

CLINICAL CASE

## RARE PRIMARY SMALL BOWEL TUMOR IN THE FORM OF CLASSIC KAPOSI'S SARCOMA: CASE PRESENTATION

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### Abstract

*The small bowel represents a rare site for primary neoplasms, with an incidence of less than 2% of all that are located in the digestive tract. The early diagnosis of these tumors is often difficult due to the lack of specificity of the clinical presentation, but it can make a difference in matters of 5-year survival rates: 83% (early detection) versus 14% (late detection). We present the case of a male patient, aged 66, with an apparently benign jejunal ulcer diagnosed enteroscopically. Clinical presentation: diffuse abdominal pain, fatigability, melena; paraclinical evaluation: anaemia, positive occult gastrointestinal bleeding test, superior and inferior endoscopy without haemorrhagic lesions. The surgical exploration guided by intramucosal dye discovers a 2 cm tumor, 250 cm distal to the angle of Treitz with multiple mesenteric adenopathies. A segmental resection is performed keeping oncological safety margins with the removal of the lymphatic drainage elements. The post-operative evolution is favourable and leads to complete healing. The histopathological assay finds pathological changes consistent for Kaposi's sarcoma and confirms the presence of HHV-8 in the nuclei of the tumor cells by means of immunohistochemistry. At the examination of the skin, multiple purple patches were discovered. The particularity of this case is that the Kaposi's sarcoma manifests itself in the absence of an immunosuppressive treatment or an immunodeficiency state induced by a HIV infection. Although rare, the small intestine tumors should be considered when non-specific abdominal pain is accompanied by positive occult bleeding test, with scarce endoscopic evidence. Since the Kaposi's sarcoma is a highly angiogenic tumor, it is prudent to include it in the differential diagnosis when suspecting a source of gastrointestinal bleeding.*

**Keywords:** primary intestinal tumor, intestinal Kaposi's Sarcoma, enterectomy, occult gastrointestinal bleeding

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### Introduction

Although the small intestine amounts to over 75% of the length of the gastrointestinal (GI) tract and 90% of its surface, less than 2% of the primary GI tumors originate from it, with the following histological types: 37% adenocarcinomas, 37% neuroendocrine tumors,

17% lymphomas, 8% stromal tumors and about 1% others (including Kaposi's sarcoma) [1,2].

The first description of Kaposi's sarcoma (KS) as 'idiopathic multiple pigmentary sarcomas of the skin' was as early as 1872 by Moritz Kaposi [3].

Today, we recognise KS as being induced by the Human Herpes Virus 8 (HHV8), known also

as Kaposi Sarcoma Human Virus (KSHV), a gamma herpesvirus that produces highly angiogenic masses which can affect the skin, the lymphatic ganglia and virtually any organ. The cutaneous pathology, with onset usually before any visceral involvement, varies from patches and plaque-like lesions to nodular red-purple dome-shaped masses. The internal organs most affected by this condition are the lungs and the GI tract; the liver, spleen and bone marrow were reported, as well [4,5].

Our case report illustrates a rare tumor arising in the gastrointestinal tract, since the incidence of a primary small intestinal tumor (PIT) is 2% reported to all GI tumors and a KS is under 1% of all PIT; therefore, in theory, a small bowel KS is just 0.02% of all GI tumors.

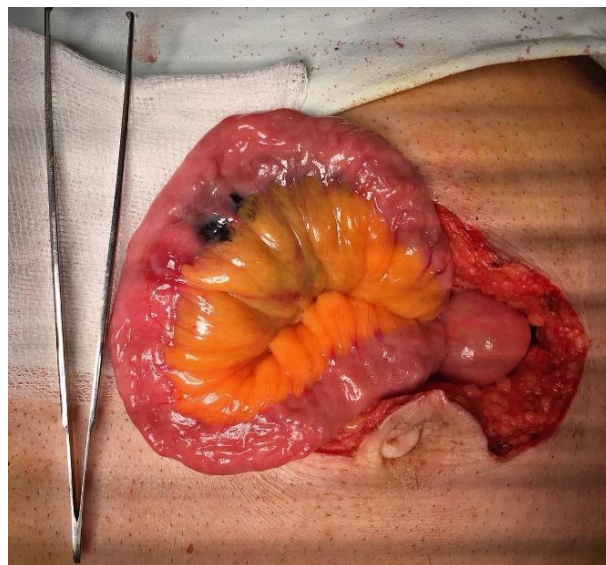
### Case presentation

A 66-year-old male patient suffering from melena, fatigability and diffuse abdominal pain was admitted to our clinic for curative surgical treatment after previously being diagnosed enteroscopically with a jejunal ulcerative lesion. The ulcer had been marked with intramucosal ink, following an initial enteroscopic biopsy that revealed inflammatory changes throughout the whole depth of the mucosa with extension in the submucosa alongside a limited lympho-granulocytic inflammatory process of the chorion, in the absence of specific changes.

Past medical history was not advantageous to case solving: NYHA class III congestive heart failure, previous coronary stenting, stroke hemiparetic sequelae and prostate adenoma. The patient denied working in toxic environment, alcohol abuse and smoking.

Since occult haemorrhage tests were positive and the patient had an increased risk of a gastrointestinal bleeding, surgical removal of the lesion was decided. After celiotomy, under intramucosal dye marking, the lesion was found at 250 cm distal to the angle of Treitz (Figure 1). The mass measured 2 cm in diameter and was accompanied by multiple mesenteric adenopathies (the largest 5 cm) (Figure 2; arrow) in the corresponding area of lymphatic drainage. Segmental bowel resection with oncological safety margins and removal of the involved mesenteric lymphatic ganglia was

performed. Following termino-terminal entero-enteral anastomosis, the mesenteric defect was closed. The patient's recovery was uneventful.



**Figure 1 – The lesion revealed by intramucosal ink marking**



**Figure 2 – Arrow points at enlarged mesenteric adenopathy (approximately 5 cm in diameter)**





**Figure 3 – Intestinal lesion close-up**



**Figure 4 – Intraluminal aspect of the ulcer-like intestinal lesion featuring indurated margins (arrow) and firm consistency**

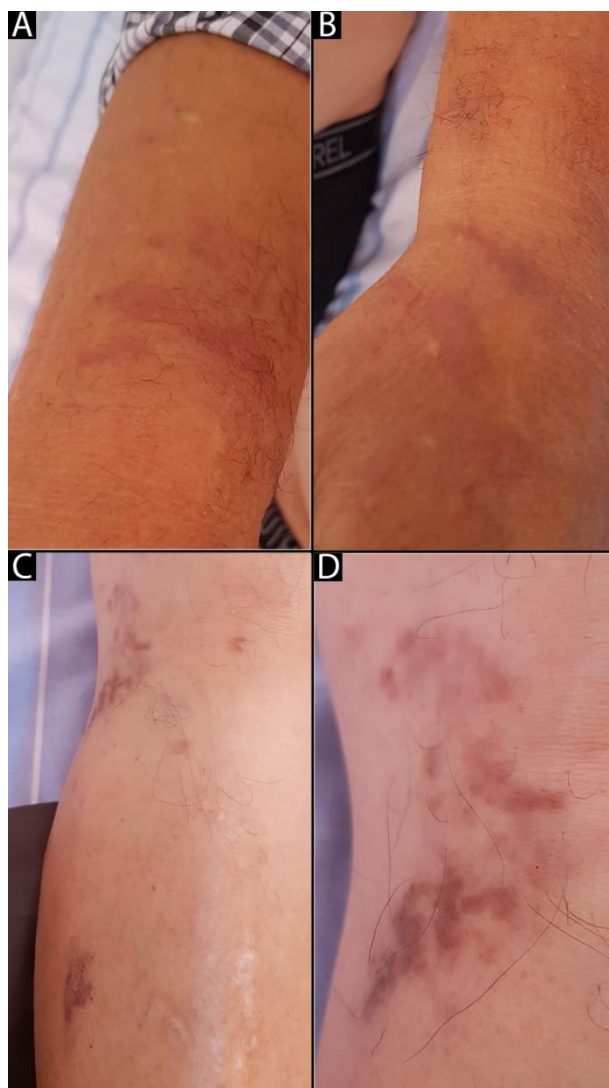
Macroscopic inspection of resected specimen resembled the ulcer-like mass previously described by enteroscopy. Additionally, it had a

firm consistency and indurated margins, besides intraluminal haemorrhagic aspect. (Figure 3), (Figure 4)

The resection piece was sent to the pathology department and was identified as Kaposi's Sarcoma.

At the time of the surgical intervention, the patient was immunocompetent. His HIV status was unknown.

The examination of the skin revealed multiple purple-brown patches at the level of the right and left upper limbs and right lower limb (Figure 5 A-D). The patient admitted to having noticed the patches for years. The lesions are consistent with a cutaneous form of KS.



**Figure 5 - Purple patches at the level of:**

- A- Right upper limb**
- B- Left upper limb**
- C- Right lower limb**
- D- Detail, right lower limb**



At the 6-month follow-up, his HIV test was negative.

### **Histopathological assay**

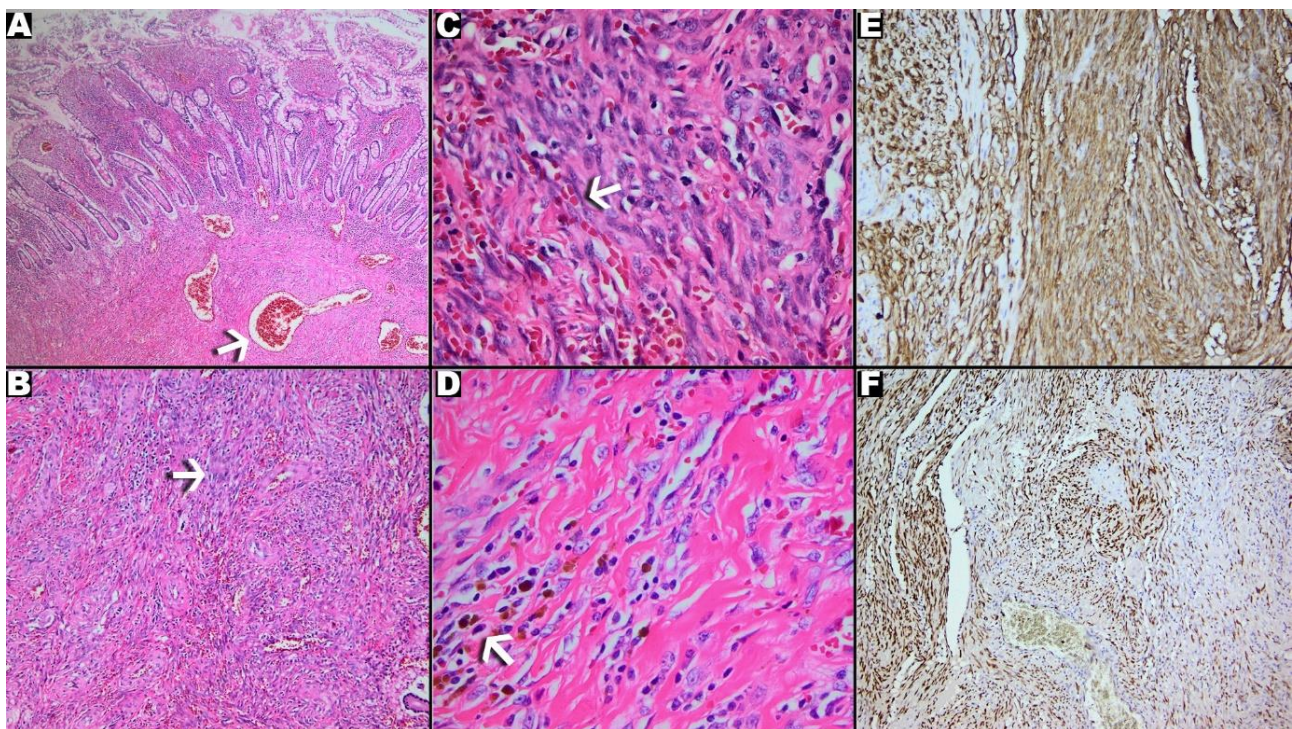
The resected specimen was found to have malignant mesenchymal proliferation with fascicular arrangement of spindle cells throughout the whole muscular layer (Figure 6 A and B). The nuclei were enlarged, polymorphic and displayed granular chromatin and rare asymmetric mitoses. Dilated vascular structures were observed in the tumoral mass and vascular intracytoplasmic clefts were present, specific to Kaposi's sarcoma. There were numerous interstitial deposits of hemosiderin (Figure 6 D). Tumor-free margins were attained.

Immunohistochemistry showed the presence of CD34 marker (Figure 6 E) and Human

Herpes Virus 8 (Kaposi Sarcoma Human Virus) (Figure 6 F) in the nuclei of the tumor cells. Immunostaining with SMA (Smooth Muscle Actin) showed that only a part of the vascular structures had its own walls. The Ki 67 index was 15%, indicative of slow neoplastic growth. The enlargement of the mesenteric lymphatic ganglia was due to reactive inflammatory changes.

For the differential diagnosis, we considered stromal gastrointestinal tumors (DOG 1 marker negative) and ectodermal tumors (S100 marker negative).

Therefore, the histopathological assay concluded that the ulcer-like mass received for evaluation was, in fact, a rare type of intestinal tumor: Kaposi's sarcoma.



**Figure 6 (courtesy of Sabina Zurac, MD; Pathology Department, 'Colentina' Clinical Hospital, Bucharest, Romania)**

- A-** Small bowel wall; submucosal solid tumor proliferation; dilated vascular structures (arrow); H&E 40x
- B-** Mesenchymal tumor morphology; spindle cells with fascicular alignment (arrow); H&E 100x
- C-** Enlarged polymorphic nuclei showing granular chromatin with rare, asymmetrical mitoses; intracytoplasmic vascular clefts pathognomonic for Kaposi's sarcoma (arrow); H&E 400x
- D-** Hemosiderin deposits in tumoral interstitial space (arrow); H&E 400x
- E-** Positive immunostaining for CD34 in tumor cells
- F-** Positive immunostaining for HHV8 (human herpes virus 8) in the nuclei of tumor cells

## Discussion

The small incidence of primary intestinal tumors is explained by several factors:

- the rapid intestinal transit which limits the contact with carcinogenic substances,
- the dilution of carcinogens due to voluminous intestinal chyme,
- the increased intestinal epithelial turn-over rate of about 2-3 days,
- the protective alkaline pH of intestinal secretions,
- a reduced intestinal bacterial population with reduced metabolic activity which leads to a reduced number of carcinogenic by-products,
- a higher density of lymphoid tissue in the intestine,
- a competent B lymphocyte population which secretes high Immunoglobulin A levels to protect against carcinogenic viruses,
- high levels of intestinal benzyl-peroxidase neutralises carcinogenic agents [2,6].

The epidemiologic evidence reported in the literature highlights the challenging diagnosis of a PIT.

Most of the benign lesions are asymptomatic and often found incidentally. On the contrary, malignant tumors are symptomatic and amount to 75% of the symptomatic surgical cases, although they become apparent in the rather late stages of the disease [1,6]. Only 31% of the patients suffering from a PIT are diagnosed in early stages to benefit from the 83% 5-year survival rate when the multimodality treatment is followed. The majority of the patients in question are diagnosed in an advanced stage, when the 5-year survival rate may drop to about 14% in cases of adenocarcinoma [1].

The symptoms at presentation are vague and non-specific, such as: dyspepsia, nausea, anorexia, König's syndrome, diffuse and intermittent abdominal pain, signs of GI bleeding (hematochezia, melena, hematemesis), signs of perforation, signs of intestinal obstruction (due to intraluminal mass, adhesions, intussusception or volvulus). [1,2,6]

The relative inaccessibility of the small bowel makes the paraclinical diagnosis no less challenging than the clinical one. The first non-

invasive, cost-effective option is the barium radiography which has a sensitivity of 53 % - 83% [1]. Upper and lower GI endoscopies have high sensitivity and allow biopsy sampling, but jejunal and ileal lesions are still out of reach. An alternative to this constraint is the double-balloon enteroscopy, which can help properly localise the lesion. It also offers the opportunity to mark the discovered pathologies with intramucosal dye to guide a future surgical intervention; nevertheless, it associates a high risk of iatrogenic perforation [7]. The video capsule endoscopy, has a sensitivity of 89% to 95%; in spite of the high detection rate, its considerable cost prevents it from being the first option when investigating this pathology [8]. CT and MRI enterography have even higher sensitivity, above 95%, but there is no clear consensus regarding the indications [1,9].

Several pathologies should be considered for the differential diagnosis: colonic neoplasm, intestinal perforation, colonic polyps, familial polyposis, Peutz-Jeghers syndrome, villous adenoma, stromal GI tumor, leiomyoma, lipoma, fibroma, hamartoma, hemangioma (Rendu-Olser-Weber disease), endometriosis, Crohn's disease, ulcerative colitis, Whipple's disease, malakoplakia, radiation enteritis [1,2,6].

In 50% of the cases only, the pre-operative diagnosis is accurate. Therefore, the surgical exploration serves as both a diagnostic and a curative tool. A thorough surgical exploration of the peritoneal cavity and its contents must be performed [1,2,6].

When identifying the intestinal lesion, the proposed surgical technique is the segmental resection of the small bowel with a 10 cm safety margin, followed by the removal of the mesenteric drainage area.

The multimodality treatment after the surgery continues with patient's referral to an oncologist pending treatment after the histopathological assay of the surgical specimen.

Less than 1% of all PIT are small bowel KS. It is believed that KS is induced by HHV8. HHV8 infects B lymphocytes, macrophages, epithelial and endothelial cells. It is transmitted supposedly by saliva or from childbirth in high incidence regions (Africa), whereas in low incidence regions (Europe, North America) it is transmitted through sexual contact (homosexual activities have a higher chance of transmitting

the virus then heterosexual ones). Blood borne transmission has not yet been demonstrated [10, 11]. HHV8 is also a pathogenic factor in B-cell lymphomas associated with AIDS, such as primary effusion lymphoma and multicentric Castleman's disease [10].

Four types of KS have been described: the classic Mediterranean, the African endemic, the immunosuppression post-transplant and the epidemic HIV-associated form.

The African form affects HIV negative individuals residing in Africa and, although indolent, it can later manifest itself more aggressively with generalised lymphadenopathies and multiple visceral involvement leading to a lower survival rate.

The classic KS is typically found in elder men from East European and Mediterranean regions. It is not associated with HIV and manifests itself mainly as cutaneous lesions, predominantly on the lower limbs. Frequently asymptomatic, it is rarely progressive, with less than 10% of internal organ involvement [4,11].

The epidemic HIV-associated form commonly occurs in AIDS patients who are not receiving HAART (highly active antiretroviral therapy)[12,13]. It is reported that increased HIV viral loads and decreased CD4 counts are prognostic factors for the severity of the disease. Visceral involvement follows quickly after more pronounced cutaneous lesions that can have disfiguring consequences. More than 50% of the AIDS patients who have visible lesions on the skin will develop GI tract KS [4,12]. The proposed management in this case combines endoscopy and HAART [4,5,12,13]. A rare case was reported when KS appeared at the site of the surgical wound in a patient with AIDS [14].

Immunosuppression, either post-transplantation or in chronic inflammatory diseases (such as Crohn's disease or ulcerative colitis) can increase the risk of KS. In such cases, KS has a 400-500 times higher incidence compared to the general population [15]. In order to suspect this form of KS, the patient must be on a long standing immunosuppression therapy and HIV infection must be ruled out. This is a more aggressive form of KS with more than half of the patients manifesting visceral lesions that can appear before the cutaneous ones [11].

There are reported cases in which KS arose from lesions previously caused by inflammatory bowel diseases [16-18]. If the complications of visceral KS are not acceptable, the medical team can consider tuning down or interrupting the immunosuppression therapy when able. In most cases of inflammatory bowel diseases, this course of action is the most fruitful [5,16,18].

The uniqueness of this case, apart from its rarity, consists of a visceral KS involvement of the small bowel in an immunocompetent, HIV negative elderly man with a non-specific initial endoscopic biopsy of the lesion. As a consequence, we believe that our patient suffered from a classic (sporadic) form of KS which was surgically treated without complications and with histopathological confirmation of tumor-free resection borders.

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## Conclusion

In cases of scarce GI symptomatology with positive faecal occult blood test, we should bear in mind that only 50% of the pre-operative diagnoses are correctly made and surgical exploration remains the most prudent course of action due to the asymptomatic nature of the intestinal tumors, when specific signs manifest themselves in the much more advanced stages. In patients with chronic inflammatory bowel diseases who stop responding to immunosuppression treatment it is prudent to consider in the differential diagnosis a KS, as it is highly angiogenic and its growth seems to be favoured by states of chronic inflammation and immunosuppression [16,17,18].

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