ANNALI ITALIANI DI CHIRURGIA

Digital Edition
e-publish on-line
ISSN 2239-253X
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Ann Ital Chir, Digital Edition 2019, 8
pii: S2239253X1903086X - Epub, Sept. 19
free reading: www.annitalchir.com

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Intrapulmonary solitary fibrous tumour: a rare neoplasm of the lung and a challenging pre-operative diagnosis.

BACKGROUND: *Case report of a 78-year-old male who came to our observation for a growing pulmonary mass of the left upper lobe without a pre-operative diagnosis. Post-operative histopathology revealed an intrapulmonary Solitary Fibrous Tumour (SFT) of the lung.*
CASE REPORT: *The pulmonary lesion was an ovoid mass centrally located in the left upper lobe; it had been known and stable in size for two years; the last Chest Computed Tomography (CT) Scan highlighted a marked increase in maximum diameter (35 versus 22 mm) with contrast enhancement. In view of the growth of the lesion, the patient was referred for surgery after multidisciplinary team evaluation, although a pre-operative diagnosis had not been reached. The patient underwent Video-Assisted Thoracic Surgery (VATS) left upper lobectomy by a biportal approach. The pulmonary nodule consisted of a proliferation of bland-looking spindle cells intermingled with fibrotic stroma and alternating sclerotic and cellular areas. At immunohistochemistry, the spindle cells expressed CD34, bcl2 and CD99. A final diagnosis of intrapulmonary SFT was reached. The 36-month follow-up was negative for relapses.*
CONCLUSIONS: *Primary intrapulmonary SFTs are extremely rare neoplasms, generally with benign biological behaviour; surgical resection is safe and essential in order to be curative and can be achieved by a minimally invasive approach. Pre-operative diagnosis is challenging, due to aspecific clinical and radiological features.*

KEY WORDS: Lung, Solitary Fibrous Tumour, Video-Assisted Thoracic Surgery Lobectomy

Introduction

Solitary fibrous tumours (SFTs) are mesenchymal neoplasms that are usually seen in the chest, generally arising from the pleura [1] and representing less than 5% of pleural tumours[2]; unusual sites of origin are reported [3]: in these settings, intrapulmonary SFTs are extremely rare [4].
Considering their rarity, the clinical course, prognosis and imaging features of intrapulmonary SFTs are not clearly understood.

We report a primary SFT of the lung successfully treated by biportal VATS left upper lobectomy; the definitive diagnosis was reached only after histopathologic examination of the lesion following surgery.

Case Report

A 78-year-old male was admitted to our Thoracic Surgery Department for a growing pulmonary lesion of the left upper lobe. Past medical history showed chronic atrial fibrillation, previous Percutaneous Transluminal Angioplasty of the left superficial femoral artery and a pituitary adenoma known since 1990.
The pulmonary lesion was an ovoid mass centrally located in the left upper lobe; it had been known and stable in size for two years; however, the last Chest CT

Pervenuto in Redazione Maggio 2019. Accettato per la pubblicazione Giugno 2019
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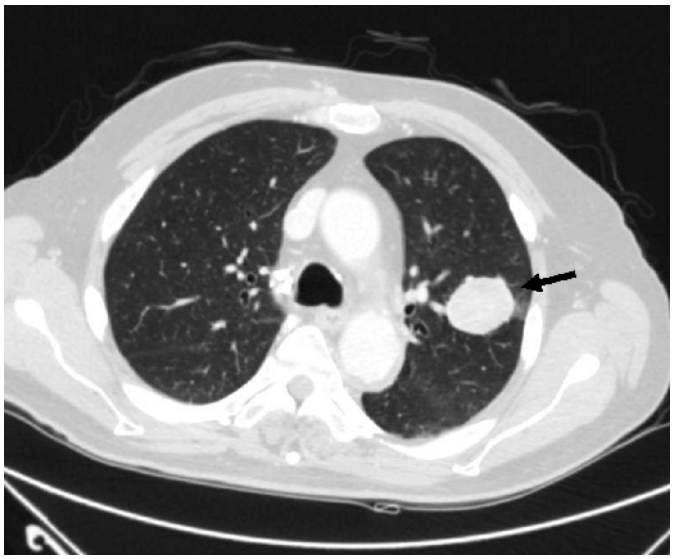

Fig. 1: Chest Computed Tomography (CT) Scan showing an ovoid mass (arrow) centrally located in the left upper lobe with contrast enhancement.

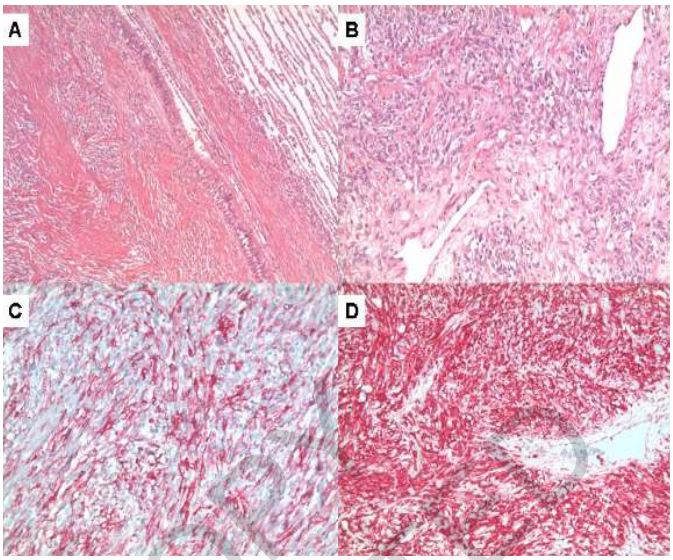

Fig. 2: The tumor showed an intrapulmonary growth partially lined by bronchial mucosa (A, hematoxylin-eosin magnification X100) and consisted of a bland-looking spindle cell proliferation with fibrotic stroma and dilated figurate vessels (B, hematoxylin-eosin stain, magnification X200). The tumor cells expressed CD34 (C, immunohistochemistry; magnification X200) and bcl2 (D, immunohistochemistry; magnification X200).

Scan highlighted a marked increase in maximum diameter (35 versus 22 mm) with contrast enhancement (Fig. 1); fluoro-deoxy-glucose positron emission tomography (FDGPET) findings were not useful, with a maximum standardized uptake value (SUVmax) less than two. In view of the growth of the lesion, the patient was referred for surgery after multidisciplinary team evaluation, although a pre-operative diagnosis had not been reached; our clinical suspicion ranged from pulmonary carcinoid to an amartochondroma or other benign lesions such as leiomyomas; despite this, a primary adenocarcinoma could not be excluded.

Pre-operative cardio-pulmonary functional tests showed no contraindications to thoracic surgery and the patient underwent VATS left upper lobectomy by biportal approach. Macroscopically, the tumour resembled a malignant neoplasm; it was a white-grey solid mass of about four centimetres with visceral pleural involvement, protruding deeply into the lobe. A wedge resection for frozen section histopathologic examination was not achievable, so we decided to perform the lobectomy directly. A sampling lymphadenectomy of the hilar and mediastinal lymph nodes was accomplished.

The pulmonary nodule consisted of a proliferation of bland-looking spindle cells intermingled with fibrotic stroma and alternating sclerotic and cellular areas (Fig. 2A). The lesion also showed peculiar haemangiopercytoma-like, figurate vessels (Fig. 2B). Mitotic figures were quite rare (one mitosis X 10 high power fields) and necrosis was absent. At immunohistochemistry, the spindle cells expressed CD34 (Fig. 2C), bcl2 (Fig. 2D) and CD99, while no staining was observed with pan-cytokeratins (AE1/AE3), desmin, smooth-muscle actin, S100, CD31, or HMB45.

A diagnosis of SFT with intrapulmonary growth (intrapulmonary SFT) was made; surgical free margins were achieved and lymph nodes examination did not show the presence of malignant cells.

The post-operative period was complicated by a delayed left pectoralis subcutaneous emphysema that occurred on post-operative day (POD) 15; only clinical and radiological observation was required and the patient was discharged five days later. The patient was followed on an outpatient basis for 36 months; this consisted of a Chest CT scan every 6 months; no relapse of disease was recorded.

Discussion

SFTs are rare, slow-growing mesenchymal neoplasms that most often occur in the pleura. They maybe rarely found in the lung, central nervous system, kidney and other extrapleural sites like breast, hearth, stomach, etc. [5]. Intrapulmonary SFTs generally are found incidentally with clinical and radiographic appearance as common lung tumours.

Chest CT-Scan radiological features are not specific: primary SFTs of the lung look like well-defined ovoid or round pulmonary nodules. On the other hand, the role of FDG PET is not commonly accepted: some Authors suggest that a high SUVmax predicts a malignant behaviour [6]; finally, pre-operative diagnosis is difficult to obtain with a transthoracic biopsy, due to the low cel-

lularity of these lesions. Surgical excision with free margins can be obtained with a broad spectrum of procedures: from wedge resection to pneumonectomy, depending on the size of the tumour and the extension to nearby structures. A low percentage of intrapulmonary SFTs present malignant pathological features and are associated with a worse prognosis. Surgery must therefore be radical with a careful follow-up.

Conclusions

Primary intrapulmonary SFTs are extremely rare. Radiological features and biological behaviour are not entirely known; pre-operative diagnosis is difficult. Radical resection is mandatory for successful treatment and to avoid recurrence, and a long-term follow-up is necessary.

Riassunto

Riportiamo il caso singolare di un tumore solitario fibroso intrapolmonare; i tumori solitari fibrosi generalmente originano dalla pleura, raramente da siti extrapleurici quali il polmone o altri organi solidi. Il paziente, un uomo di 78 anni ci era stato inviato dopo discussione multidisciplinare per una lesione polmonare del lobo superiore sinistro in accrescimento volumetrico; come spesso accade per i tumori solitari fibrosi del polmone, la diagnostica per immagini non è stata dirimente. Il nodulo si presentava a margini lisci e ovoidali; la captazione PET non era significativa. Il nostro sospetto spaziava dal carcinoide polmonare all'amartoma condroide, pur non potendo escludere lesioni neoplastiche maligne. Il paziente è stato sottoposto ad una lobectomia superiore sinistra in VATS con approccio biportale: in sede intra-operatoria non fu possibile eseguire una resezione atipica per un esame estemporaneo in quanto la lesione era centrale e voluminosa.

La diagnosi anatomo-patologica post-operatoria si è avvalsa di tecniche immunoistochimiche ed ha rivelato una proliferazione cellulare mesenchimale compatibile con un tumore solitario fibroso polmonare. Il decorso post-operatorio è stato caratterizzato dalla comparsa tardiva di enfisema sottocutaneo dei tessuti molli dell'emitorace sinistro, che non ha comunque richiesto il posizionamento di un nuovo drenaggio pleruico. Il follow-up è attualmente negativo per recidive a tre anni dall'intervento chirurgico.

Concludendo, i tumori solitari fibrosi del polmone sono rare neoplasie mesenchimali dal comportamento biologico incerto. Una bassa percentuale presenta caratteristiche di malignità: per questo l'atto chirurgico deve essere sempre finalizzato a rispettare i canoni di radicalità oncologica ed il follow-up a medio-lungo termine è necessario.

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