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Isolated intestinal Kaposi's sarcoma in a kidney transplant patient: Diagnostic difficulty

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ABSTRACT

A 33 year old man with unknown cause of end stage renal disease received a renal graft from a deceased donor. Triple immunosuppression with cyclosporine A (CyA), mycophenolate mofetil and prednisone was prescribed. Four months after transplantation the patient developed general weakness, fever, diarrhea and bleeding to the intestinal tract leading to severe anemia. Due to the fact that no bleeding site was found during gastroduodenoscopy and colonoscopy, open surgery was performed and revealed bleeding ulcerations in small intestine. Partial resection of small intestine and hemicolectomy was performed and histopathological examination revealed Kaposi's sarcoma (KS) infiltration in the ulceration. CyA was withdrawn and rapamycine started. After 4 years post surgery the patient has been doing well, without symptoms of KS.

Implication for health policy/practice/research/medical education:
This case suggests that although GI bleeding is extremely rare presentation of KS, it should be considered in the differential diagnosis of tumors of this region among renal transplant recipients.

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1. Background

Kaposi's sarcoma (KS) is a malignant tumour, characterized with a vascular proliferation as a most prominent feature. Seroepidemiologic studies have made a strong association between HHV-8 infection and KS (1). Clinical manifestation include: cutaneous nodules and blue or purplish colour plaques, lymph node enlargement, with signs and symptoms of visceral involvement. Skin lesions are observed in more than 90% of cases while visceral localization is less frequent (2). The prevalence of KS in the gastrointestinal (GI) tract is observed in AIDS patients, but is relatively rare in solid organ recipients. Primarily tumor growth is in the submucosa and the disease is usually asymptomatic, diagnosed at endoscopy as small multiple lesions (3). We report a case in which severe, life threatening bleeding from the intestinal tract was caused by KS infiltration in the small intestine undetectable during routine endoscopic procedures.

2. Case presentation

A 33 year old man with unknown cause of end-stage renal disease, with a three years history of hypertension,

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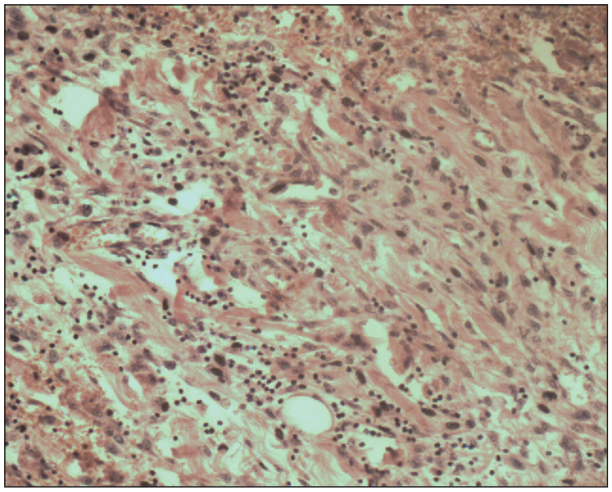

Figure 1. KS infiltration. On low power the picture is similar to granulation tissue containing sparse, mixed-cellular inflammatory infiltrate (H & E, x200).

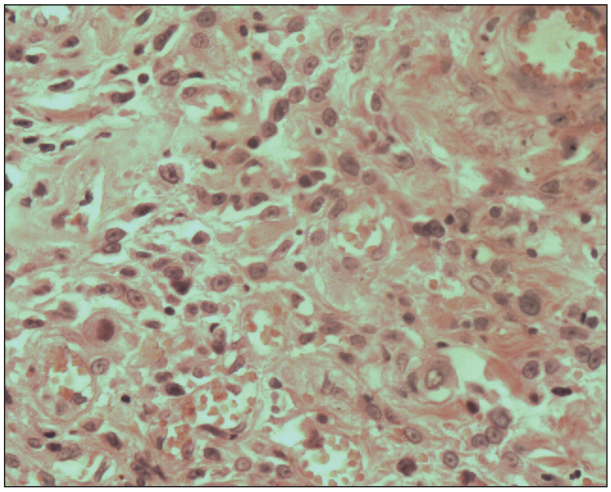

Figure 2. KS infiltration. On higher magnification plump elongated cells are visible. There are many vascular spaces filled with erythrocytes (H & E, x 400).

hemodialyzed since April 2004, received a renal graft from a deceased donor in September 2006. Cyclosporine A (CyA), mycophenolate mofetil (MMF 2 g/day) and prednisolone (P) were given for immunosuppression. Delayed graft function was observed after transplantation. At the time of discharge, serum creatinine was 2.16 mg/dl. There were no rejection episodes. Post-transplant diabetes mellitus was diagnosed and insulinotherapy started. On January 2007, the patient developed general weakness, diarrhea, melena and fever. During hospitalization in a regional hospital due to GI bleeding suspicion, laboratory results revealed severe anemia with hemoglobin (Hb) - 4.0 g/dl, thrombocytopenia (73,000/μl), leucopenia (2,000/μl) and progression of renal failure (serum creatinine 409 μmol/l). The gastroduodenoscopy showed signs of an erosive gastritis. Blood clots with no bleeding site were found during colonoscopy. After blood transfusions, Hb level increased to 9.8 g/dl. Due to leucopenia and thrombocytopenia, MMF was stopped, and CyA + P continued.

Patient was transferred to the Department of Nephrology because of fever and unexplained graft failure (serum creatinine 325 μmol/l at admission). There was no evidence of skin lesions, abnormalities on chest X-ray (CXR) and changes of abdominal organs on ultrasonography (US). Wide spectrum antibiotics and fluconazole were ordered because of fever (all cultures were negative). Due to Cytomegalovirus (CMV) infection suspicion (later confirmed by CMV PCR), endobulin and gancyclovir intravenously (IV) and later on valgancyclovir orally were prescribed. Because of sudden, massive GI bleeding (loss of 1100 ml of blood, Hb - 6.8 g/dl) the patient was transferred to the Surgical Department. Due to negative visceral angiography and no active bleeding site during gastroduodenoscopy and colonoscopy, open surgery was performed on 19th January 2007 and revealed bleeding ulceration in the small intestine located 40 cm from the Bauchin's valve and superficial ulcerations in distal part of the intestine. Partial resection of the small intestine and hemicolectomy was performed. Histopathology revealed KS infiltration of the ulceration *(Figure 1, Figure 2)*. After surgery the patient returned to the Nephrological Department where CyA was withdrawn and rapamycine started (3-4 mg daily, blood levels - 7.0 ng/ml). On February 2007, kidney function was stable (creatinine -194 μmol/l, creatinine clearance 33.1 ml/min). On March 2007, fever and exacerbation of graft failure with temporary 2 hemodialyses sessions were observed due to symptomatic CMV infection. It was treated successfully (no fever and CMV PCR negative after therapy) with cytotect i.v. every 4 weeks (5 courses). From April 2007 to January 2011 the patient has been doing well with creatinine ranging between 180-210 μmol/l. The abdominal US, CXR, colonoscopy and barium swallow of the small bowel performed on December 2009 were negative.

3. Discussion

KS is an extremely rare neoplasm in the general population, whereas the risk of its development is substantially increased in immunocompromised patients including AIDS and solid organ recipients (3). Patients receiving more intense immunosuppressive therapy are at a significantly high risk of developing post transplantation KS (4). The reported incidence ranges from 0.5% in Western European countries and USA, up to 5.3% in Saudi Arabia (5). Even lower incidence (0.45%) was observed by Abbeszadeh and Taheri in the population of 2211 living kidney allograft recipients (6). The similar low results were observed by Einollahi *et al.* (7). They observed KS development in 55 patients (0.65%) of 7,939 kidney recipients from living donors.

The presented case is worthy to describe due to unusual isolated intestinal KS localization, severe life threatening gastrointestinal bleeding and diagnostic difficulties. There are many reports of atypical KS localization in the

central nervous system, musculoskeletal system, heart, bone-marrow, liver, lungs, and urinary bladder (8-12). Localization of KS in the GI tract is rare in solid organ recipients (11, 13). Reduction or withdrawal of immunosuppression caused complete remission of KS in most patients (9). The conversion of calcineurin inhibitors such as CyA or tacrolimus to rapamycine (possessing antiproliferative properties) is the most effective treatment (13-15). Additional strategies like local radiotherapy, chemotherapy, cryotherapy or systemic chemotherapy as well as surgery may be useful especially in AIDS related KS (8).

Stallone *et al.* reported 15 kidney transplant recipients with KS who underwent a switch from CyA to rapamycine. In these patients, remission was histologically confirmed (16). Harfi *et al.* (13) described successful treatment of a post renal transplant patient with gastric and pulmonary KS after conversion of tacrolimus to rapamycine and initial 50% reduction of MMF, followed by complete cassation, combined with two doses of monoclonal anti-CD20 antibodies. Yaich *et al.* (17) reported simultaneous KS and Hodgkin's disease nine months after renal transplantation, treated with good results after MMF discontinuation and switch from CyA to rapamycine and additional poly-chemotherapy associated with 4 courses of rituximab. Twelve months post treatment; the patient had normal graft function and complete remission of both malignancies. In the presented case, lifesaving was the surgical procedure eliminating bleeding ulceration but one must also note the importance of the switch from CyA to rapamycine, according to data from the literature, which may protect patient from KS recurrence (14, 16, 18).

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Conflict of interest

None declared.

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