CASE REPORT

A case of disseminated recurrent retroperitoneal plasmacytoma with left obstructive hydronephrosis

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SUMMARY

Retroperitoneal extramedullary plasmacytoma (EMP) is a rare condition that often poses a diagnostic challenge. This is mainly due to its unusual location and non-specific symptoms especially in its early stages. Retroperitoneal plasmacytoma with renal involvement is an extremely rare entity and to date, less than 25 cases have been reported in the literature. We describe a case of a 65-year-old woman with primary retroperitoneal plasmacytoma, confirmed by ultrasound-guided biopsy and histopathological examination, with unilateral left obstructive hydronephrosis and metastasis to the lungs. Our patient was started on chemotherapy to which she did not respond. Subsequently, she resorted to palliative measures. Our case highlights key aspects of clinical presentation, diagnosis and treatment options available for management of retroperitoneal plasmacytoma.

BACKGROUND

Extramedullary plasmacytoma (EMP) is a neoplastic proliferation of plasma cells in non-osseous sites without the involvement of bone marrow. It accounts for about 3% of all the plasma cell neoplasms. The majority of EMPs occur in the head and neck region¹ and retroperitoneal involvement is rare.^{2–3} They often pose a diagnostic challenge because of their unusual location and absence of symptoms until advanced stages. A high index of suspicion is required for the diagnosis of this condition at an earlier stage. Small and confined EMPs are usually highly radiosensitive.^{4–5}

CASE PRESENTATION

A 65-year-old Caucasian woman, a heavy smoker with a medical history of EMP of the retroperitoneum, presented with a 1-month long history of progressively worsening abdominal pain, discomfort and generalised weakness. Two years ago, she was diagnosed with retroperitoneal plasmacytoma. On diagnosis, the tumour measured 4.9 cm×3.8 cm and involved mainly the left side. She had a complete response to radiotherapy.

Currently, she denied any history of fever, night sweats, nausea or vomiting. However, she did have significant anorexia along with a 15 kg weight loss over the past 3 months. She denied any alcohol or drug use. There was no family history of malignancy in any of her first-degree relatives.

Physical examination revealed an ill-appearing woman with stable vital signs. There was no icterus, pallor or lymphadenopathy. Heart and lung examination were within normal limits. Abdominal examination revealed distension and fullness in the

left flank region. The abdomen was soft, non-tender without any hepatosplenomegaly. Bowel sounds were normal. Neurological examination was grossly intact.

INVESTIGATIONS

Laboratory tests revealed white cell count of 4900/ μ L, haemoglobin 10.4 g/dL and platelet count 375 000/ μ L. Glucose, electrolytes, renal and liver function tests were within normal limits. Urinalysis revealed no Bence-Jones proteinuria. CT of the abdomen and pelvis (figure 1) showed a 10 cm×16 cm left paraspinal soft tissue mass at the level of left renal hilum with significant hydronephrosis. CT of the chest showed a 6.1 cm×4.6 cm left hilar mass (figures 2 and 3). Bone marrow biopsy revealed normocellular marrow with trilineage haematopoiesis with less than 10% plasma cells, non-diagnostic for multiple myeloma. Fine-needle aspiration cytology of the left retroperitoneal mass was positive for plasmacytoma. A complete skeletal survey was negative for osteolytic lesions. Serum IgA level was elevated at 1150 g/L (normal range 0.76–3.90 g/L) with normal IgG and IgM levels. Immunohistochemistry was positive for CD138 and CD38 but negative for CD45, CD19 and CD20.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses of lymphoplasmacytic lymphoma and immunoblastic lymphoma were considered but were eventually ruled out by characteristic findings on immunohistochemistry.

TREATMENT

Although she had complete remission with radiotherapy during the previous episode, radiotherapy was not considered this time because of large tumour size along with metastases to the lungs. Chemotherapy was initiated with daily oral lenalidomide 25 mg (for days 1–21 of 28-day cycle) and intravenous dexamethasone 4 mg every 6 h with an initial dose of 10 mg.

OUTCOME AND FOLLOW-UP

The patient did not respond well to chemotherapy. Ultimately, she resorted to comfort measures due to poor prognosis.

DISCUSSION

EMP is a rare tumour with a worldwide annual incidence of 3/100 000 populations. It is three times more common in men compared with women.³ Retroperitoneal plasmacytoma is an even

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Figure 1 CT of the abdomen and pelvis with 16×10 cm left paraspinal soft tissue mass (× for approximate dimension) at the level of left renal hilum (white arrow).

rarer entity. Owing to the rarity of this entity, epidemiology including prevalence, age and sex distribution has not been well defined in the literature. In published case reports, involvement of variable age groups, ranging from 25 to 70 years, has been described.^{2 3 5–7}

Retroperitoneal plasmacytoma can present with abdominal pain or discomfort, back pain, obstructive jaundice,⁶ renal failure if there is invasion of the bilateral kidneys.² Our patient had significant left-sided hydronephrosis, indicating compression of the left ureter by the mass. She did not have renal failure at the time of presentation. We believe that her serum creatinine was normal due to the non-involvement and normal functioning of the right kidney. Unfortunately, excisional biopsy could not be performed to see whether there was invasion of the kidney because of large size of the tumour and patient refusal.

The diagnosis of EMP is complex requiring multimodal approach involving radiological, haematological, biochemical and histopathological testing.⁷ EMP should be suspected in cases when a retroperitoneal mass is identified.⁵ CT scan with/without positron emission tomography scan or MRI may show evidence of mass at the specific location and its extension. Definitive diagnosis requires histopathological examination of the biopsied specimen and immunohistochemistry.^{5 6} Immunohistochemistry not only supports the diagnosis but also helps in ruling out other diagnoses such as lymphoplasmacytic lymphoma and immunoblastic lymphoma.⁸ CD138 and CD38 are characteristically positive in plasmacytoma⁹ and failure to stain for CD20 and CD45, as seen in our case, is highly characteristic for plasmacytoma.¹⁰ Once the diagnosis of EMP has

Figure 2 CT of the chest showing 6.1×4.6 cm left hilar mass (white arrow) in lung window.

Figure 3 CT of the chest showing 6.1×4.6 cm left hilar mass (white arrow) in mediastinal window.

been established, a complete blood count, serum and urine protein electrophoresis, immunoelectrophoresis, skeletal survey and bone marrow examination should be performed to rule out multiple myeloma.⁷ Our case did not meet the criteria for multiple myeloma as there were less than 10% plasma cells on bone marrow biopsy, lack of osteolytic lesions on skeletal survey and absence of end organ damage along with normal calcium and creatinine levels.

EMP, when small in size and detected early, is usually sensitive to local radiotherapy (40 to 50 grey over 4 weeks) leading to complete remission in most of the cases.⁶ Although highly radio-sensitive, radiotherapy to a large retroperitoneal plasmacytoma is associated with significant morbidity.⁵ Current literature supports use of radiotherapy^{11 12} or combined radiotherapy and surgery depending on the resectability of the tumour^{7 13} or combined radiotherapy and chemotherapy.¹⁴ Usual chemotherapy regimens suggest use of bortezomib as one of the newer agents or lenalidomide along with dexamethasone. In our case, because of the tumour size with metastasis to the lungs, local radiotherapy was no longer an option. Systemic palliative chemotherapy was undertaken to which our patient did not respond.

Owing to the rarity of the condition, there are no clear guidelines for follow-up. Periodic follow-up of the patients with retroperitoneal plasmacytoma is crucial due to the probability of relapse as seen in our case and progression to multiple myeloma.⁷

Learning points

- ▶ Retroperitoneal extramedullary plasmacytoma is an extremely rare entity and early diagnosis necessitates high index of suspicion.
- ▶ Biopsy and immunohistochemistry are essential to confirm the diagnosis.
- ▶ Treatment options include local radiotherapy alone or in combination with surgery and/or chemotherapy with varying results.

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literature search and discussion section of the manuscript. SJ reviewed and edited the final draft of the manuscript.

Competing interests None.

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