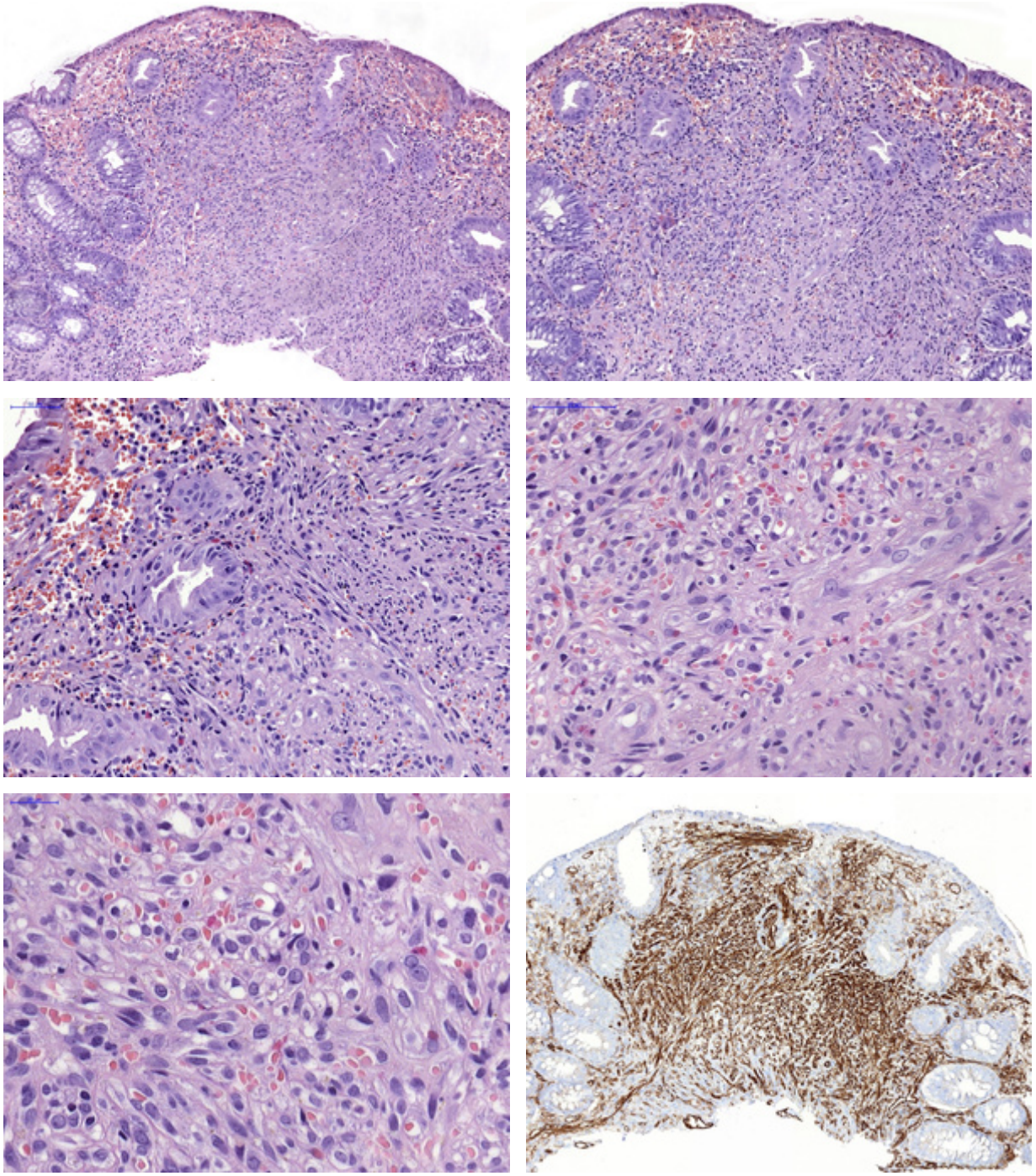
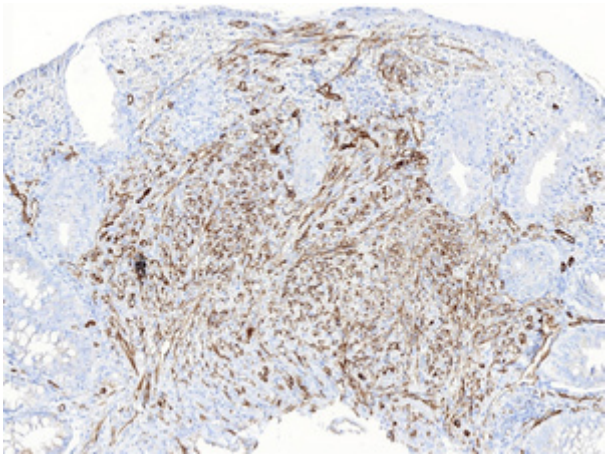
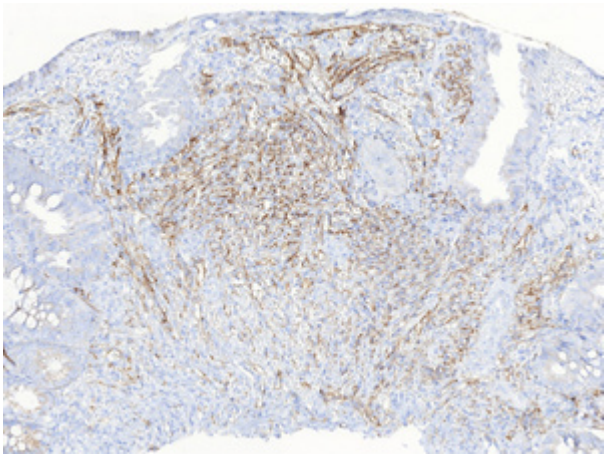
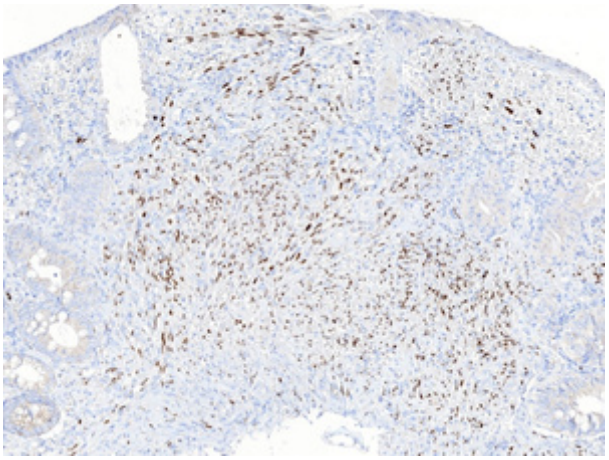
February 2015

Biopsy material from the colon of a 63-year-old male with psoriatic skin changes lasting for 30 years and treated with methotrexate.

What is your diagnosis?



Diagnosis:

Kaposi's sarcoma.

Comment:

On the microphotographs we can appreciate a lesion situated in the colonic mucosa. The lesion is composed of minimally atypical spindle cells forming slits filled with erythrocytes. In some areas extravasated red blood cells and eosinophilic hyaline bodies are found (Panels A-E). Upon immunohistochemistry, the spindle cells are positive for CD31 (Panel F), CD34 (Panel G), D2-40 (Panel H) and human herpesvirus 8 (HHV-8; Panel I).

In our patient endoscopy revealed a total of four lesions, two in the ascending and two in the descending colon. Several skin lesions were also found, excised and sent for examination. Histological findings were compatible with those of the colonic lesions.

Kaposi´s sarcoma (KS) is a malignant spindle-cell neoplasm caused by HHV-8 infection. The lesion was first described by Moritz Kaposi in 1872. It is classified into four subtypes according to different clinical manifestations, prognosis and treatment: classic KS (more indolent), AIDS-related KS, immunosuppression-associated KS and endemic KS.

KS lesions are usually found on the skin, but other sites can be involved, mostly oral mucosa, gastrointestinal and respiratory tract. Characteristic lesions are raised, red to violet nodules. Gastrointestinal lesions can be seen anywhere along the GI tract. They are often asymptomatic and clinically silent, but sometimes pain, weight loss, vomiting, diarrhea and GI bleeding or obstruction can occur. On histology, we see typical changes, such as proliferation of spindle cells that are separated by prominent, slit-like vascular spaces containing erythrocytes. Extravasated red blood cells, haemosiderin-laden macrophages, lymphocytes, and PAS positive intracellular eosinophilic hyaline bodies can also be present. Mitoses are rare and cellular pleomorphism is usually absent. The lesion shows positivity for CD31, CD34, Factor VIII-related antigen, D2-40 (podoplanin) and HHV-8.

In differential diagnosis we should consider arteriovenous malformations, pyogenic granuloma and other vascular lesions together with other spindle cell neoplasms. Treatment of KS is based on the subtype and clinical presentation. Most of the patients can be treated with local resection or radiotherapy. In HIV positive patients, antiretroviral therapy is used. In immunosuppressed or immunodeficient patients the reason for immune response dysfunction should be identified. In patients under immunosuppressive therapy the treatment should be discontinued, which was the case in our patient.

For further reading:

- Kaposi M. Idiopathisches multiples Pigmentsarkom der Haut. Arch Dermatol Syph 1871;4:265–273.
- Campbell DM, Rappocciolo G, Jenkins FJ, Rinaldo CR. Dendritic cells: key players in human herpesvirus 8 infection and pathogenesis. Front Microbiol. 2014;5:452.
- Thrash B, Patel M, Shah KR, Boland CR, Menter A. Cutaneous manifestations of gastrointestinal disease: part II. J Am Acad Dermatol. 2013;68:211.e1-33.
- Antman K, Chang Y. Kaposi’s sarcoma. N Engl J Med 2000; 342:1027-1.

Presented by:

Dr. Ayşegül Erol and Dr. Arzu Ensari, Ankara, Turkey, and by Dr. Iva Brcic, Zagreb, Croatia