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Case Report

Extramedullary gastric plasmacytoma

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This report describes a case of gastric plasmacytoma which was diagnosed following an episode of haematemesis and melaena. The patient had a previous orbital plasmacytoma and subsequently developed aggressive disseminated disease with extensive skin and subcutaneous deposits but without marked marrow involvement until the terminal phase. Following the episode of haematemesis and melaena, endoscopy revealed two gastric ulcers and histological examination showed extensive infiltration of the gastric mucosa by a monoclonal population of plasma cells. Gastric plasmacytoma is a rarely described cause of haematemesis and melaena and should be suspected in a patient with a history of plasmacytoma or multiple myeloma. The extensive extramedullary disease in the absence of marked marrow involvement is unusual in the spectrum of plasma cell dyscrasia.

Extramedullary plasmacytoma is relatively uncommon, accounting for approximately 4% of all plasma cell neoplasms.[1] For a diagnosis of primary extramedullary plasmacytoma a normal radiological skeletal survey is required together with an absence of bone marrow involvement.[2] Some patients with extramedullary plasmacytoma, particularly in the upper aerodigestive tract, achieve long term remission following radiotherapy while others progress to multiple myeloma.[1,3] This report describes a patient with extensive extramedullary plasma cell infiltration in the absence of marked marrow involvement which is unusual in plasma cell dyscrasia. Gastric involvement was diagnosed following an episode of haematemesis and melaena.

CASE REPORT A 53-year-old man presented with a two day history of haematemesis, melaena and epigastric discomfort. His past medical history included a right orbital plasmacytoma five years previously. At that time he had an IgA paraprotein of 57g/dl with no associated immune paresis. Bone marrow aspirate and trephine biopsy were normal as was a skeletal survey. He had been treated with five courses of combination chemotherapy. This resulted in the disappearance of his paraprotein and he was maintained on interferon until his present admission. One year previously he underwent radiotherapy treatment for a solitary lucent area in his right hip. Bone marrow aspirate and trephine and skeletal survey again showed no evidence of multiple myeloma. On examination he was mildly anaemic and tender in the epigastrium. Rectal examination confirmed the presence of melaena.

On admission there was an IgA paraprotein of 3.6gldl. Endoscopy revealed two large peptic ulcers on the greater and lesser curvatures of the stomach (figure 1). These had a characteristic bull's eye appearance. Biopsy was performed and histological examination showed extensive infiltration of the lamina propria of the stomach by plasma cells (figure 2a). Immunohistochemical staining with kappa and lamda light chains (Dako, Copenhagen, Denmark) showed light chain restriction with a preponderance of kappa light

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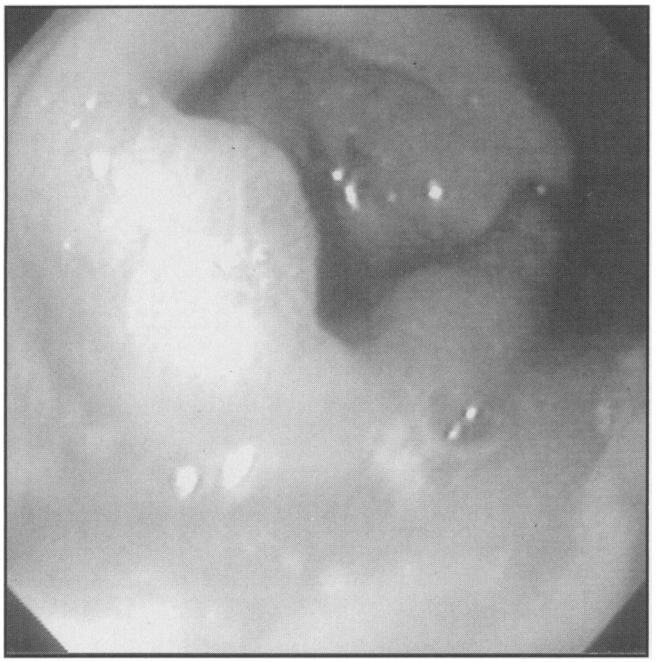

Fig 1. Gastric ulcer curve of stomach.

chains. Bone marrow aspirate was normal but the trephine revealed a small excess of plasma cells which, in the clinical context, was thought to be in keeping with multiple myeloma. He underwent further combination chemotherapy similar to the previous regime. There was no further haematemesis or melaena. Follow up endoscopy at two months showed resolution of the lesion on the lesser curve with healing of the greater curve ulcer. Eight months later he developed multiple skin and subcutaneous nodules, especially involving the head and neck. Fine needle aspiration of one of these showed numerous plasma cells (figure 2b). Bone marrow trephine biopsy showed extensive infiltration by plasma cells in keeping with multiple myeloma. His disease became resistant to further chemotherapy and he died of a chest infection.

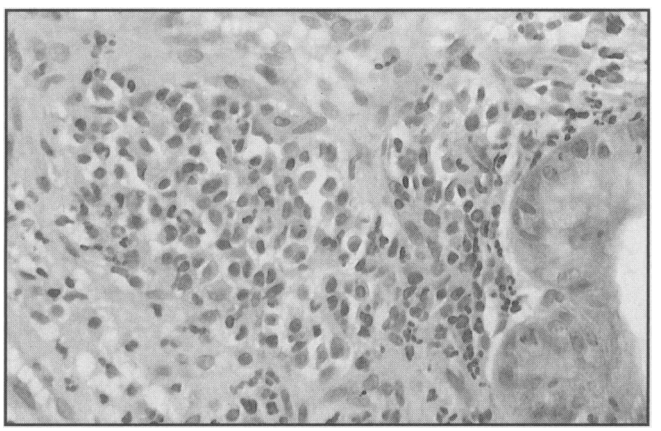

Fig 2a. There is extensive infiltration of the lamina propria of the stomach by plasma cells (haematoxylin and eosin).

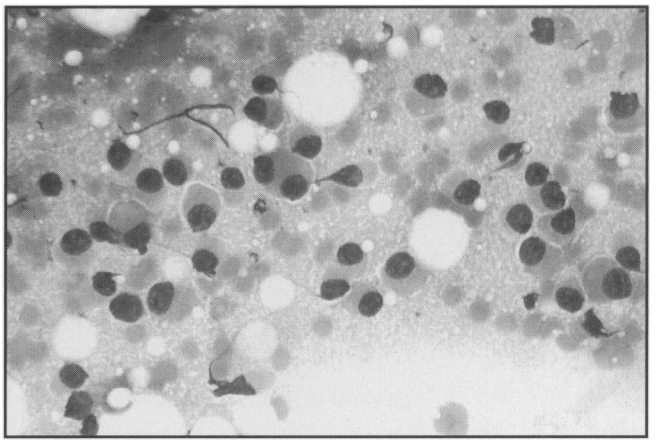

Fig 2b. Fine needle aspirate of skin nodule showing many plasma cells (Giemsa).

DISCUSSION

This report describes an unusual case of gastric involvement by extramedullary plasmacytoma. Extramedullary plasmacytomas are relatively uncommon, the most common site being the upper aerodigestive tract. A recent review of 400 publications between 1905 and 1997 revealed 869 cases of which 714 were confined to the upper aerodigestive tract with 155 cases found in other sites.[1] Only 17 of these involved the stomach and 26 the skin hence confirming the rarity of this presentation. Presenting features in cases with gastric involvement are often non-specific with epigastric pain, weight loss and anorexia. Haematemesis and melaena are unusual.[4-6] Treatment options tor extramedullary plasmacytoma include surgery, radiotherapy and chemotherapy. Surgery and radiotherapy are favoured in many cases, particularly with lesions in the upper aerodigestive tract where long-term remission is common.[1,3] In view of the previous response this patient received further combination chemotherapy. There was no further haematemesis or melaena and follow up OGD showed resolution of one ulcer with healing of the other. There was disappearance of the IgA paraprotein. Such a favourable response to chemotherapy is well documented in extramedullary plasmacytoma.[3,4]

The history of an orbital plasmacytoma and the subsequent development of multiple cutaneous and subcutaneous lesions without extensive

marrow involvement until the terminal phase is rare within the spectrum of plasma cell dyscrasia. The development of multiple skin and subcutaneous lesions coincided with the development of extensive marrow infiltration.

In summary, we describe a rare case of gastric plasmacytoma in a patient with a previous history of orbital plasmacytoma. Gastric plasmacytoma should be considered in the differential diagnosis of a patient with a history of multiple myeloma or plasmacytoma who develops haematemesis and melaena.[7] This case is also unusual in that marked extramedullary disease was present without extensive marrow involvement until the terminal phase.

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