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Case report

Not to be confused, this is a pleural fibroma: Case report and literature review

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ABSTRACT

Solitary fibrous tumors of the pleura are rare mesenchymal tumors. The diagnosis is often made incidentally. We report the case of 65 year old patient consulting for dry cough evolving for 1 year. The clinical examination revealed a mattness of the right hemi thorax. The chest X-ray detected a right lower lobar opacity. The CT scan showed a right lateral basal mass, connected at an obtuse angle with the wall. A CTguided biopsy with an anatomopathological and immunohistochemical examination were performed, thus confirming the diagnosis of a solitary fibrous pleural tumor. The evolution after treatment was favorable. Complete surgical resection of the tumor is usually sufficient, but there are reported cases with recurrence.

1. Introduction

Less than 10 % of all pleural tumors are pleural solitary fibrous tumors, a rare kind of mesenchymal tumor. A typical radiograph is used to diagnose a pleural tumor. The mass can be described using the thoracic CT scan, which can also be used to look for signals that show the tumor is benign or malignant. The diagnosis will be verified by immunohistochemical and anatomicopathological tests.

2. Case report

65-year-old patient consulting for dry cough evolving for 1 year. The clinical examination revealed a mattness of the right hemi thorax. The chest X-ray detected a right lower lobar opacity. The CT scan showed a right lateral basal mass that was linked to the wall at an obtuse angle. This tissue mass was heterogeneously enhanced after injection without significant individualisation of infiltration or invasion of adjacent structures (Fig. 1). The diagnosis of a solitary fibrous pleural tumor rising from the visceral pleura was confirmed after a CT-guided biopsy and an anatomopathological and immunohistochemical study. Histologically, it was a more or less dense proliferation of spindle-shaped cells resembling fibroblasts, dispersed in an orderly fashion and supported by a framework of variable abundance collagen fibers. Fibrous tumor immunohistochemistry typically reveals diffuse expression of vimentin and CD34, variable intensity expression of CD99 and Bcl-2 protein, and negativity of epithelial markers (cytokeratin, EMA, and S100 protein). Pleural fibroma is a rather uncommon condition. It is frequently asymptomatic and the result of an unintentional radiological discovery.

3. Discussion

Klemper and Rabin first referred to the solitary fibrous tumor of the pleura (SFTP) as "localized mesothelium" in 1931. Since then, it has gone by a number of names that incorporate the words mesothelial, pleural, fibrous, localized, benign, and solitary [1]. Formerly known as benign mesothelium, solitary fibrous tumors were once believed to have mesothelial origins. However, immunohistochemical investigations have shown that they actually come from subpleural connective tissue [2]. They are rare mesenchymal tumors as a result, accounting for less than 10 % of all pleural tumors [1]. However, they have also been observed in the pericardium, meninges, liver, and pancreas. They grow at the cost of the visceral layer of the pleura [3]. They typically start to show up after age 50, with no predilection for both sexes [4]. Asbestos and tobacco do not cause one another.

SFTPs are frequently asymptomatic in more than half of patients with an unintentional radiological finding. Although they can occasionally present as compressive or irritative symptoms, a haemothorax, or a paraneoplastic syndrome like Pierre Marie-Foix Syndrome or Doege-Potter Syndrome [2].

On the chest X-ray, a well-contained, round or oval opacity of varied

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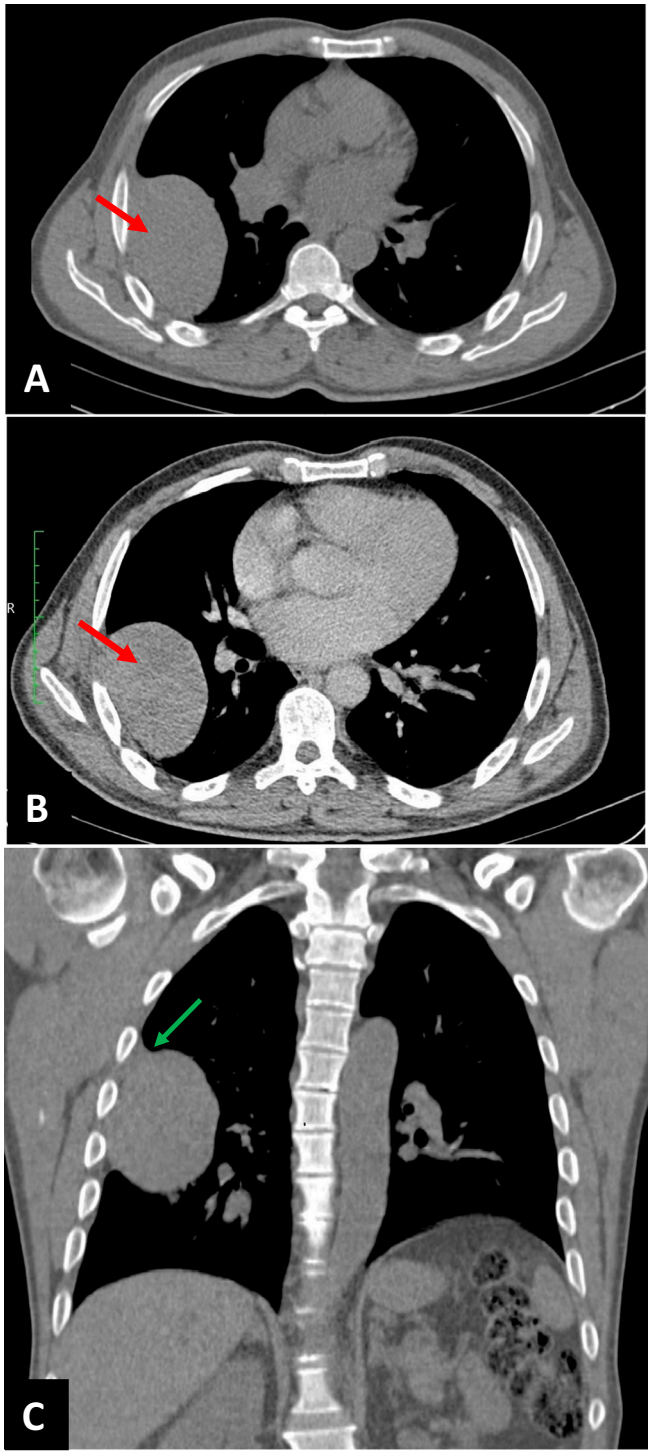

Fig. 1. Chest CT scan in cross-section (A, B) and coronal section (C) showing a non-infiltrative, 6 cm long, heterogeneously enhancing tissue process arising from the right costal pleura (red arrows) after contrast injection. Note the obtuse angle connection with the wall (green arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

sizes that is sometimes lobulated and frequently located at the base of the thorax and connects with the wall at an acute angle is visible. It might or might not be connected to a pleural effusion. Depending on how the patient is positioned in the pedunculated fibroids, this opacity may change [5].

On scanning, the soft tissue mass is tightly packed, occasionally polylobed, and its composition might be homogeneous or heterogeneous. Heterogeneity is affected by the size of the tumor, the presence of necrotic or hemorrhagic changes, and calcifications (which are present in 5 % of instances) [1]. It could develop at the expense of the costal, diaphragmatic, or mediastinal pleura or have a scissural localization [2]. A big tumor or the presence of a pedicle might cause the insertion angle to be acute or even obtuse and acute at the same time. The pedicle being seen in its entirety is remarkable [5]. Since there is a significant vascular contingent, enhancement is typically severe and early [4].

MRI can provide a more accurate description of the mass's internal structure, as well as its relationship to the wall, the apex, and the diaphragm [4]. [2]. The signal is variable and often heterogeneous depending on the histological makeup of the mass [4].

Malignancy in an SFTP can develop spontaneously or as a side effect of a benign type [1]. A high mitotic index continues to be the best indication of cancer, despite the lack of a clear test for it [2]. Imaging reveals cancer if the surrounding structures are invaded and there are lung nodules or other secondary locations [2].

The definitive diagnosis is made via histology, which is typically done on resections [5].

The differential diagnosis includes additional causes of pleural tissue loss, such as metastases, localized mesothelioma, lymphoma, sarcoma, lipoma, desmoid tumor, hemangiopericytoma, and fibrous malignant histiocytoma [2]. Thymoma and neurinoma may present a differential diagnosis challenge with the single fibrous tumor if it originates from the mediastinal or costal pleura [2].

The typical course of treatment for SFTPs is large surgical excision [1]. Neo-adjuvant chemotherapy may be used in the case of large tumors that cannot be immediately removed [2]. In more aggressive kinds, it can be strengthened by chemotherapy and/or radiotherapy, but its efficacy has not been established [1]. Given that it is challenging to evaluate the evolutionary profile, long-term radio-clinical surveillance is advised [1]. Preoperative embolization of some major blood vessels can be used to treat large tumors with an abundant blood supply [6]. In our case, the tumor was completely removed via a right posterolateral thoracotomy. There were no other nodules to be found. After a 4-day stay in the intensive care unit, the postoperative course was straightforward, and the patient was discharged 10 days later. Six months later, the evolution was favorable, with the resumption of a good respiratory dynamic and a satisfactory control radiography.

This work has been reported in line with the SCARE criteria [7].

4. Conclusion

In conclusion, SFTPs are uncommon tumors that benefit most from complete surgical resection. In this series, increased tumor growth is a sign of malignancy. Although it is also conceivable after malignant SFTP resection, long-term survival after benign SFT resection is excellent. On the other hand, malignant SFTPs are more likely to recur, which is associated with a poor prognosis.

Guarantor of submission

The corresponding author is the guarantor of submission.

Consent statement

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Ethical approval

Article produced in compliance with legal, ethical and institutional rules.

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The authors do not declare any conflict of interest.

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