

## Letters to the Director

### Extramedullary Plasmacytoma That Simulates Pulmonary Metastasis

#### *Plasmocitoma extramedular que simula una metástasis pulmonar*

To the Editor:

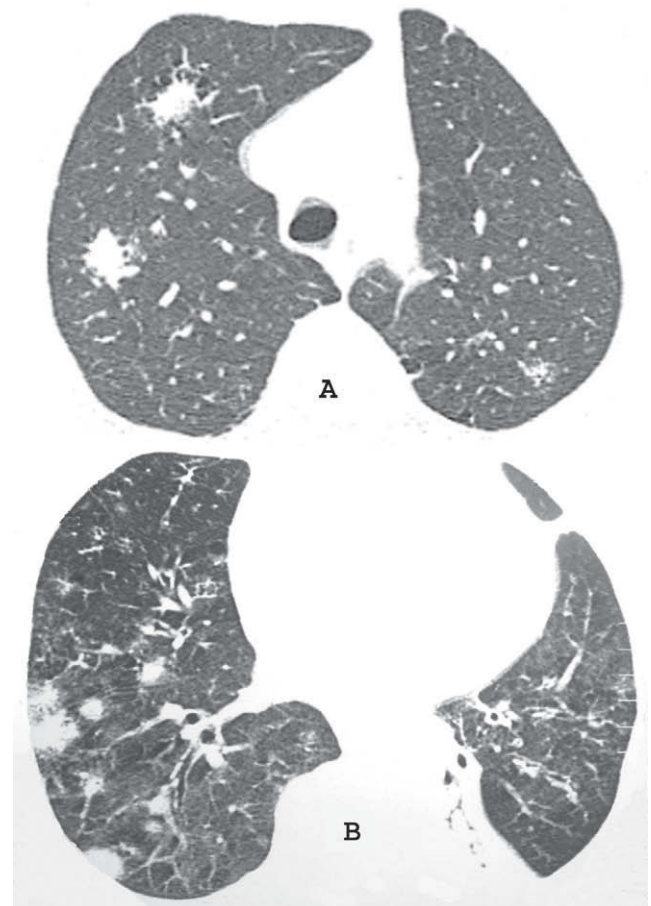
We read with great interest the report of three cases of extramedullary plasmacytoma (EMP) published by Montero et al.<sup>1</sup> In all the cases, the radiological presentation was of a tumour adjacent to the bronchi, with or without thickening of the lymph nodes, which simulated lung cancer. The final diagnosis was obtained through lung biopsy.

Recently, we observed a case of EMP with different radiological findings. The patient was a 66-year-old woman with progressive dyspnoea on exertion, dry cough and malaise, but without signs of fever or other symptoms. The patient reported having lost some 10 kg over the previous year and had a history of tuberculosis, for which she had received standard treatment six years earlier. The physical examination was normal, except for the presence of bilateral crepitanant rales.

Computed tomography (CT) showed nodular opacities with poorly defined margins in both lungs, particularly in the right lung, and atelectasis in the lower left lobe with bronchiectasis (fig. 1). The laboratory tests were normal. The fiberoptic bronchoscopy showed stenosis in the lumen of the lower left lobe without mucosal lesions. The bronchoalveolar lavage was negative for neoplastic cells, fungi and mycobacteria. The bronchial brushing and biopsy results were negative for malignant neoplasm. The open lung biopsy revealed proliferation of well-differentiated plasma cells. The immunohistochemistry showed tumour cells that were positive for kappa light chains. No Bence-Jones proteins were detected in the urine. An examination of the bones and a bone marrow biopsy provided negative results for myelomatosis. The patient received chemotherapy, and three years after the diagnosis, she is well and without symptoms.

The diagnosis of EMP is based on the discovery of monoclonal plasma cell tumour in an extramedullary site and of clonal cell tumour in the bone marrow of less than 10%.<sup>2</sup> It is unusual that the primary origin is the lung, although some cases have been reported. Almost all cases of pulmonary plasmacytoma are presented as solitary nodular mass lesions.<sup>1-3</sup> As far as we are concerned, only one case of EMP with multiple nodules has been described,<sup>4</sup> and unlike our patient, that case was associated with mediastinal mass.

Surgical resection is the best treatment for localised pulmonary plasmacytomas, which occasionally is combined with chemotherapy or radiotherapy. Plasma cell tumours are radiosensitive; however, the number of therapeutic options is reduced in patients with diffuse lung involvement, since the surgery and radiotherapy are not feasible.<sup>3,5</sup> Diffuse multiple pulmonary plasmacytoma, as seen in our patient, can be controlled effectively through a combination of chemotherapeutic agents treatments. The prognosis of patients with



**Figure 1.** CT scans of the upper (A) and lower (B) lobes, showing ill-defined nodules of varying sizes, predominantly in the right lung, and atelectasis in the lower left lobe.

EMP is generally better than that of patients with myelomatosis. Unlike solitary myelomas, EMP develops into myelomatosis in very few cases.<sup>4</sup> Our patient was treated with chemotherapy, and during the course of three years, she did not experience any growth, and her condition remained clinically stable.

#### References

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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 1979<sup>1</sup> 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.<sup>2</sup> We present a case of a pPNET intrapulmonary tumour that does not involve the chest wall.

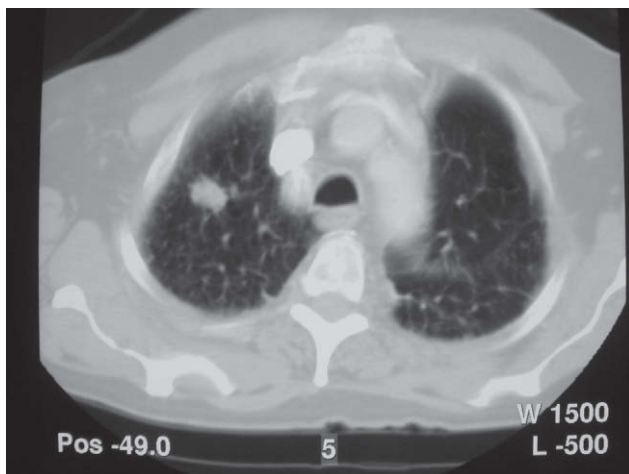
Man of 75 years of age being studied for COPD who showed in a chest X-ray (not shown) a tumour in the right lung apex. The patient did not report chest pain. A chest CT was performed (fig. 1), and in the apical segment of the right upper lobe a solid tumour was seen, with a spiculate contour, of 3.5 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.<sup>3</sup> 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.<sup>4</sup>

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.<sup>3</sup> This type of neoplasia of the chest wall is not frequent and intrapulmonary locations (such as we found) have been very rarely reported.<sup>5</sup>

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.<sup>2,3</sup>



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

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