Acute leukaemia masquerading as acute abdomen

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INTRODUCTION

This article reports three patients with acute leukaemia who presented with acute abdomen as the first manifestation of the disease. Leukaemic infiltrates of the bowel and appendix may have caused the abdominal symptoms in these patients. Acute leukaemia should be included in the differential diagnosis of severe abdominal pain associated with an abnormal blood count. This report highlights the importance of early diagnosis of acute leukaemia as initiation of cytotoxic chemotherapy results in prompt resolution of the abdominal catastrophe and obviates the need for surgery.

DISCUSSION

Many conditions can present to the general surgeon that are primarily medical in origin. Overlooking a diagnosis of malignant haemopathy before surgery has disastrous if not fatal consequences as a result of infective and bleeding complications associated with surgery. These complications occur at a much higher rate among this group of patients, as observed in the first case. The incidence of acute surgical abdomen in leukaemic patients has been reported as between 4.6% and 5.6% (Kuffer et al, 1968; Exelby et al, 1975).

The primary differential diagnoses for acute abdomen in patients with acute leukaemia include appendicitis, typhlitis, pancreatitis, and intestinal obstruction or infection. The incidence of acute appendicitis, however, is low (0.5%) (Angel et al, 1992). Postoperative morbidity and mortality rates following appendicectomy are 25% and 8% respectively (Skibber et al, 1987).

Of patients with leukaemia, 10% are found to have typhlitis at autopsy (Angel et al, 1992). The reported incidence of typhlitis as the initial manifestation of leukaemia is extremely low (less than ten reported cases) (Kaste et al, 1997; Quigley et al, 2001).

Typhlitis (neutropenic colitis) is a necrotizing colitis that occurs subsequent to therapy- or disease-induced neutropenia (Sloas et al, 1993; Paulino et al, 1994). It usually presents as fever, diffuse abdominal pain, right lower quadrant tenderness, diarrhoea and haematochezia, and is associated with a high mortality rate (Jain et al, 2000). Paucity of gas in the right colon, a soft tissue mass with a 'target' sign in the right lower quadrant corresponding to a fluid-filled caecum, circumferential thickening of the caecal wall, and a 'thumbprinting' sign on barium enema (caused by oedema of the intestine) are some radiographic

CASE REPORT 1

A 40-year-old man was admitted to the surgical ward with a 10-day history of right lower quadrant (RLQ) pain, fever, vomiting and constipation. The pain began at the paraumbilical region and then transferred to the RLQ. On examination, the patient appeared unwell and had a temperature of 38.4°C. There was tenderness at the RLQ, but no guarding and no palpable mass. He was treated with antibiotics while being prepared for appendicectomy. The blood count report showed a haemoglobin level of 11.1 g/dl, leucocytes 345 x10^9/litre and platelets 40 x10^9/litre. A grossly inflamed retrocaecal appendix measuring 15 cm long was discovered at operation that was performed 3 hours after admission. Three days after surgery, he developed fever, diarrhoea, abdominal pain and distension, and petechial rashes over the lower limbs. His condition rapidly worsened and he required mechanical ventilation for severe bronchopneumonia on the fourth postoperative day. Subsequent investigations including microscopic examination and immunophenotyping of the peripheral blood confirmed the diagnosis of acute myeloid leukaemia. Histopathological examination of the appendix was consistent with leukaemia infiltration (*Figure 1*). Cytotoxic chemotherapy was deferred in view of worsening of septicaemia, coagulopathy and respiratory failure despite maximal supportive treatment. The patient succumbed to his illness 1 week after admission to hospital.

CASE REPORT 2

A 13-year-old boy was referred to the general surgeon with a 3-day history of severe right lower quadrant (RLQ) pain. On examination, he looked unwell, pyrexial (temperature 40°C) and dehydrated. There was marked tenderness with guarding and rebound tenderness over the RLQ region. The liver, spleen and lymph nodes were not enlarged. A clinical diagnosis of acute appendicitis was made and the patient was initially planned for surgery. However, the full blood count showed a haemoglobin level of 10.9 g/dl, leucocytes 318 x10^9/litre with 98% blasts and 1% neutrophils (absolute neutrophil count 3.18 x10^9/litre), and platelets 57x10^9/litre (*Figure 2*). Ultrasound of the abdomen showed thickening of the wall of the caecum and a normal appendix. Bone marrow examination confirmed the diagnosis of acute lymphoblastic leukaemia. Surgery was deferred and the patient received induction chemotherapy with vincristine, daunorubicin, L-asparaginase and prednisolone, and broad-spectrum antibiotics that resulted in prompt resolution of the abdominal symptoms. Following a course of chemotherapy he developed *Escherichia coli* septicaemia associated with profound neutropenia and died 14 days after admission.

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CASE REPORT 3

A 33-year-old man presented to the surgical unit with a 2-day history of central abdominal pain and fever. Abdominal examination revealed tenderness with guarding and rebound tenderness over the paraumbilical region. Initial blood tests showed a haemoglobin of 8.4 g/dl, leucocytes 45.8 x10^9/litre with 65% blasts and 16% neutrophils (absolute neutrophil count 7.32 x10^9/litre), and platelets 24 x10^9/litre. The serum amylase level was normal. A plain radiograph of the abdomen was unremarkable. The abdominal ultrasound showed a normal-looking appendix. Computed tomography of the abdomen revealed marked thickening of the wall of the duodenum and jejunum. Barium enema showed a coin-stacking appearance of the duodenum and jejunum. The patient was treated with broad-spectrum antibiotics with no improvement in the abdominal pain. An urgent bone marrow examination confirmed the diagnosis of acute myeloid leukaemia French-American-British (FAB) subtype M2. The patient received combination chemotherapy consisting of daunorubicin and cytarabine that resulted in rapid resolution of the abdominal symptoms. He successfully underwent allogeneic peripheral blood stem cell transplantation in July 2001 and has remained well ever since.

signs that suggest the diagnosis of typhlitis (Horton et al, 2000). Even though thickening of the caecal wall was noted in case two, this condition is less likely in both patients in the absence of neutropenia and previous chemotherapy. Most patients with typhlitis can be treated conservatively with intravenous fluids and antibiotics, although surgery may be necessary if medical treatment fails and a mass develops (Urbach and Rotstein, 1999).

Imaging studies including ultrasound, barium enema and computed tomography (CT) could accurately diagnose the aetiology of the acute abdomen. Frick et al (1984) stated that differentiation of typhlitis from appendicitis, intramural haemorrhage and leukaemic or lymphomatous deposits should be possible in most cases using CT. At CT, a dilated, thickened appendix suggests appendicitis (Horton et al, 2000). The results of the radiographic tests in cases two and three excluded lesions in the appendix, pancreas, gall bladder and kidneys.

Figure 1. Histopathological examination of the appendix showing acute leukaemic infiltration (haematoxylin and eosin stain x400 at image size of 11cm).

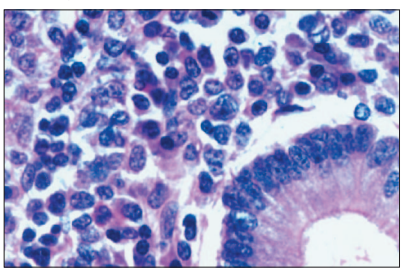
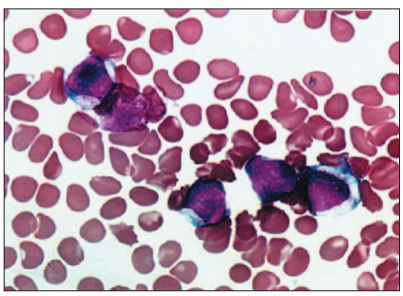


Figure 2. Microscopic examination of the peripheral blood smear showing acute lymphoblastic leukaemia (Romanowsky stain x 400 at image size of 11cm).



Systemic fungal infection, in particular invasive candidiasis, can cause severe abdominal pain but this condition is less likely in these patients in the absence of risk factors, including prolonged periods of neutropenia, the use of immunosuppressive or broad-spectrum antibiotic therapy and invasive instrumentation. The radiographic findings in these patients were consistent with infective or infiltrative lesions in the duodenum in case two and the caecum in case three.

Leukaemic infiltration of the gastrointestinal wall is uncommon (Muller et al, 1997), but should be considered in patients with abdominal pain and an abnormal blood count when other causes cannot be found. The first patient was initially treated for acute appendicitis with antibiotics and appendicectomy; he developed septicaemia and respiratory failure following surgery resulting in his death 1 week after hospital admission. Acute myeloid leukaemia was only diagnosed 5 days after surgery, by which time the patient was too ill to receive cytotoxic chemotherapy. In cases two and three, the high leucocyte count at presentation, failure to isolate organisms from the blood and stool, and the resolution of abdominal symptoms only after initiation of chemotherapy favoured the diagnosis of leukaemic infiltration over that of an infection.

CONCLUSIONS

Despite the fact that leukaemia rarely presents for the first time as acute abdomen, these cases illustrate that it must be included in the differential diagnoses of an acute abdomen and that surgeons and physicians have to be aware of this condition. CT and/or ultrasound should be performed in all leukaemic or neutropenic patients with severe abdominal pain for accurate diagnosis and prompt treatment of the underlying abdominal conditions. In addition, microscopic examination of the peripheral blood smear should be done in all patients presenting with an abnormal blood count. HM

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