Unusual presentation of more common disease/injury

Mesenteric amyloid deposition as the initial presentation of multiple myeloma

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Summary

Despite many recent progresses in diagnostic modalities, occasionally the initial manifestation of the diseases may be misleading. Therefore, to consider the uncommon presentations of prevalent diseases may be of help. Plasma-cell dyscrasia is one of the most well-known haematological malignancies. Clonal expansion of plasma cells results in diverse clinical findings, such as renal failure, lytic bone lesion, anaemia, hyperviscosity syndrome and so forth. However, this disease entity rarely presents with abdominal mass due to mesenteric amyloid deposition. Here we report a case of a 53-year-old Iranian woman who presented with a 4-month history of abdominal pain and fullness; she was finally found to suffer from small bowel mesenteric amyloidosis in the context of multiple myeloma.

BACKGROUND

Multiple myeloma is characterised by the classic triad consisting of tissue plasmacytosis, presence of M component in plasma or urine and evidence of organ damage. Plasma cells are derived from lymphocyte lineage. In the case of expansion of an aberrant clone of plasma cells in marrow, normally residing haematopoietic cells would be suppressed in number and function. The tumourous products, either abnormal immunoglobulins or inflammatory cytokines, may bring about such deleterious effects as renal failure, hyperviscosity syndrome and lytic bone lesions. Increased susceptibility to certain infections is another common manifestation in these patients.¹⁻⁴ Occasionally, the disease presents with an extramedullary mass consisting of abnormal plasma-cell nests.⁵ However, the disease rarely presents with an abdominal mass due to amyloid infiltration of omentum or mesentery. Here, we present a case of multiple myeloma presenting initially with a 4-month history of abdominal mass; the mass was due to mesenteric amyloid depositions.

CASE PRESENTATION

The patient was a 53-year-old, married, Iranian woman. Four months prior to her admission to our ward, she developed a sense of fatigue and weight loss. A few weeks later, she noticed a vague abdominal pain. At the same time she was surprised to find her abdominal girth had increased and there was fullness on abdominal palpation. Her physician ordered an ultrasound study of the abdomen followed by a CT scan of abdomen. There was evidence of retroperitoneal lymph node enlargement; the presence of discrete abdominal mass was suspicious (figure 1). She had no gastrointestinal symptoms and there was no evidence of intestinal wall thickening according to bowel studies. She was referred to our ward to undergo lymph node biopsy.

She also had anaemia, renal failure and nephrotic range proteinuria.

INVESTIGATIONS

Urine protein electrophoresis showed heavy light-chain protein excretion; surprisingly, serum protein electrophoresis revealed hypogamaglobulinaemia. There were also multiple lytic bone lesions in the bone survey. These features led us to perform bone marrow aspiration and biopsy. The pathological and flow cytometric studies were consistent with plasma-cell dyscrasia. She was a candidate for receiving thalidomide and dexamethasone regimen. Yet, the abdominal mass within the context of multiple myeloma was unjustified; so she underwent an open biopsy from the mass. Due to her clinical condition, the treatment was started while the result of the biopsy was pending.

There was a huge small bowel mesenteric mass with firm consistency. The mass had been encased in the superior mesenteric artery and was in a close approximation to the aorta posteriorly. The result of Congo red and immunohistochemistry staining revealed immunoglobulin light chain (κ) depositions. The abdominal mass was, surprisingly, due to an amyloid L (AL) mesenteric amyloidosis (figure 2).

DIFFERENTIAL DIAGNOSIS

Plasma-cell dyscrasia, lymphoma.

TREATMENT

She received thalidomide and dexamethasone regimen.

OUTCOME AND FOLLOW-UP

She has been undergoing treatment and close follow-up for 6 months. She feels much better now and the abdominal mass has decreased in size significantly.

DISCUSSION

Amyloidosis is caused by extracellular deposition of insoluble protein fibrils. According to the type of fibrils, the diseases are categorised to such types as AL amyloidosis,

Figure 1 (A,B) Selections of abdominal CT views illustrates amyloid deposition presenting as an intra-abdominal mass.

Figure 2 Photomicrographs of histopathological specimen obtained from mesenteric mass showing amorphous amyloid depositions, giant cell formation (A) and plasma-cell infiltration (B).

amyloid A amyloidosis, familial type and so forth. The pathogenesis of different types varies considerably. AL amyloidosis is the most prevalent one in most series. It is due to deposition of immunoglobulin light chain fragments in extracellular tissues. Any organ may be involved and its function may be deranged by these toxic fibrils; most frequently, kidneys, heart and liver may be involved.^{6–9} AL amyloidosis usually occurs in the context of a clonal B cell proliferative disease. Multiple myeloma is the most common. It is not discretely obvious what the prompting agent is: the expanded population of abnormal plasma cells produces such amounts of overwhelming light chain proteins to be deposited in other tissues or the depositions play the initial role? This means they stimulate the bone marrow plasma cells to proliferate inexorably. Up to 20% of AL patients have multiple myeloma. The myeloma may be diagnosed at any time within the AL course. However, very few cases of multiple myeloma present with mesenteric amyloid deposition.^{10 11}

AL amyloidosis may initially present locally in such organs as kidneys, heart and the gastrointestinal tract. However, as the disease progresses, it generally affects several organs in the body. The gastrointestinal tract may be frequently involved in the disease. Up to 70% of patients develop gastrointestinal symptoms, such as weight loss, bleeding, malabsorption and changes in bowel habit. Yet, it is uncommon for the disease to be discovered as peritoneal or omental mass intervening intestinal loops.^{12–14}

Solitary plasmacytoma might infrequently arise in the abdominal cavity. Intestinal obstruction due to

plasmacytoma polypoidosis and mesenteric adenopathies has also been reported. Mesenteric panniculitis and intestinal necrosis due to mesenteric vein thrombosis have been reported in the context of multiple myeloma. Aggregates of malignant plasma cells can produce various manifestations anywhere in the body, including the abdominal cavity, but the disease rarely presents with an abdominal mass due to mesenteric amyloid deposition.^{15 16}

To our knowledge, there are very few cases describing mesenteric and omental amyloid deposition as the initial presentation of plasma-cell dyscrasia and, as omental involvement is rarely considered to be a common manifestation of amyloidosis, we believe that presentation of this case may be of help for future patients to be diagnosed earlier in their disease course.

Learning points

- ▶ To consider an uncommon presentation of plasma-cell dyscrasia.
- ▶ To consider mesenteric amyloidosis as another abdominal or gastrointestinal manifestation of AL amyloidosis.
- ▶ To consider the patient's clinical findings as a whole.

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Competing interests None.

Patient consent Obtained.

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