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Scientific letter

Posterior Mediastinum Plasmocytoma: An Infrequent Location☆

Plasmocitoma del mediastino posterior, una localización poco frecuente

Dear Editor,

We present the case of a patient with a rapidly growing intrathoracic plasmacytoma, which is a particularly interesting location due to its rarity. Extramedullary plasmacytoma (EMP) is a plasma cell neoplasm affecting soft tissues without bone marrow involvement. Other myeloma multiple's systemic characteristics should be also excluded.[1–3] Usually, EMPs have a good prognosis but our patient did not have a final good outcome.

A 69-year-old male, ex-smoker and with history of spontaneous pneumothorax 2 years ago, was admitted to the hospital with persistent chest pain. In the last year, he had presented episodes of pleuritic chest pain in the right hemithorax and occasional low-grade fever associated with night sweats. In the last 3 months he started with intermittent grade III mMRC dyspnea. He visited several times the emergency department without finding signs of new episodes of pneumothorax or any other pathology.

Careful physical examination was taken, not finding palpable lymphadenopathy. Chest X-ray showed a lower right paravertebral mass, not visible on the radiological study taken 2 months ago. Thoracic computed tomography (CT) scan showed an extraparenchymal mass of 10 cm × 9 cm × 7.2 cm, located in the posterior mediastinum, near the right posterior-paravertebral costophrenic sinus.

The biopsy was compatible with high proliferation plasma cell neoplasia. Immunohistochemistry revealed CD138 + and EMA + tumor cells; with high proliferative activity, estimated with KI-67 (Fig. 1a, b).

He was referred to the Hematology Service to rule out multiple myeloma. Blood counts, calcemia, and renal function were normal. Monoclonal component was not detected in the serum protein electrophoresis, neither Bence-Jones protein in the urine. No lytic lesions were observed on CT scan. A positron emission tomography (PET) scan showed hypermetabolism in the mass without evidence of increased metabolic activity at another level (Fig. 1c, d). Bone marrow biopsy did not show plasma cell infiltration. Therefore, multiple myeloma was ruled out and the diagnosis of solitary plasmacytoma was confirmed.

Firstly, the patient was treated with a bortezomib– dexamethasone (Vd) chemotherapy regimen to reduce tumor size. He achieved initial clinical improvement, with cessation of pain. But a few months later, he did not obtain a good response to chemotherapy, with a significant increase in tumor mass and multiple complications such as respiratory infections, pleural and pericardial effusion, and pulmonary embolism. The patient received a second-line chemotherapy treatment alternating vincristine, carmustine, melphalan, cyclophosphamide and prednisone, with vincristine, carmustine, doxorubicin and dexamethasone (VBMCP/BVAD). It was not sufficient to combat the tumor. Consequently, the patient did not receive radiotherapy and decided to continue with palliative treatment for pain and symptoms management.

In summary, our case is an intrathoracic extramedullary plasmacytoma that arise in posterior mediastinum.

Extramedullary plasmacytomas (EMPs) represent 3–5% of plasma cell neoplasms.[1,4] They can appear in any part of the body, being more frequent in head and neck, upper airway and gastrointestinal tract.[5] Although there are cases described in the thorax, the number of EMP found in the low tract (including trachea, bronchi and lungs) and mediastinum is limited.[6–8] Therefore, the diagnosis may be delayed due to absence of clinical suspicion.

Tumors in the posterior mediastinum are usually neurogenic, lymphomas or non-neoplastic lesions, being plasmacytoma a rare cause.[2] EMPs are more frequent in men between 50 and 60 years old.[9] However, there are also cases described in younger individuals.

The usual clinical presentation is a mass located in soft tissues that generates symptoms related to compression of nearby structures. It can spread locally, even presenting at diagnosis with metastatic lymph node involvement in up to 40% of cases.[10] Our case debuted with chest pain appearing some months earlier, with occasional low-grade fever.

PET-CT is considered the technique of choice for the detection of a solitary EMP, to evaluate the response to treatment and for subsequent follow-up.[5] It also has an important role in prognosis with high metabolic uptake related to larger masses and a higher risk of progression.

EMPs can associate a monoclonal component in less than 30% of the cases, and it usually disappears at the start of treatment. Most EMP are located in respiratory and digestive mucosa where IgA is produced. Therefore, it would be reasonable to expect predominance of IgA type myelomas.[4]

In up to 5% of cases, EMPs can be found at the same time as multiple myeloma, increasing up to 30% in case of mediastinal mass.[1,9] Subsequent progression to multiple myeloma within two years can occur in up to 30% of cases,[3,5] being more likely in older patients, larger tumors, and patients with monoclonal component (especially if the latter persist > 1 year after treatment).[11] Although multiple myeloma is ruled out at initial diagnosis, as in our case, follow-up and subsequent studies are necessary.

☆ This manuscript does not require either approval from an Ethics committee or informed consent from the patient as only anonymous clinical records have been used.

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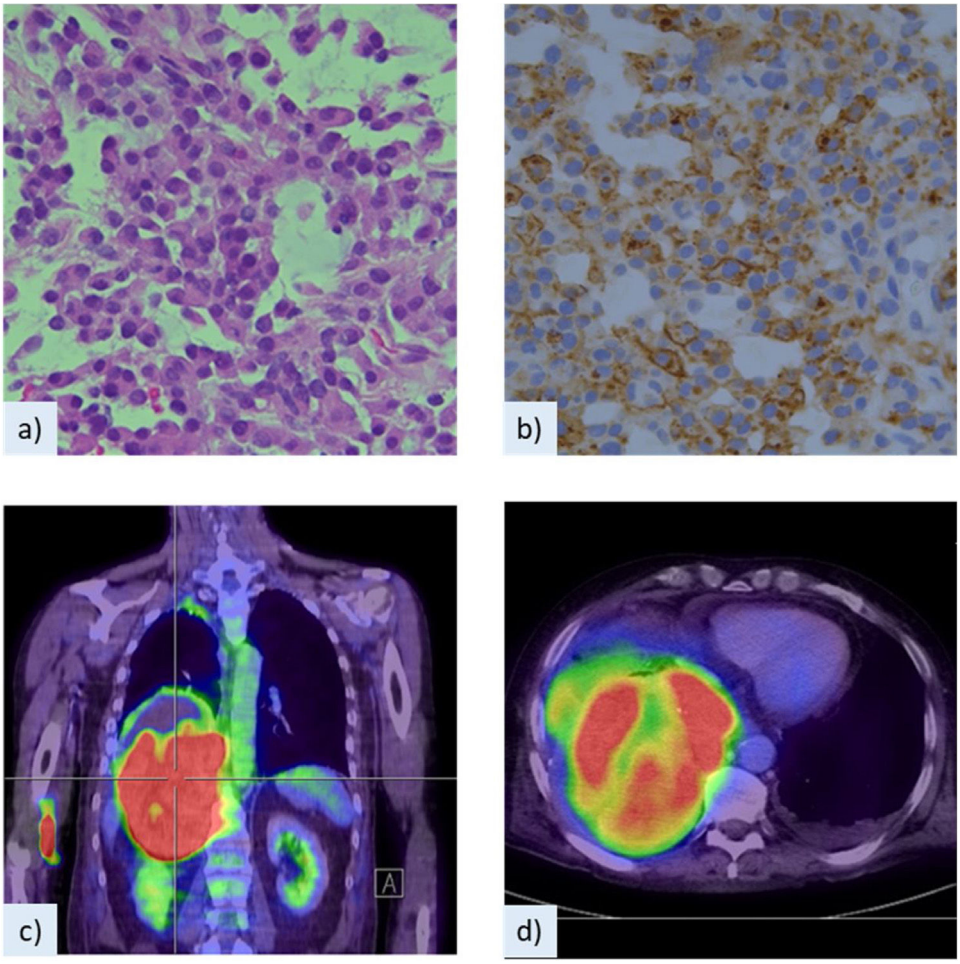

Fig. 1. (a) Microscopic examination showing infiltration of plasma cells (hematoxylin and eosin stain). (b) Immunohistochemical staining revealed CD138+ and EMA+ tumor cells. (c, d) PET-TC with increased metabolic uptake in the mass.

EMPs are highly radiosensitive and normally have a good response to it with a favorable prognosis. Other treatment options are chemotherapy or surgery. There is controversy in the literature about which treatment is the best one, considering equally valid the combination of different options. In our case, the patient received firstly chemotherapy to reduce the tumor's size. Due to poor outcome, it was not possible to start RT as a curative treatment and symptomatic palliative treatment was finally chosen.

Usually, EMPs have a good prognosis with a 5-year survival rate ranges from 70% to 90%.[10] However, after treatment of EMP, up to 20–30% of cases can develop local recurrence.[3,7] Some studies claim that head and neck plasmacytomas have a better prognosis, perhaps due to their good response to RT; however, others do not consider the location as a relevant factor, observing a similar evolution. Nevertheless, metastatic lymph node involvement and monoclonal component is clearly related with worse prognosis and a more aggressive treatment is recommended.[10]

In conclusion, plasmacytoma should be included in the differential diagnosis of a mass in the posterior mediastinum. In addition, we should take into account its association with multiple myeloma and the possibility of progression. This is why multiple myeloma has to be ruled out at the moment of diagnosis and on top, subsequently to carry out a careful and long-term follow up of it.

Informed consent

This consent has been obtained.

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Authors' contributions

All the authors have contributed substantially to the elaboration of the manuscript.

Conflicts of interest

None.

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