CASE REPORT

Extramedullary Plasmacytoma Presenting with Acute Abdomen

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ABSTRACT

Multiple Myeloma (MM) involving the Gastrointestinal (GI) Tract is an extremely rare entity. A 34 year old man with history of MM for 3 years who had chemotherapy and allogenic bone marrow transplant presented with an acute abdomen. CT scan showed pneumoperitoneum with thickened ileal loops. Laparotomy confirmed ileal perforation. The ileal segment was resected and an end loop ileostomy was fashioned. Histopathology confirmed a diagnosis of Plasmacytoma.

Keywords: Extramedullary plasmacytoma, plasma cell myeloma of small bowel

CASE REPORT

A 34 year old Caucasian male presented with an acute abdomen. He had a history of MM (Lambda light chain Multiple Myeloma) diagnosed 3 years previously. He suffered from multiple relapses despite different chemotherapy regimens. The patient had an allogenic BMT (Bone Marrow Transplant) a year ago. CT Scan performed 2 months previously for similar presentation showed terminal ileum thickening causing small bowel obstruction. A follow-up colonoscopy revealed ulceration of the terminal ileum and biopsy confirmed chronic GVHD (Graft vs Host Disease) which was thought to be a complication of his recent BMT. Bone marrow transplant is also complicated by cutaneous GVHD and renal failure. He was managed successfully with conservative treatment and discharged home. However, two months later, he was referred from haematology clinic and re-admitted with right lower abdominal pain and constipation. Repeat CT abdomen revealed distal small bowel obstruction with pneumoperitoneum necessitating emergency laparotomy. Intra-operatively there was grossly thickened and dilated small bowel with ischemic perforation. A small bowel resection was performed with the formation of end ileostomy. Postop recovery was uneventful. Histopathology confirmed plasma cell infiltration of resected small bowel specimen rather than GVHD contradicting the previous diagnosis. Two months later follow up PET scan showed residual plasmacytoma in small bowel loops adjacent to the stoma site. The patient presented 3 months later with bleeding from stoma site which was successfully treated with conservative management. The case was discussed at Multidisciplinary Team meeting where no further surgical intervention was advised. The pt is currently receiving chemotherapy.

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DISCUSSION

Plasma cells are derived from Hematopoietic stem cells in the Bone Marrow and form an integral part of our immune system. Any defect in the formation of plasma cells can lead to abnormal plasma cells production and can present in any of 4 four ways. MM, Solitary myeloma of the bone, plasma cell leukemia or EMP (Extamedullary Plasmacytoma).

EMP involving the GI Tract is an extremely rare entity that accounts for only 5% of all plasma cell tumors and occur mainly in the upper GI Tract[1]. EMP in the setting of MM involving the terminal ileum is an even rarer presentation[2]. Non specific abdominal symptoms, intra-operative findings and similar histological appearances with other tumors make it a diagnostic challenge for clinicians. Immunohisto-chemistry is a valuable diagnostic test that will confirm the diagnosis.

The treatment of EMP depends on the site of the disease, mode of presentation and whether primary or secondary in nature at the time of presentation. The treatment options must be offered in a multidisciplinary setting to patients diagnosed with EMP secondary to MM. The following four factors are important in planning treatment: tumor burden (staging), patient factors (age, performance status, comorbidities), disease biology, aggressiveness and response to therapy. The prognosis of MM involving the GI Tract is unfavourable[3].

For patients with EMP of small bowel, the indications of for surgical intervention include impending or frank obstruction, perforation[4] and bleeding. Surgical resection may be indicated for diagnostic purposes if a biopsy specimen cannot be obtained due to difficult access. However this needs to be balanced against the risks of surgery in potentially high risk patients[5].

CONCLUSION

EMP should be considered in the differential diagnosis of acute or chronic small bowel related presentations in patients with MM.

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