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A Rare Incidental Isolated Colonic Extramedullary Plasmacytoma

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Abstract:

Plasmacytomas are rare tumours that develop from B lymphocytes. In 10% of cases the gastrointestinal tract is involved, usually in the small intestine. We report an unusual case of colonic extramedullary plasmacytoma at the splenic flexure. This was an incidental finding in a patient being investigated with right groin discomfort and culminated in resection of the affected splenic flexure. 1 year following resection there is no evidence of disease recurrence.

Key words: Plasmacytoma, Colon, Intestine, Gastrointestinal Tract, Large Intestine, Humans.

Introduction

Plasma cell tumours develop from B lymphocytes and can be subdivided into plasmacytoma and multiple myeloma. The International Myeloma Working Group recognises three distinct groups of plasmacytoma; solitary plasmacytoma of bone, extramedullary plasmacytoma or multiple solitary plasmacytomas [1]. Extramedullary plasmacytomas are most frequently located in the upper respiratory tract [2]. In 10% of cases the gastrointestinal tract is involved, and usually in the small intestine [3]. We report an incidental finding of a case of solitary colonic extramedullary plasmacytoma in a patient presenting with right groin discomfort.

Case Report

A 68 year old lady was referred as an outpatient to the general surgery clinic with a 1 year history of intermittent right groin discomfort which had worsened over the preceding 2 months. There were no exacerbating or relieving factors and the patient did not report any lumps in the area with no alteration in her bowel habit or weight. Past surgical history included appendicectomy and hysterectomy. There was no significant medical history and she was not on any regular medications. She smoked 20 cigarettes per day and drank no alcohol. Clinical examination was unremarkable with no palpable inguinal or femoral hernia with a normal range of movement of the right hip. X-ray of the right hip and pelvis and ultrasound of the right groin were also normal.

In view of the non-specific nature of the patient's symptoms a CT abdomen and pelvis was requested. This demonstrated sigmoid diverticulosis with no

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evidence of herniae in the right groin. In addition, an incidental area of thickening and altered calibre of the colon at the splenic flexure was reported.

The patient went on to have a colonoscopy with a biopsy of patchy mucosal erythema at the splenic flexure. Histological examination demonstrated abnormal large bowel mucosa consistent with a neoplastic process of haematological origin. Immunohistochemistry raised the possibility of a plasma cell dyscrasia. FISH studies were suggestive of a plasmacytoma, multiple myeloma or mantle cell lymphoma.

Urine Bence Jones proteins, serum protein electrophoresis, monoclonal bands, bone marrow biopsy, renal functions, calcium and full blood counts, serum free light chains Kappa/ Lambda ratio were normal or negative. A haematology opinion was sought and the patient went on to have a normal staging CT of the chest, abdomen and pelvis with

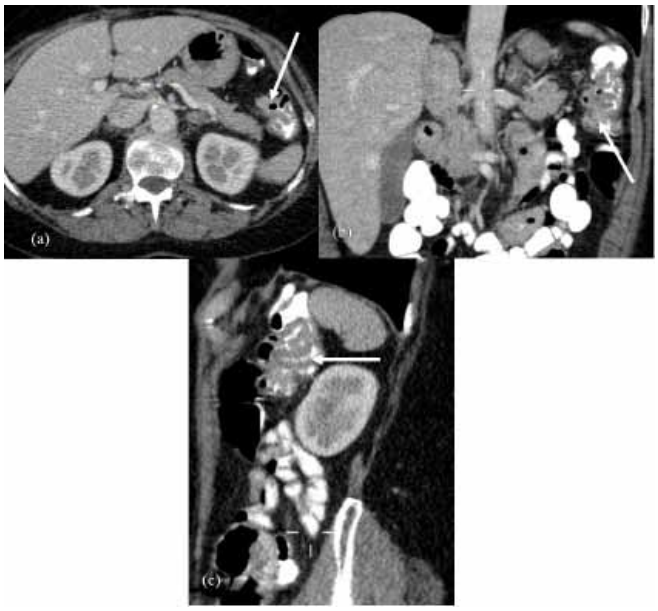

Fig.1: CT abdomen with oral and Intravenous contrast. (a) Axial, (b) Coronal and (c) Sagittal images showing thickened walls of large bowel at the level of splenic flexure (white arrows).

further random colonic biopsies to exclude the possibility of an underlying mantle cell lymphoma. The biopsies of the remaining colon were normal while histological appearances at the splenic flexure were similar to the previous occasion.

The consensus of opinion between the haematologist and oncologist was that resection of the lesion was indicated and an uneventful segmental resection of the splenic flexure was performed. The final pathology confirmed the diagnosis of an isolated extramedullary colonic plasmacytoma of the splenic flexure limited to the colonic mucosa with no lymph node involvement.

Six months following colonic resection the patient had a normal CT of the abdomen on account of

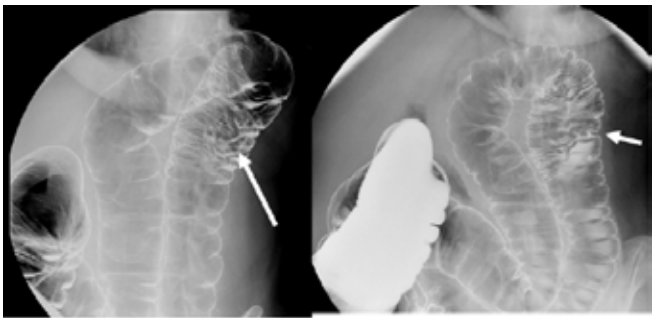

Fig.2: Barium enema images showing linear and nodular filling defects at the splenic flexure (white arrows).

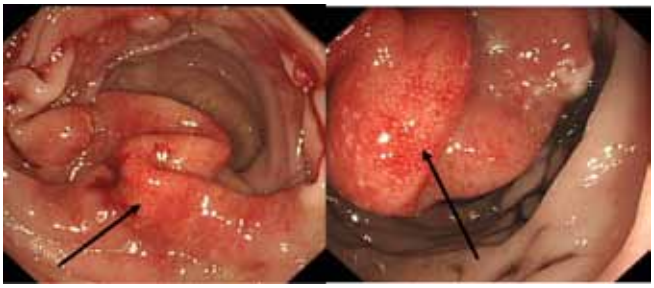

Fig.3: The colonoscopy images showing thickened erythematous mucosa at splenic flexure (black arrows).

non-specific left upper quadrant pain and a follow up colonoscopy was also unremarkable

Discussion

The International Myeloma Working Group and World Health Organisation have classified plasma cell tumours into two main subgroups: plasmacytoma and multiple myeloma [1,4]. Plasmacytomas may be limited anatomically to bone or outside the bone (extramedullary plasmacytoma). Clinicians are likely to gain some exposure to multiple myeloma at some point in their career, but are less likely to have experience of plasmacytoma, particularly extramedullary plasmacytoma (EMP) as this is rare. A diagnosis of EMP requires the exclusion of multiple myeloma following negative urinary Bence Jones protein, normal serum protein electrophoresis and bone marrow biopsy. The most common site of solitary EMP are within the upper respiratory tract but have also been described in a number of other locations e.g. lung [5], spleen [6], colon [7,8], thyroid [9].

Approximately 40 cases of solitary colonic EMP have been described in the literature to date. Presenting symptoms vary with anatomical location and may include diarrhoea and weight loss [7], abdominal pain [3,8] and rectal bleeding [10,11]. Only rarely these are incidental. Due to rarity of these tumours, there are no specific treatment guidelines. However, surgical treatment in the form of surgical resection is usually undertaken which is curative. Plasma cell tumours are radiosensitive and radiotherapy has in some cases been employed as an alternative to surgical resection with good effect [12]. Chemotherapy has also been employed in cases with associated systemic disease. The rationale for this is based upon the proposed staging system for solitary colonic EMP by Allison *et al*. [13].

Our case of primary colonic plasmacytoma was very unusual as it was diagnosed as an incidental finding in a female patient being investigated for nonspecific discomfort in the right groin. There were no features of colonic obstruction, bleeding or change in bowel habit. The initial abnormality at the splenic flexure was detected on CT of the abdomen and further investigated with barium enema then colonic biopsy. Histopathology raised the suspicion of a haematological pathology and specialist input was sought. The patient went on to have a segmental resection of her splenic flexure. Pathology of the resected specimen confirmed the suspicion of extramedullary plasmacytoma with no evidence of lymphatic spread and the resection margins were also clear. As a result, no further treatment was undertaken and after 1 year of follow up there is no evidence of disease recurrence.

In summary, isolated colonic plasmacytomas is a rare clinical condition specially when seen as an incidental finding. The radiological appearances are nonspecific but a high index of suspicion is required to suspect an abnormality as an incidental finding. The initial diagnosis can be difficult and management requires close collaboration between a number of specialists to ensure the correct diagnosis and treatment for best possible outcome for the patient concerned. Specialist involvement in this case included the expertise of a colorectal surgeon, haematologist, pathologist and radiologist. Colorectal surgeons will be unlikely to see this condition during their career but they should be aware of the nature of the condition and the importance of seeking specialist help prior to undertaking any form of surgery.

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