CLINICAL IMAGES

Massive postoperative splenulosis

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Symptomatic thrombocytopenic purpura occurred in an otherwise normal 62-year-old man. Bonemarrow studies showed features typical of peripheral sequestration and the condition was diagnosed as autoimmune. There was no response to corticosteroid administration. Laparoscopic splenectomy was uneventful and the patient remained well off all treatment. Four years later a packed cell volume, previously around 43%, started to rise beyond 50%. Investigations revealed no cause and whole-blood viscosity was controlled with occasional venesection. The patient subsequently developed nagging recurrent pain in the left iliac fossa, and the scan, normal 1 year previously, showed a large mass (Fig. 1) on the posterior gastric wall. No other pathology was noted and a clinical diagnosis of leiomyoma sarcoma was made. At operation hundreds of splenuculi were seen scattered throughout the whole abdominal cavity (Fig. 2). It transpired that the lower abdominal discomfort was due to a sizeable collection of splenic tissue in the anterior abdominal wall. With difficulty the stomach was mobilised to reveal a tumour that, on frozen section, proved to be a leiomyoma. Complete removal was possible and a distal gastrectomy was performed. Postoperative recovery was uneventful.

Commentary

Two unrelated clinically important observations are illustrated. Embryologically the spleen is derived from the foregut and retains the capacity for extramedullary blood formation and removal of effete cells from the circulation.[1] It also has immunocytes that process and present antigen for elaboration of antibodies and creation of memory T-cells that combine to mediate humoral and cellular defence.[2] The organ is anatomically restrained by a fibrous capsule and elective removal, without breaching this barrier, may nevertheless be followed by enlargement of otherwise quiescent accessory

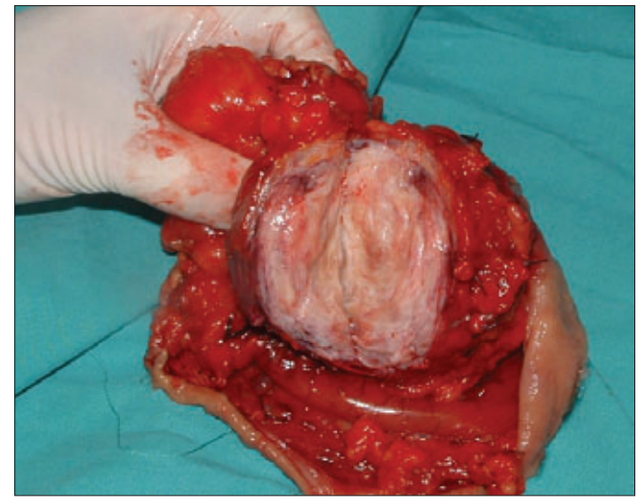

Fig. 1. The large gastrointestinal stromal cell tumour, arising from the posterior wall of the stomach, is readily evident in the surgically resected specimen.

Fig. 2. At laparotomy widespread splenuculi covering the entire peritoneal surface was a surprise finding since they were asymptomatic. The pain in the left iliac fossa was shown to represent a larger accumulation of regrown splenic tissue.

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equivalents known as a splenuculi. Rupture with liberation of the tissue contents may result in splenulosis, with function returning in children but less frequently so in adults.[3]

There is no proof that heterotopic autotransplantation provides adequate protection from infection.[4] This principle is confirmed by the extent to which splenic tissue was distributed over the surface of the peritoneal cavity, presumably at the time of laparoscopic removal of the organ. The fact that there was no commensurate fall in platelet count is unexplained. Precautions were taken against infections and these did not occur.

Gastrointestinal stromal tumours, which are the most common mesenchymal neoplasms of bowel, are of further interest. These share immunohistochemical, ultrastructural and other features with cells described by Cajal,[5] and their neoplastic transformation is ominous since chemotherapy remains ineffective. Understanding of tumour growth has improved with demonstration that in these cancers the physiological regulation of growth is via a receptor, tyrosine kinase. It is now possible to treat these patients effectively either initially or after resection with adjunctive small molecule known as imatinib mesylate, STI 571 or Gleevec.[6] There is now evidence that these neoplasms also produce immunoreactive peptides that stimulate red cell production.

Thus a specific mutation makes it possible to categorise the gastrointestinal stromal tumours precisely and to provide a highly selective small molecule that produces major response rates in a previously refractory tumour, an example of experimentally derived new knowledge in the field of cancer biology applied to clinical practice.[7]

Conclusion

The dissemination of peritoneal inoculation with cells released at the time of operation is a reminder of the potential that immunohaematopoietic progenitors have for autologous grafting. This principle underlies bone marrow or peripheral blood stem cell transplantation.

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