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Intrapulmonary Askin Tumour: an Unusual Form of Presentation

Tumor de Askin in trapulmonar: Una forma inusual de presentación

To the Editor:

Askin in 1979¹ drew attention to the existence of an aggressive malignant tumour of the chest wall that affected young people; currently this tumour is included within the classification of peripheral primitive neuroectodermic tumours (pPNET), and their most frequent location is the chest wall.² We present a case of a pPNET intrapulmonary tumour that does not involve the chest wall.

Man of 75 years of age being studied for COPD who showed in a chest X-ray (not shown) a tumour in the right lung apex. The patient did not report chest pain. A chest CT was performed (fig. 1), and in the apical segment of the right upper lobe a solid tumour was seen, with a spiculate contour, of 3.5 cm in diameter, with no associated adenopathies or pleural effusion. After surgery, the anatomopathological diagnosis was of a primitive neuroendocrine

tumour (pPNET-Askin tumour). The patient received adjuvant chemotherapy treatment (adriamycin, vincristine and cyclophosphamide) and died at 6 months.

The term pPNET is currently used to describe a family of tumours that are characterised by a specific chromosome translocation, t(11;22)(q24;p12), and that present a variable degree of neuroectodermic differentiation characteristics.³ The associated clinical symptoms are usually chest pain (39%), deformity or palpable mass, and these tumours have been described in association with other haematological type neoplasias such as Hodgkin's disease.⁴

On X-ray it presents as a soft tissue mass on the chest wall that can be associated with costal erosion and pleural effusion, and is easily diagnosed by CT and MR; which furthermore, make it possible to determine its extension, and to assess the effects of chemotherapy and possible recurrences after surgery.³ This type of neoplasia of the chest wall is not frequent and intrapulmonary locations (such as we found) have been very rarely reported.⁵

The definitive diagnosis of these tumours is carried out by means of an anatomopathological study of the surgical specimen. Treatment consists in radical resection of the tumour, accompanied by chemotherapy with or without radiotherapy. Local or distant recurrences after surgery have been described and their prognosis is generally extremely poor.^{2,3}

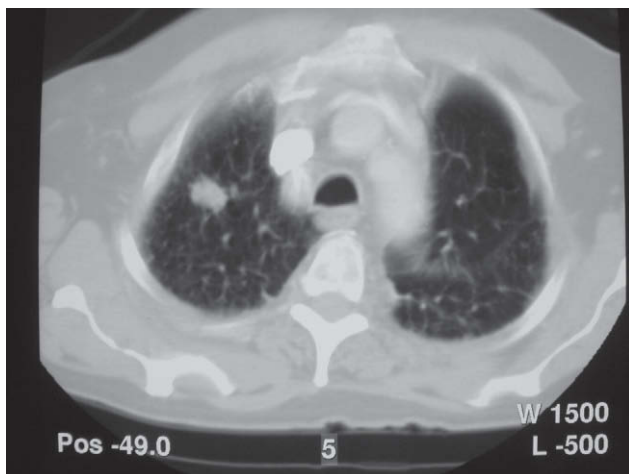


Figure 1. Chest CT: Tumour with spiculate contour in right lung apex with no pleural contact.

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