CASE REPORT

Severe gastrointestinal haemorrhage due to primary intestinal kaposi's sarcoma – a case report

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Abstract

Kaposi's Sarcoma (KS) was previously a relatively rare disease. With the advent of HIV/AIDS pandemic however, AIDS-related KS has been on the increase and so has interest in the disease. Ninety percent of patients with KS present with skin lesions. While the gastrointestinal tract is a fairly common site of metastatic KS, primary gastrointestinal KS is uncommon. The presentation of gastrointestinal KS with severe gastrointestinal bleeding is rarer still. In this report, we present a 56-year-old HIV-negative patient who presented with severe gastrointestinal bleeding without any skin lesions. Multiple hemorrhagic polypoidal lesions were found on the walls of the jejunum and ileum as well as the liver at exploratory laparotomy and these were found to be KS on histopathologic examination. We also discuss the diagnostic and therapeutic challenges we had with this rare cause of severe GI bleeding.

Keywords Kaposi's Sarcoma · AIDS · Lesions

Introduction

Kaposi's Sarcoma, (KS), was first described in 1872 by Moritz Kaposi as idiopathic multiple pigmented sarcomas of the skin [1]. It was previously a rare disease, comprising 0.1% of all malignancies worldwide. KS occurs within two clinical contexts, the classic or endemic type found primarily in elderly Mediterranean, Jewish and equatorial African men, as well as the epidemic type found in immunosuppressed patients [2].

With the advent of the AIDS epidemic in the 1980s however, AIDS-related KS has been on the increase, and a third of AIDS patients may present with KS. Skin lesions are seen in up to 90% of all KS cases [3]. The epidemic KS has propensity for metastasis, which most frequently involves the lymph nodes, gastrointestinal tract and the lungs [3]. The most common location for KS visceral metastasis is the gastrointestinal tract. The small intestine is most frequently affected, followed by the stomach, esophagus and colon. Primary Kaposi's sarcoma of the gastrointestinal tract is very rare [4, 5].

Gastrointestinal KS is usually clinically silent and when apparent, it commonly present as anaemia from chronic blood loss, intestinal obstruction, perforation, intussusception, and protein losing enteropathy [4, 5, 6] rather than massive gastrointestinal bleeding which is rare [4, 5], and despite the prevalence of HIV/AIDS in Africa, there are no reports of severe gastrointestinal haemorrhage from KS.

We present here an HIV-negative patient who was admitted with severe GI bleeding from jejunal and ileal Kaposi's sarcoma, to highlight the need to consider KS in the differential diagnosis of acute severe GI bleeding.

Case report

A 56-year-old administrator presented to us with recurrent passage of melaena stool of six months duration. He

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had chronic duodenitis from *Helicobacter pylori* infection diagnosed five years earlier by gastroduodenoscopy and gastric biopsy. He received full treatment for *Helicobacter pylori*. Post treatment gastroduodenoscopy showed healed anterior duodenal ulcer. He was not on maintenance anti-ulcer therapy. He was however well until he re-presented 5 years after discharge with recurrent passage of large amount of bloody stool.

He was very pale, with thready low volume pulse on admission. The blood pressure was 80/50mmHg. He was not jaundiced. Liver span was 10 cm and there was no demonstrable ascites. The initial working diagnosis was recurrent bleeding duodenal ulcer. He was negative for HIV I & II antibodies.

He was transfused with five units of whole blood. Gastroduodenoscopy and proctosigmoidoscopy revealed no ulcers or any other abnormality. Liver function tests, clotting profile and blood urea and electrolytes were within normal limits. Repeat HIV screening test was negative.

With continuous passage of large amounts of bloody stools, he was immediately worked up for exploratory laparotomy, which revealed widespread haemorrhagic, polypoidal lesions on the wall of the small intestine extending from the proximal jejunum to the terminal ileum. These lesions varied from 1 to 3 cm in diameter with a large concentration in the mid-portion of the ileum. A 3 x 2cm highly vascularised lesion with poorly defined edges was seen on the diaphragmatic surface of the left lobe of the liver. Multiple enlarged mesenteric lymph nodes were seen. There was no ascites. A 45 cm portion of the ileum displaying the largest concentration of the masses was resected with end-to-end anastomosis. A biopsy of liver lesion was also taken. Histopathologic examination of the resected specimen revealed multiple nodular masses in the small intestinal mucosae some of which are umbilicated. On microscopy, the submucosal, muscular and serosal layers of the ileum and jejunum showed numerous malignant slit-like vascular channels, many of which contained no red blood cells. There were numerous haemosiderin-laden macrophages. Resection margins were not free from the lesion. The liver lesions and the lymph nodes were similar to the intestinal ones. A pathological diagnosis of small intestinal Kaposi's sarcoma with hepatic and lymphatic secondaries was made.

The patient had two units of blood postoperatively and the recovery was smooth though patient had superficial wound infection. He had an episode of melaena three weeks after the surgery, which stopped after two days. He was placed on haematinics and was discharged. He was seen twice in the outpatient clinic with fairly stable general condition and was to commence chemotherapy on the next visit but suddenly developed severe abdominal pain and distension at home and was brought to the hospital four hours later in hypovolaemic shock. He died in the process of resuscitation. Postmortem examination revealed haemoperitoneum from rupture of the hepatic lesion.

Discussion

AIDS related form of KS is on the increase in Africa and other parts of the world though the classic form of the disease in HIV negative individuals still exists. Regardless of its clinical context, cutaneous kaposi is the most common form of presentation of the disease. Up to one third of patients with the disease have gastrointestinal secondaries, [6] but primary gastrointestinal KS is very rare. When present, the lesions of gastrointestinal KS are usually multiple, involving both the upper and lower parts of the tract as well as solid organs. Gastrointestinal KS, either primary or metastatic, is usually indolent [4, 5, 7] and when it occurs, may present as intestinal perforation, intestinal obstruction, multiple intra-abdominal masses or occult blood loss and chronic anaemia. Severe GI bleeding necessitating repeated blood transfusion is rare and there are no documented reports of this eventuation in our sub region and scanty reports worldwide [4, 5, 8]. Earlier cases reported include GI secondaries in a patient with stable cutaneous disease [5], HIV-infected individual [8] and a patient on immunosuppressive therapy following cadaveric renal transplantation [4]. The patient we report here is a heterosexual, HIV-negative man who is not on any immunosuppressive therapy and has no cutaneous lesions. This picture informed our judgement that this lesion is probably a primary gastrointestinal KS with hepatic and lymphatic metastasis. The possible antecedents of this primary gastrointestinal KS as well as the rare presentation of severe bleeding in this patient is unclear.

This case was a diagnostic challenge initially since the patient was previously treated for chronic duodenal ulcer due to *H.pylori* infection five years prior to this presentation. Recurrence after *H. pylori* eradication occurs in a third of patients [1] hence at representation, our preoperative diagnosis was severe bleeding from a recurrent duodenal ulcer. There were therapeutic challenges in this patient too. The persistent life-threatening blood loss necessitated exploratory laparotomy. In many tertiary health centers in the developing world, facility for selective angiography and or contrast enhanced CT scanning with possible embolization prior to operative intervention is not readily available and not infrequently is such diagnosis made only at exploratory laparotomy as it was in this patient. The site of the lesions also made it inaccessible to visualization at oesophago gastroduodenoscopy and proctosigmoidoscopy.

In our opinion, this case should serve as a reminder that visceral KS may occur in the absence of HIV infection, even in Africans, and that it should be borne in mind in the differential diagnosis of severe gastrointestinal bleeding.

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