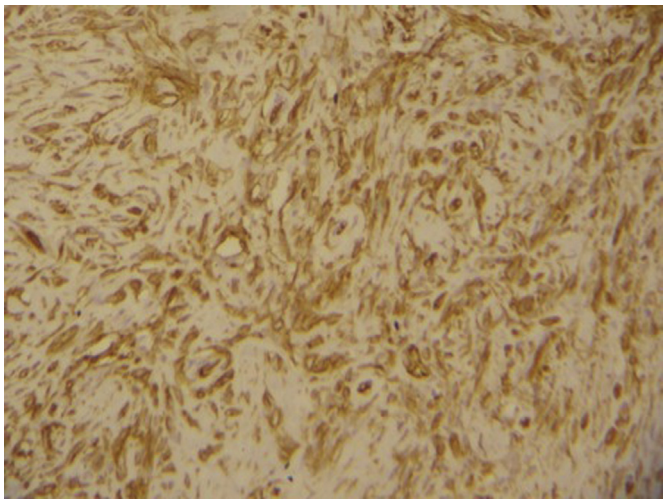

Fig. 3. The laboratory picture of the immunohistochemical staining.

4. Conclusion

Solitary fibrous tumor of the pleura is a rare localized mesenchymal tumor which is often radiologically and clinically misdiagnosed as primary lung carcinoma. However, it can be benign or malignant and differential diagnosis is only feasible after complete resection of this tumor which is the standard approach in these cases. Immunochemistry of the resected tissue is an important complementary tool for verifying the diagnosis of SFTP. Despite that the majority of SFTPs are benign and resection maintain good prognosis with a high 5-year survival rate, patients must be regularly followed-up since some have malignant histologic features and local recurrences or metastatic lesions can be found.

Contributors

AP gathered the clinical information and drafted the manuscript. GC provided the operation findings and IP and AG helped in writing this manuscript.

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