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CASE REPORT

Two sides of one coin: pan-gastrointestinal Kaposi sarcoma presenting with small bowel intussusception and severe gastrointestinal haemorrhage

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Summary
We report a case of multi-focal pan-gastrointestinal Kaposi sarcoma in an HIV positive patient, presenting with two life-threatening complications of the disease, intestinal obstruction and upper gastrointestinal haemorrhage. The patient responded well to combined surgical and oncological treatment. The reported complications of the disease are regarded as relatively rare events, and this report aims to highlight the need for more intensive screening for Kaposi sarcoma in immune-compromised individuals.

Case presentation

A 39-year old, HIV positive African male presented with acute onset of abdominal pain and vomiting. He had completed treatment for abdominal tuberculosis six months earlier and had known pica disorder. At the time of presentation, HIV viral load was low (< 40cps/ml) with highly active anti-retroviral therapy (HAART). An initial abdominal radiograph was unremarkable, but due to ongoing symptoms, a computed tomography (CT) scan was arranged 48 hours later that showed proximal small bowel obstruction due to apparent intussusception (Figure 1a). Decision for surgery was made and, at laparotomy, two intussuscepting segments of proximal small intestine with large mucosal polyps as lead points were confirmed (Figures 1b and 1c). Segmental resections of the polyps along with further polypectomies of several palpable, smaller and non-obstructive polyps via enterotomies were carried out. Postoperative recovery was uneventful and the patient was discharged on postoperative day 7.

The patient was admitted again two weeks after initial discharge with acute upper gastrointestinal bleeding (haematemesis and melaena, haemoglobin 4.0 g/dl, haematocrit 0.14). Following haemodynamic stabilisation and blood transfusion, bidirectional endoscopy demonstrated multiple oesophageal (Figure 2a), gastric (Figure 2b) and proximal colonic (Figure 2c) polypoid lesions, which were biopsied. Histology of the obstructing and non-obstructing small bowel polyps, and the gastro-oesophageal and colonic biopsied polypoid lesions all revealed intestinal-type Kaposi sarcomata, characterised by strongly positive human gamma herpesvirus (HHV-8) stains. A discussion was held with the patient and he was referred for urgent oncological

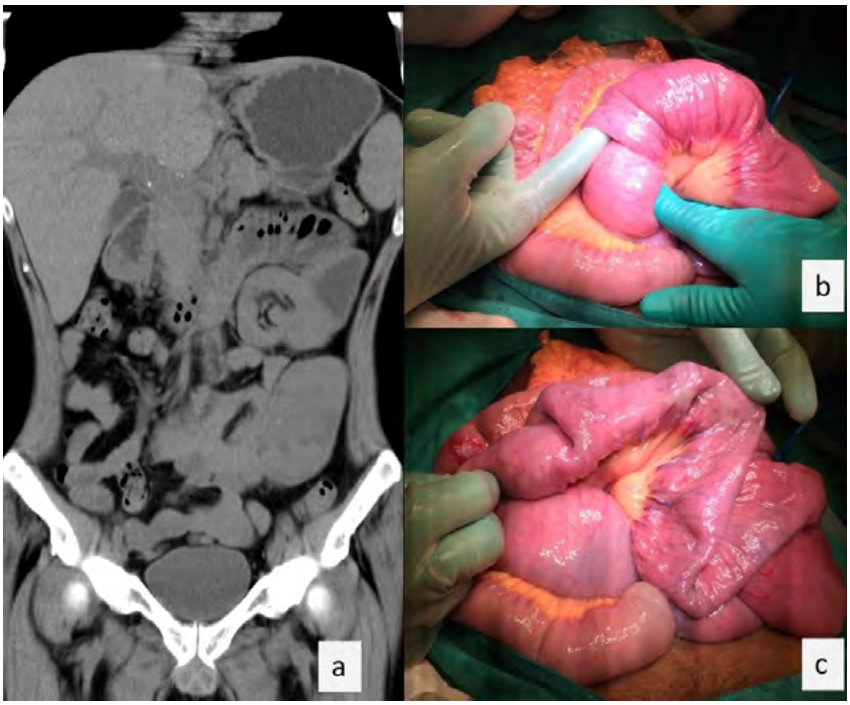

Figure 1: CT scan demonstrating proximal small intestinal intussusception (a); Operative findings of intussuscepting polypoid tumours causing small bowel obstruction (b & c)

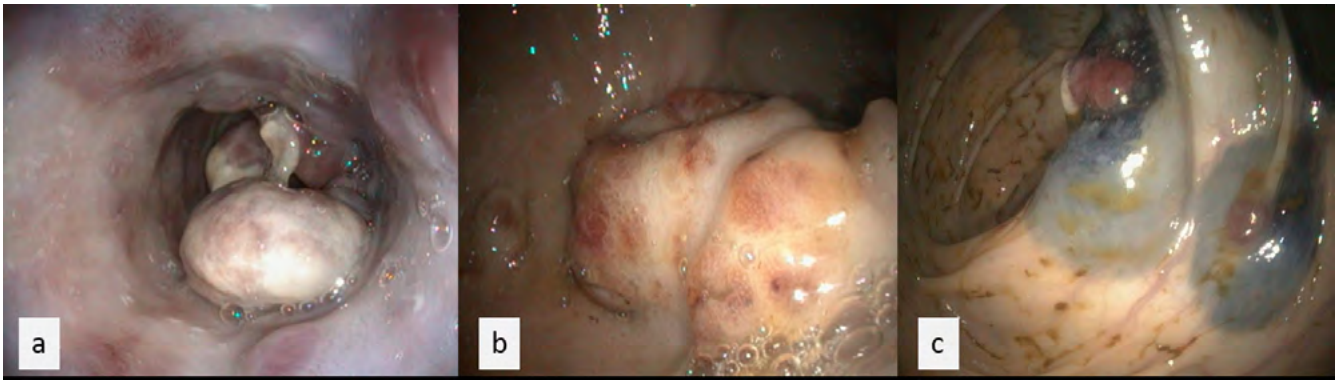

Figure 2: Oesophageal (a), gastric (b) and colonic (c) Kaposi sarcomata – colonic lesions tattooed for site recognition

intervention. He received chemotherapy with curative intent consisting of six cycles of adriamycin, bleomycin and oncovin. He remains well two years later, with ongoing anti-retroviral treatment (HAART) and no sign of recurrence.

Discussion

Kaposi sarcoma (KS) is a low-grade vascular tumour associated with HHV-8 infection,[1] classically seen in the elderly of Mediterranean and Eastern European heritage as slow-growing tumour of the limbs ("classic KS"), and in the African population in a more aggressive form ("endemic KS"). It is now more often associated with attenuated immune status, including HIV ("epidemic KS"), and immunosuppression ("iatrogenic KS"). The incidence of KS in HIV positive patients has significantly reduced with the onset and widespread use of HAART since the 1990s.[2] Following cutaneous sites, the gastrointestinal tract is the most common extracutaneous site, but often with little or no symptomatology.[3,4] The macroscopic findings on endoscopy range from red maculopapular lesions to nodular or polypoid lesions, which may display central ulcerations, as in this case.[5] Complications of gastrointestinal KS include intestinal haemorrhage, hollow viscus perforation and obstruction, although such events are rare.[6–8] Whilst HAART alone may suffice in localised disease, multi-focal and visceral disease require combination management of HAART, surgical intervention (particularly for complications), and chemotherapy and/or biologics. Our patient did have an excellent clinical response to a classic triple chemotherapy regimen, although liposomal doxorubicin alone is favoured due to both improved efficacy and tolerability,[9] however, despite low local relapse rates a high mortality rate due to other causes is evident.[10]

In conclusion, the here reported patient had a combination of two rare complications from gastrointestinal KS and responded well to systemic chemotherapy and HAART. For patients with known HIV-positive status, despite the significantly lower incidence of KS in the HIV positive population in recent years due to HAART, a high index of suspicion for gastrointestinal disease and its complications must be applied to enable early diagnosis and commencement of multi-modal intervention. Equally, in rare cases of HIV negative patients with intestinal Kaposi sarcoma, iatrogenic and lymphoproliferative disorders must be considered.

Conflict of interest

The authors declare no conflict of interest.

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