Kaposi's Sarcoma in the Spleen

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Özet: *DALAKTA KAPOSİ SARKOMU*

Multifokal neoplazm olarak bilinen Kaposi Sarkomu'nda (KS), primer splenik yerleşim son derece nadir olup günümüze kadar sadece bir vaka bildirilmiştir. Bu yazıda dalakta bir kadın hastayı takadim etmekteyiz. Kaposi Sarkomu'nun bu alışılmamış formunda ilginç olan özellik geniş dalak tutulumuna rağmen deri lezyonların yokluğudur.

Anahtar kelimeler: Kaposi sarkomu, dalak

Summary: *In Kaposi's sarcoma (KS) which is known to be multifocal neoplasm, primary splenic involvement is extremely rare and only one case has been reported to date. In this presentation, we describe a 50 year old female patient who developed a mass in the spleen that proved to be Kaposi's sarcoma afterwards. In such an unusual clinical form of KS, the interestin feature is the absence of skin lesions despite wide involvement of spleen.*

Key words: Kaposi's sarcoma, spleen.

Kaposi's sarcoma (KS) a malign tumor of the skin is endemially seen in Central Africa (1,2). In some studies from Uganda, KS has been reported to account for approximately 20% of all malignancies (2). On the other hand, Kaposi's sarcoma is the most common malignancy encountered in patients with AIDS (3, 4). Also, KS accors as an important complication in renal transplant patients recieving immunosuppressive agents (5, 6, 7). When skin lesions are known to be the most frequent findings of KS, some times, visceral manifestations have also been reported (8). We present here a case of KS arising from spleen to emphasize an uncommon but a likely entity in terms of differential diagnosis in abdominal masses.

CASE REPORT

A 50 year-old woman was admitted to the hospital in November 1992, with a five month history of abdominal pain in the left side. Her pain had become severe and had been radiating to back for the last two weeks. She denied nausea, vomiting, fever or weight loss. On physical examination, no skin lesions of Kaposi's sarcoma were notied on arms, legs, or any part of the body. Spleen tip was palpable 8 cm below the left costal margin, but liver was no palpable. Abdominal ultrasonography revealed ascites and gave information regarding splenomegaly, hepatomegaly and enlargerd abdominal lymph nodes. Computed tomography helped tumoral masses oringinating from spleen to be demonstrated. Initial laboratory investigations disclosed the following values; hemoglobin 12 g/dl, leukocyte count 5.800/mm^3, platelet count 150.000/mm^3, erythrocyte sedimentation rate 12 mm/h. The coagulation profile was normal and liver function tests showed normal values. The patient was clinically diagnosed as having abdominal lymphoma. At operation, approximately 2 L of ascites fluid was noted in the abdominal cavity and all was removed. Spleen was observed much greater than normal and there were also tumor nodules being felt within it. Similar, however smaller nodules were in the liver superficially. Abdominal lymph nodes, especially celiac nodes showed enlargment. After removal of spleen, a liver biopsy was made. Operation was completed with biopsies from celiac lymph nodes Although the patient carried no risk factors regarding AIDS, some investigations were performed which failed to reveal any evidence of AIDS. In addition to surgery, we started a combined therapy of vincristine (1.4 mg/m^2), adriamycin (20 mg/m^2) and bleamycin (10 units/m^2). On control examination, one year later, the patient remains well and is apparently free of any symptoms showing dramatic improvement.

Macroscopic Findings : The materials of splenectomy which weighed a total of 1450 g measured

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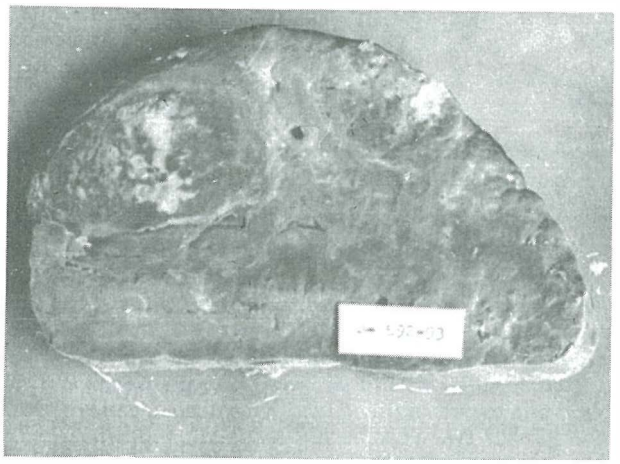

Fig. 1. Macroscopic appearance of thespleen showing two tumor nodules which are well demarcated from the surrounding tissue.

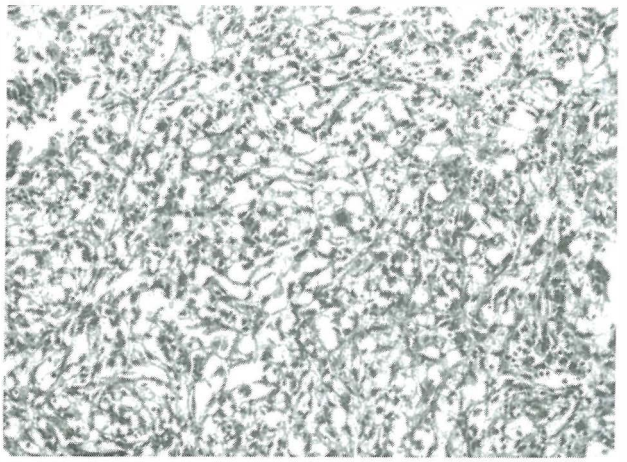

Fig. 2. Tumor composed of thin vascular structures lined by single layer epithelium, most of which contained red ccells in their lumens and spindle cells which arrange between slitlike vascular spaces. Notice atypia and mitoses in the middle. (hematoxylin - eosin x 200).

25x14x8 cm. On sectioning the spleen revealed two tumor nodules, larger on of which measuring 8x6x5 cm and smaller one of which is 4 cm in diameter. Both nodules were also displaying central necrosis, peripheral irregularity, whereas including some solid areas (Fig. 1).

Microscopic Findings : Microscopic examination of sections prepared from tumoral process revealed proliferation of atypic spindle cells arranged along the vascular structures, thin splits in shaped. Also, afluent erytrocytes were noticeable in the lumen of vessel.

Tumor cells showed several mitotic figures in some areas and PAS positive globules were present in cytoplasms of some (Fig.2). It was possible to see wide necrotic areas sometimes. Microscopic appearance on the sectioning of the liver was similar to that of the spleen. Furthermore, there was a response of mononuclear cell at the periphery of tumor, No metastases of tumor to the abdominal lymph nodes were demonstrated.

DISCUSSION

At least three distinct clinical forms of Kaposi's sarcoma have been described : African endemic, classic and epidemic (1, 2, 3, 4, 5). African endemic type of Kaposi's sarcoma (KS) in known as having better prognosis than others and is particularly presented with skin nodules at the lower extremites (1, 9). In contrast, the other types of disease generally exhibit very aggressive features associated with internal visceral dissemination (4, 5, 6, 7, 10). Without any cutaneous lesions, some patients with KS can pose internal organ involvements which remain unsuspected for years (10, 11). Small bowel, stomach, esophagus and colon are the most affected organs (12).

In disseminated form of KS, splenic involvement has well been described (10, 13). However, Kaposi's sarcoma of the spleen is a very rare condition as the primary focus for malignancy. The fisrt description of primary splenic involvement in Kaposi's sarcoma was documented in detail by sarode et al (8). In a wide description of that case, obvious involvement of spleen by single large mass which was previously thought to be abdominal lymphoma had been emphasized. In addition, wide spread of other internal organs had also occurred. Despite treatment, patient had rapidly become poor and died.

We have also described a primary case of KS arising in the spleen without skin lesions. Interestingly, two tumor nodules were present in the spleen (Fig 1). Except for that some metastatic nodules were present in the liver and some enlarged abdominal lymph nodes, all other abdominal organs were apparently clear. Primary splenic mass which was displayed on computed tomography was also confirmed grossly at the opiration, however definitive diagnosis could be made only after histologic examination of surgical specimens. To our knowledge, primary site of

the tumor was spleen and nodules in the liver were likely metastates from the spleen.

Kaposi's sarcoma is a tumor histologically charaterized by the presence of interlacing bundles of spindle cells and vascular channels (14, 15). However, angiomatous variant of KS has been documented to contain well-formed capillaries in focal areas (15, 16). In the patient presented here the tumor tissue contained criss-crossing bundles of spindle ells intermingled with the vascular spaes which is quite demonstrative for KS. In addition, some tumor cells contained PAS positive hyaline globules within their cytoplasms (Fig. 2). This is one of the most important diagnostic findings of KS. All histologic findings together seemed to be suffiient for making a firm diagnosis of Kaposi's sarcoma in the spleen.

Kaposi's sarcoma may be confused with other epithelioid vascular tumors such as hemangioma, hemangioendothelioma and angiosarcoma (17). Among these entities, the most likely differential diagnosis lay between Kaposi's sarcoma and primary splenic angiosarcoma (14). Histological resemblence of both tumors has been explained by a theory based on special figure of disease is that of vascular or lymphatic endothelium as the cell type of the origin (18). Our case has two main histologic evidences of KS. These are presence of characterestic spindle cells with slitlike vascular spaces and identification of the PAS positive globules within the cytoplasma of tumor cells which provide strong support for the diagnosis of KS. Histologically, the current tumor revealed similar features and we rejected the diagnosis of angiosarcoma.

Although the etiology of KS is unclear, a variety of factors such as immune deficiency, various infective agents, environmental factors have been proposed (18). Virtually, there seems to be a close relationship beetween KS and immune deficiency (10). On the other hand KS has been found at a high rate of 30% in patients with the AIDS (3). Also, incidence of KS is much greather in transplant recipients treated with immunosuppressive agents than normal population (5, 6, 7). Our patient had neither of these factors that could be cause for KS. In treatment of KS, various chemotherapeutic agents have been used and satisfactory results have been reported (19,20). We also used a combination of vincristine, adriamycin and bleomycin after surgery. Patient remains free of disease one year after operation and so we hope long term remission.

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