Un tumor intrapulmonar “out of the box”

An intrapulmonary tumor out of the box

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ABSTRACT

Primary intrapulmonary thymomas (PIT) are very rare tumors of unclear pathogenesis. We report a case of a 60-years-old woman who present with an accidental finding of a mass on her chest. Histology analysis shown to be primary intrapulmonary thymoma and she underwent surgical resection, remaining currently asymptomatic and without evidence of recurrence or metastasis. Since its rarity, reporting these cases is essential to better understand its clinical course and management.

Keywords: Thymus neoplasms, thymoma, lung, ectopic.

RESUMEN

Los timomas intrapulmonares primarios (TIP) son tumores muy infrecuentes de patogenia incierta. Presentamos el caso de una mujer de 60 años que presenta el hallazgo accidental de una masa en el tórax. El análisis histológico demostró que se trataba de un timoma intrapulmonar primario y fue sometida a resección quirúrgica, permaneciendo actualmente asintomática y sin evidencia de recurrencia o metástasis. Dada su rareza, la comunicación de estos casos es esencial para comprender mejor su curso clínico y manejo.

Palabras clave: Neoplasias del timo, timoma, pulmón, ectópico.

CLINICAL CASE

A woman of 60 years old was referred to our department for evaluation of an abnormal mass incidentally detected on chest x-ray (Figure 1). Her only medical known condition was a small uterine fibromysma. She was not taking any medications and has never smoked. Although she was asymptomatic, at examination she had a decreasing in pulmonary breath sounds over the left lung and rales slightly heard in left base.

Her laboratory analysis, including tumor marker levels, were unremarkable. CT chest shown a heterogeneous mass on the half lower of the left lung (10x15 centimeters), provoking collapse of the left lower lobe bronchus; no adenopathies was identified. PET/CT scan revealed slight ¹⁸F-fluorodeoxyglucose uptake (SUVmax value of 4) in this lesion, without any other significant sites of abnormal metabolic uptake along with no evidence of other lesions in the anterior mediastinum. In flexible bronchoscopy was seen a light reduction in diameter of the left inferior lobar bronchus by convergence of folds, whilst cytopathological examination of the bronchial brush, bronchial washing and bronchial biopsies were negative. It was then performed a transho- racic needle biopsy which histopathological examination was consistent with primary intrapulmonary thymoma, type B1 (according to WHO classification). The evaluation was complemented with CT angiography with cardiac gating, which shown that the mass was vasculared by branches from left internal mammary artery and branches from left brachiocephalic arterial trunk (Figure 2). The patient underwent thymectomy resection by left thoracotomy. The histology of the surgical sample confirmed a tumor 15cm length, encapsulated, without signs of invasion and surgical margins without neoplasm, compatible with Primary Intrapulmonary Thymoma type B1 (WHO classification), pT1aN0M0R0, and Masaoka-Koga clinical stage I.

In 2 years of follow-up, the patient has remained asymptomatic, with favorable evolution and no evidence of recurrence.

DISCUSSION

Primary intrapulmonary thymomas (PIT) are a very rare phenomenon and to the best of our knowledge there have been reported less than 40 cases since 1951, when it was first described by McBurney et al₁. They are defined as intrapulmonary tumors with the typical histological features of thymoma, yet without evidence of a thymic lesion in the anterosuperior mediastinum. Its pathogenesis is unclear, but it was hypothesized that an embryological displacement might account for the occurrence of ectopic thymomas.

Since its clinical presentation is non-specific and its location unexpected, the diagnosis of these type of tumor can be extremely challenging and they may often be misdiagnosed at first as other more common pathologic processes. In this case the patient was asymptomatic, and the diagnostic workflow just arose after a routine chest x-ray had shown a huge mass. Fine needle aspiration and core needle biopsy are commonly part of the initial diagnostic workup of these lesions, although in some cases they were correctly diagnosed solely after surgical resection.

Because of its low incidence, clinical studies about treatment and outcome are very limited and there are no standard guidelines for PIT. Its treatment approaches are then largely based on that of anterior mediastinal thymomas, which comprises complete surgical resection as the best treatment strategy. Adjuvant radiotherapy and/or chemotherapy have been suggested in specific inoperable cases or if there was invasion of local structures or incomplete resection were performed.

Although PIT has usually an indolent course when completely resected, there are a few cases reported of ectopic thymomas with distant metastasis and tumor recurrence, therefore, keeping close and long-term follow-up is crucial in these cases.
BIBLIOGRAPHY


