Case Report

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A Case of Neglected Classic Kaposi's Sarcoma

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Introduction

Kaposi's sarcoma (KS) is a rare disease in the community. It was first described in 1872 as an idiopathic, multipigmented sarcoma of the skin [1]. It is a vascular or lymphatic-focused tumor type originating from the endothelium, caused by Human Herpes Virus 8 (HHV8), called KS-associated herpes virus (KSHV) [2]. There are 4 clinical subtypes of the disease. These are: Classical KS, African endemic KS, iatrogenic KS, and AIDSrelated (epidemic) KS. It usually occurs with pink-red patch and blue-purple-black nodule, polyp or plaques. Most of the cases in classical KS are over 50 years old. Although the female/male ratio varies between studies, it is more common in males [3]. Here, a neglected case of Classic KS is presented.

Case report

An 84-year-old female patient was admitted to the dermatology outpatient clinic due to purplish swelling on both feet. In the anamnesis, it was learned that these complaints had been present for three years and started on the soles of the feet and gradually spread to the upper parts of the feet. On dermatological examination, purplish solid papules and nodules in the dorsal and plantar regions of both feet; In addition, skin-colored nodular lesions were observed in both plantar regions. (Picture 1) (Picture 2). The lesions of the patient, who had received various local treatments before, did not regress. The patient reported that there was no pain in this region. There were no features in her CV and family history. Routine hematological examinations and biochemical tests were within normal limits. The patient was not HIV positive. Excisional biopsy was taken from the patient with the preliminary diagnosis of Kaposi's sarcoma. In the histopathological examination of the skin and subcutaneous tissue, a spindle cell neoplasm with nodular development was observed in the superficial dermis. Neoplastic cells are cells with light sandy chromatin, usually indistinct nucleoli, oval-spindle nuclei, syncytial cytoplasm, and do not show significant pleomorphism. The presence of intracytoplasmic hyaline globules was noted in some of them. Mitotic activity was evaluated as 4/10 BCI. The neoplasm was accompanied by inflammatory cells rich in lymphocytes. In immunohistochemical examination; neoplastic cells are nuclear positive with HHV-8, diffuse and strongly positive with CD34; It was found to be negative with SMA. Ki-67 proliferation index reached 10% in focal areas. With all these data, a diagnosis of Kaposi's sarcoma was made. Cryotherapy was applied to hyperkeratotic areas and some regression was observed. The patient who was scheduled for radiotherapy was referred to the Oncology department.



Picture 1: Purplish solid papules and nodules in the dorsal regions of both feet.



Picture2: Purplish solid papules and nodules in the plantar area of the left foot, as well as skin-colored hyperkeratotic nodules in both plantar areas.

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Picture 3: Vascular neoplasm consisting of spindle cells is seen. (x200, Hematoxylin&Eosin).



Picture 4: The fascicular pattern of extravasated erythrocytes, hyaline globules and oval-spindle cells is observed. (x400, Hematoxylin&Eosin).



Picture 5: Widespread positive expression with immunohistochemical CD34 (x200, CD34).



Picture 6: Diffuse positive nuclear expression by immunohistochemical HHV-8 (x400, HHV-8).

Discussion

Classical KS mostly occurs multiple and bilaterally. Purple-blue or red-brown nodules and plaques formed in the extremities tend to coalesce over time. In a recent study [4], it was reported that 70% of the lesions are localized in the lower extremities. Classical KS progresses slowly. In our case, the lesions were limited to the lower extremities and developed over a long period of time. There are two types of approaches, local and systemic, in the treatment of classical KS. However, there is no curative treatment. Local treatment methods include surgery, intralesional vinblastine, topical retinoid injection, cryotherapy, and radiotherapy. Factors such as the extent, location, and rate of clinical change of lesions are determinative in the choice of local treatment [6] Rapidly progressive KS is characterized by the development of 10 or more new cutaneous lesions per month. In addition, pulmonary KS, symptomatic visceral involvement and lymphedema are indications for systemic chemotherapy. Although the first choice chemotherapy agents for KS are pegylated liposomal doxorubicin and paclitaxel, cytotoxic agents may cause significant side effects, especially in elderly patients [6]. Classical KS is different from our case. It mostly occurs in elderly male patients. Treatment options vary according to clinicians' experience and disease stage. For patients with limited extent and in the early stages, observation without treatment or local treatments are preferred. Classical KS is very sensitive to radiotherapy. Radiation therapies are an appropriate treatment option for patients presenting with multifocal but relatively localized KS [7]. In elderly patients with limited lesions, only follow-up without specific treatment is also an option [8]. For our case, five cycles of cryotherapy were applied at two-week intervals. If deemed necessary for some persistent lesions, it was considered to be evaluated together with Oncology in terms of applying radiotherapy.

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