Mesenteric plasmacytoma: An unusual cause of an abdominal mass

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Plasmacytoma is a rare plasma cell tumour that arises from plasma cells. The tumour accounts for about 3 - 5% of all plasma cell malignancies and most often affects the head and neck, but may also occur in the gastrointestinal tract. To our knowledge, mesenteric plasmacytoma has not been described previously. We describe the presentation and management of a case of mesenteric plasmacytoma presenting as an abdominal mass in a 69-year-old HIV-positive man.

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Plasmacytoma is a rare plasma cell tumour that arises from plasma cells and accounts for about 3 - 5% of all plasma cell malignancies.[1] These tumours most often develop in the head and neck, but may also occur in the gastrointestinal tract. Extramedullary plasmacytoma is a rare cause of an intra-abdominal mass. The gastrointestinal tract is involved in 10% of cases, with the stomach and small intestine being the commonest sites.[1] The colon is very rarely affected.[2] We present the case of a 69-year-old man who presented with an abdominal mass. A preoperative ultrasound-guided fine-needle aspirate showed it to be a plasmacytoma, and this was confirmed by histological examination of the excised tumour.

Case report

A 69-year-old HIV-positive man who was not on antiretroviral therapy and had an absolute $CD4^+$ count of 632 cells/μl presented to the Department of Medicine at Chris Hani Baragwanath Academic Hospital, Johannesburg, South Africa, complaining of abdominal pain and constipation. Systemic examination revealed shotty lymph nodes in the neck, axillae and groin. Abdominal examination revealed a mobile, solid, right paraumbilical mass with no tenderness or abdominal distension. The rest of the findings were unremarkable. Admission laboratory results showed a serum urea level of 8.2 mmol/l, a creatinine level of 166 μmol/l, a normal calcium level and a markedly raised total protein level of 98 g/l. The albumin level was 34 g/l. Subsequent protein electrophoresis demonstrated a monoclonal band (gammaglobulinaemia 29.50 g/l).

An abdominal ultrasound scan showed a mass with mixed echogenicity measuring 96 × 57 × 66 mm, which was separate from vessels and the vertebral bodies. An ultrasound-guided fine-needle aspirate revealed a monomorphic population of abnormal plasma cells suggesting a plasma cell dyscrasia. On referral to the Department of Surgery, a computed tomography (CT) scan of the abdomen and colonoscopy were performed. The CT scan revealed a poorly defined heterogeneous mass near the uncinate process of the pancreas, with a poor plain of separation between the mass, the third part of the duodenum and the superior mesenteric vein. No common bile duct or pancreatic duct dilatation was evident. Multiple sub-centimetre mesenteric nodes (that may not be picked up on CT because they are too small) were present. The liver and spleen were normal. There were no lytic lesions in the spine or pelvis, and no clinical or radiological signs of obstruction. The findings on colonoscopy were normal.

A mobile mass measuring 125 × 90 × 90 mm arising from the mesentery of the proximal small bowel was found at exploratory laparotomy (Fig. 1). The liver, spleen and peritoneum appeared normal. The mass was excised with margins including an area of mesentery, 260 × 25 × 25 mm of jejunum and a regional lymph node. Primary anastomosis was performed. The cut surface of the mass revealed multiple nodularity and multiple areas of haemorrhage. Histological examination subsequently confirmed a neoplasm comprising sheets and circumscribed nodules populated by mature plasma cells. Areas of coagulative necrosis were present within the tumour. No morphological evidence of plasmablastic lymphoma was identified. The small-intestine specimens showed no obvious neoplastic infiltration, but prominently dilated lymphatic channels were present throughout the wall of the small intestine. The lymph node had an atypical plasma cell infiltrate similar to that seen in the excised mass. Bone marrow aspirate and trephine biopsy showed no neoplastic infiltration. The patient's postoperative course was uneventful.

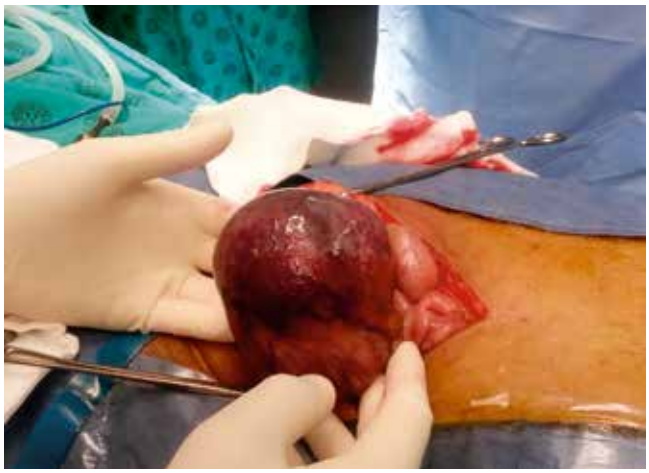

Fig. 1. Image taken intraoperatively showing the mass arising from the mesentery of the small bowel.

Discussion

Plasmacytomas are rare tumours arising from plasma cells and are divided into solitary bone plasmacytomas (SBPs) and extramedullary plasmacytomas (EMPs), with the latter accounting for less than 3% of all plasma cell tumours.[1] SBPs account for less than 5%.[1] EMPs most commonly occur in the upper gastrointestinal tract or upper airways (80 - 90%) and often cause symptoms of upper airway obstruction, swelling and epistaxis.[2] Presentation of the tumour at other sites depends on the site and the tissues involved.

The differential diagnosis includes monoclonal gammopathy of undetermined significance (MGUS), SBP and multiple myeloma. MGUS is a condition resembling multiple myeloma in which a paraprotein is found in the blood.[3]

Multiple myeloma is characterised by monoclonal proliferation of plasma cells in either bone or soft tissue and over-expression of monoclonal paraprotein (M protein). These proteins give rise to complications such as renal failure. Local symptoms caused by tumour mass invasion include bone pain and spinal cord compression. Hypercalcaemia is a common associated feature.

Extramedullary disease involves discrete, neoplastic proliferation of monoclonal cells and tends to spread locally.[1] Diagnostic criteria for EMP include a tissue biopsy showing monoclonal plasma cell activity, absence of evidence of multiple myeloma, a low or absent M-protein concentration and absence of hypercalcaemia or renal failure.

Laboratory tests should include plasma electrophoresis to exclude M proteins, which are present in 14 - 25% of cases. A tissue biopsy is necessary to demonstrate plasma cells, and a bone marrow aspirate and trephine biopsy should demonstrate no clonal plasma cells. Imaging should include ultrasound, CT imaging of the upper aerodigestive and gastrointestinal tracts and complete bone imaging (CT or magnetic resonance imaging) of the spine and pelvis.

The disease is staged according to the Wiltshaw classification[4] as follows: I – limited to extramedullary site; II – involvement of regional lymph nodes; III – metastatic disease. Our patient's regional lymph nodes were involved. It is important to note that the classification has a poor correlation with prognosis.

The prognosis after local treatment, either surgical or by radiation, is favourable. Plasmacytomas are sensitive to radiation therapy, and this is recommended as a treatment modality of choice. Surgical resection has similar results, and results in fewer side-effects if clear margins are obtained.[1] If the plasmacytoma is resected, regional lymph nodes should be removed. Incomplete resection should be managed with adjuvant radiotherapy rather than reoperation. Patients should be closely followed up for residual or metastatic disease, with investigations including monitoring of serum and urine protein levels, skeletal radiological studies, abdominal ultrasonography and bone marrow studies.[1]

The prognosis for EMP is good, with a low risk of development of multiple myeloma. A median survival of approximately 10 years has been reported, with 56% of patients being free of systemic disease at 5 years.[5] SBP, however, has a higher risk of progression.[6]

Conclusion

Mesenteric plasmacytoma is very rare, and to date there has been no published literature on plasmacytoma of the small-bowel mesentery. However, the prognosis of EMP is good. It is important to note the difference between EMP and multiple myeloma, as the treatment and prognosis differ. Follow-up is crucial, as transformation to multiple myeloma can occur. Local treatment, either surgical or by radiation, has a favourable prognosis.

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