Intestinal pseudo-obstruction due to amyloidosis of the colon in association with an intestinal plasmacytoma

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Summary: A case of large bowel pseudo-obstruction due to colonic amyloidosis associated with an intestinal plasmacytoma is described. The association of an intestinal plasmacytoma with massive local amyloid deposition has not to our knowledge been previously reported.

Introduction

Amyloidosis is an extremely rare cause of mega-colon and intestinal pseudo-obstruction.\(^1\)\(^2\) Plasmacytoma of the large intestine is also a rare entity with some 18 published cases.\(^3\) The association between caecal mucosal plasmacytoma and massive local deposition of amyloid with intestinal pseudo-obstruction has not, to our knowledge, been described previously.

Case report

A 70 year old retired gardener presented with a 13 day history of constipation and increasing abdominal distension. For several years he had suffered from frequent bowel action 2–5 times daily. His problem had previously been attributed to intestinal hurry, following a selective vagotomy and pyloroplasty 19 years earlier for recurrent duodenal ulceration.

On examination, he was dehydrated with a palpable mass in the left iliac fossa and clinical and radiobiological signs of intestinal obstruction. Full blood count, ESR, biochemical profile and chest X-ray were normal. A provisional diagnosis of large bowel obstruction secondary to a mass in the descending colon was made.

At laparotomy, an acquired megacolon with a huge faecal mass, approximately the size of a melon, was found in the descending colon. The caecum looked necrotic. No tumour was found macroscopically and a subtotal colectomy with ileo-sigmoid anastomosis was performed.

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Figure 1  Colonic amyloid deposition. (Methyl Violet x 300).

Figure 2  Plasma cells. (Haematoxylin and Eosin x 786).

avidin–biotin immunoperoxidase technique. This showed them to be expressing IgA with lambda light chain restriction indicating that they were monoclonal. Immunohistochemistry revealed the amyloid to be of an AL lambda type with negative staining for A protein.

Although no large discrete tumour mass was present, the diffuse monomorphous plasma cell infiltrate and light chain restriction indicated that this was an extramedullary caecal plasmacytoma. No lymphoepithelial lesions, such as small centrocyte-like cells infiltrating crypts, characteristic of other intestinal non-Hodgkin’s lymphomas could be seen. The colonic lymph nodes identified showed reactive changes only. In view of these histological findings, a bone marrow aspiration and trephine biopsy, and skeletal survey were performed, which were normal, with no evidence of myeloma. Serum and urine electrophoresis were normal. Aspiration of the subcutaneous abdominal fat was negative for the presence of amyloid. He had an uneventful post-operative recovery and has remained well since.

Discussion

As many as 98% of patients with systemic amyloidosis also have involvement of the gastrointestinal tract. Hence, colonic amyloidosis usually occurs as part of generalized gastrointestinal involvement in the various known types of amyloidosis. Thus, in our patient it was important to look for evidence of systemic amyloidosis. Biopsy of the rectal submucosa has been traditional but also biopsy of other tissues, renal, liver, splenic and endomyocardial, has been used. Abdominal fat aspiration provides a high yield of positive results (up to 84%) with minimal risk. Scintigraphy with 125I-labelled serum amyloid P component where available may offer a non-invasive and highly sensitive technique for the evaluation of systemic amyloidosis.

The majority of extramedullary plasmacytomas are found in the oral cavity and upper respiratory tract, rather than in the intestine. Plasmacytoma is most prevalent in the 40–70 year age group with a male-to-female ratio of 3:1. The diagnosis of an extramedullary plasmacytoma should only be made after excluding multiple myeloma by the histological findings, skeletal survey, bone marrow aspiration, and serum and urine electrophoresis. Several authors point out that some of the published cases have not in fact met all of these criteria.

In retrospect, this patient's prolonged history of diarrhoea suggests that the plasmacytoma with associated amyloidosis was a long-standing problem. There would appear to have been local secretion of a potently amyloidogenic light chain as a result of the plasmacytoma. Impaired motor activity with subsequent diarrhoea is believed to be a common presentation of colonic amyloidosis. It is thought that intestinal hyperactivity is a result of mechanical compression of nerve endings. Later, signs of obstruction can occur as a consequence of neuronal atrophy.

In a similar case, an abdomino-perineal resection was performed on a patient with a solitary rectal tumour. Although there was no histological evidence of plasmacytoma, the immunochemical findings of AL-amyloid with lambda chains pointed to a plasmacytoma as the most likely underlying cause when the tumour recurred. Our case however demonstrates a more clear-cut histological association between a plasmacytoma and a large local deposition of amyloid in the large bowel.

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References


