

Granulocytic sarcoma of the rectum: a rare complication of myelodysplasia

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Abstract

A 67 year old man with myelodysplasia was admitted as an emergency with a six week history of rectal bleeding and diarrhoea. Barium enema showed an irregular polypoid filling defect in the lateral wall of the proximal rectum near the rectosigmoid junction. Histology showed this to be a granulocytic sarcoma (extramedullary granulocytic leukaemia; chloroma) infiltrating the bowel. A low index of suspicion of this lesion results in an incorrect diagnosis in many such cases. A chloroacetate esterase immunoperoxidase stain will confirm the granulocytic nature of the tumour cells.

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Keywords: granulocytic sarcoma; myelodysplasia; rectal cancer

Case report

A 67 year old man with myelodysplasia requiring blood transfusions was admitted as an emergency with a six week history of rectal bleeding and diarrhoea. Eleven months earlier a bone marrow trephine biopsy showed hypercellular marrow with marked erythroid hyperplasia with megaloblastic and dysplastic changes. Granulopoiesis was relatively decreased with no increase in blasts. No ring sideroblasts were present.

A full blood count showed a haemoglobin of 7.5 g/dl, white cell count of $1.0 \times 10^9/l$ and a platelet count of $18 \times 10^9/l$. The blood film failed to show any blasts. Sigmoidoscopy revealed an inflamed mucosa with contact bleeding, and a barium enema showed an irregular polypoid filling defect in the lateral

wall of the proximal rectum just beyond the rectosigmoid junction, which was interpreted as a primary rectal carcinoma.

On the basis of the above investigations, a Hartmann surgical resection was carried out. The postoperative period was complicated by a septicæmic shock probably secondary to severe cellulitis at the colostomy site, which progressed to a fistula. He finally succumbed to bronchopneumonia four weeks after the operation. No necropsy examination was carried out.

Pathology

The specimen of rectum showed an ulcerated tumour 2 cm in diameter, 3.5 cm from the distal resection margin. Histology showed a full thickness mucosal ulcer with regenerative glands at the sides, just deep to the base of which there was a dense infiltrate of mature plasma cells (fig 1). In the deep submucosal and serosal tissue there was an infiltrate of neoplastic haemopoietic cells containing medium sized nuclei with indistinct nucleoli. Occasional bilobed nuclei and mitoses were identified and most of the cells contained cytoplasmic eosinophilic granules (fig 2). Some lymph nodes dissected from the serosa in the region of the ulcer showed sinusoidal infiltration by immature myeloid cells.

The atypical cells did not stain with low molecular weight cytokeratin (CAM 5.2) and epithelial membrane antibody markers, but did stain with common leucocyte antigen. The chloroacetate esterase (CAE) stain showed cytoplasmic positivity. The large majority of the tumour cells were CD43 positive. These features confirmed the diagnosis of a granulocytic sarcoma (extramedullary granulocytic leukaemia; chloroma) infiltrating the bowel.

Histology also showed intimal obliteration of some medium sized arteries and veins in the serosal tissue, with focal fibrosis of the muscularis propria and haemosiderin deposition in the submucosa. This pathology implies a degree of ischaemia in the aetiology of the ulcer, possibly produced by tumour involvement of vessels.

Discussion

Granulocytic sarcoma is an extramedullary tumour composed of immature cells of the granulocytic series (myeloid cells). It was first described by Burns in 1811,¹ and because such tumours display a greenish colour owing to the presence of myeloperoxidase in the cells, the term chloroma was coined in 1853 by King.² This tumour mass may arise in four clinicopathological situations: (1) a harbinger of acute myeloid leukaemia in a non-leukaemic patient;

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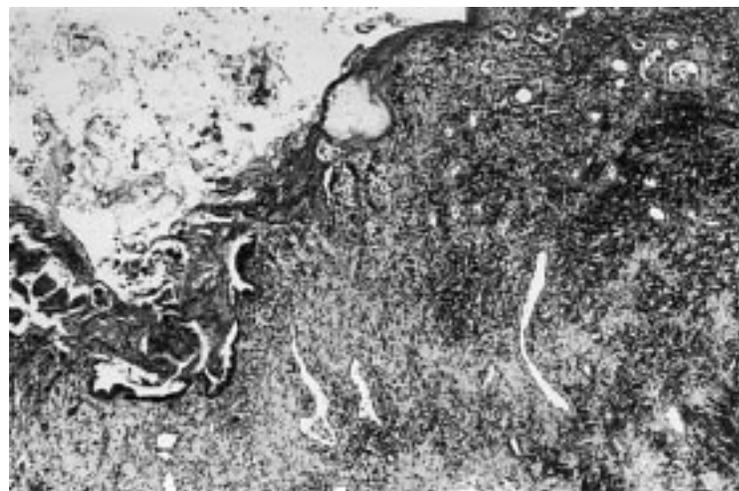


Figure 1 Full thickness mucosal ulceration in the rectum. (Haematoxylin and eosin, magnification $\times 9$.)

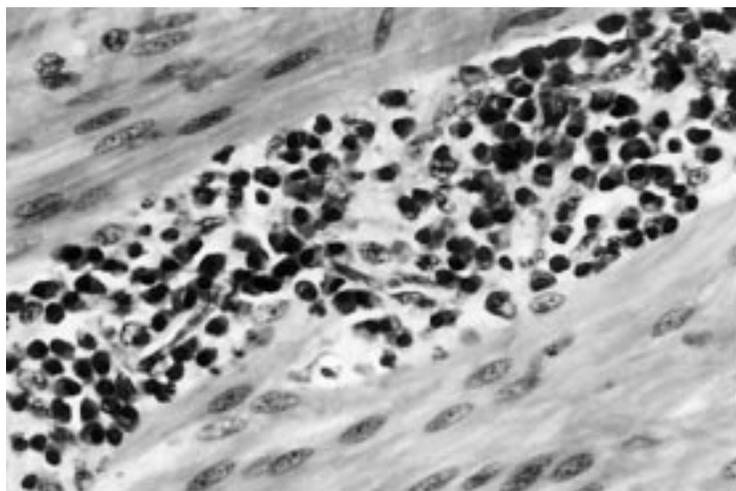


Figure 2 Dense infiltrate of monotonous medium sized nuclei with inconspicuous nucleoli. (Haematoxylin and eosin, magnification $\times 36$.)

(2) impending blast crisis in chronic myeloid leukaemia or leukaemic transformation in myelodysplastic disorders; (3) an additional complication of known acute leukaemia; and (4) an isolated lesion.³

The most common presentation is a tumour confined to a single site, although one third of patients with chronic myeloid leukaemia are found to have foci of extramedullary leukaemia at necropsy.⁴ Granulocytic sarcoma may occur in virtually any organ, although the central nervous system, bones, ovaries, and lymph nodes are most frequently involved. Tumours

of the small or large intestine have seldom been described; this involvement is usually asymptomatic, but patients may present with acute abdominal pain or change in bowel habit with rectal bleeding.^{3 5 6}

Granulocytic sarcoma may range from well differentiated tumours on histological examination to those with virtually no evidence of myeloid differentiation. A low index of suspicion of the lesion results in an incorrect diagnosis in many cases⁷; however, the chloroacetate esterase immunoperoxidase stain will confirm the granulocytic nature of the tumour cells. The lesion in the present case presented both radiologically and macroscopically as a common rectal carcinoma; consideration of the alternative correct diagnosis of granulocytic sarcoma in the appropriate clinical context is important, since chemotherapy or radiotherapy are then treatment options.³

- 1 Burns A. *Observations of surgical anatomy in head and neck*. Edinburgh: Thomas Royce, 1811:364–6.
- 2 King A. A case of chloroma. *Monthly J Med* 1853;17:97.
- 3 Catalano MF, Levin B, Hart RS, et al. Granulocytic sarcoma of the colon. *Gastroenterology* 1991;100:555–9.
- 4 Rosenthal S, Cannellos G, De Vita VT, et al. Characteristics of blast crisis in chronic granulocytic leukaemia. *Blood* 1977;49:705–19.
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- 6 Rottenbery GT, Thomas BM. Case report: granulocytic sarcoma of the small bowel—a rare presentation of leukaemia. *Clin Radiol* 1994;49:501–2.
- 7 Meiss MJ, Butler JJ, Osborne BM, et al. Granulocytic sarcoma in nonleukaemic patients. *Cancer* 1986;58:2697–709.