The Unpredictable Behaviour of the Solitary Fibrous Pleural Tumour

El comportamiento impredecible del tumor fibroso solitario pleural

To the Editor:

Solitary fibrous tumours of the pleura (SFTP) are rare neoplasias of mesenchymal origin, of frequently unpredictable behaviour which does not always correlate with their histology. They are usually benign and remain asymptomatic for long periods of time until they become malignant.\(^1,2\) We present the case of a male with a SFTP without a malignant histology but with aggressive behaviour.

A male patient, 66 years old, smoker, 40 packs a year, with HT, dyslipidaemia, intolerance to glucose and peripheral arteriopathy with a left iliofemoral bypass in 2001. Referred to pneumology due to findings on a chest CT requested because of left pleural effusion after tumour surgery.\(^1,3,5\) The treatment of choice is surgery, with perioperative mortality is low (0-1.5 %).\(^3\)

In some months’ evolution. During physical exam left lung base hypoventilation up to the middle field during auscultation was found. Basic laboratory analyses were normal and on echocardiogram moderate pulmonary hypertension was seen. A MR was performed (fig. 1B) in which the CT findings were confirmed. The tumour was seen to contact the thoracoabdominal aorta without infiltration and signs of spinal compression were seen so treatment with dexamethasone was begun.

Finally, by means of Trucut puncture a neoplasia was seen with immunohistochemical expression of CD34 and bcl-2, negative for cytokeratin and actin, with overexpression of p53 complying with criteria for non-malignant SFTP.

Respiratory function tests were normal and surgery was programmed. Only the intraspinal tumour was removed as it was very vascularised and bled abundantly, which caused postoperative instability and the patient died of respiratory distress. The anatopathology of the resected surgical specimen confirmed diagnosis.

SFTP is a rare type of tumour with about 800 cases described in the literature.\(^1,3\) This tumour constitutes 5 % of tumours of the pleura and the second primitive tumour in that location, after diffuse malignant mesothelioma.\(^2,3,5\)

About 80 % are benign and it is difficult to distinguish them from malignant tumours due to their histological similarities, a larger size and invasion of neighbouring structures are more typical of malignant tumours and can serve as orientation.\(^1,5\)

The immunohistochemical profile of SFTP is characteristic with positive reactions to vimentine and CD34 and a negative reaction to cytokeratin, that distinguish it from malignant mesothelioma. Overexpression of p53 and an increase in neoplastic Ki67 positive cells are associated with a poor prognosis.\(^2\) It is not associated with exposure to asbestos, tobacco or other carcinogens.\(^1,5\) These tumours are more frequent between 60-70 years of age and there is no difference between sexes.\(^1,3,5\) 50 % of cases are asymptomatic and their finding is casual in an imaging study.\(^1,2,5\) They present as well circumscribed peripheral pulmonary masses which are sometimes lobulated or have pedicles. Their density is usually homogeneous, it can be heterogeneous in malignant variants or benign ones with necrosis, haemorrhage or myxoid degeneration. Pleural effusion is rare and is usually a sign of malignancy. Calcifications in contrast are non-specific. MR better defines the relationship with neighbouring structures.\(^1,5\) The most frequent symptoms are cough, dyspnoea and chest pain. The can cause paraneoplastic syndromes: Hypoglycaemia due to production of insulin like substances, hypertrophic pulmonary osteoarthropathy and drumstick fingers (10-20 %) that disappear after tumour surgery.\(^1,5\) The treatment of choice is surgery, with complete resection of both benign and malignant masses and perioperative mortality is low (0-1.5 %).\(^1\)

References


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