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CASE REPORT

Atypical presentation of Kaposi's sarcoma in the external ear

Dış kulakta Kaposi sarkomu: Olgu sunumu

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We presented a case of Kaposi's sarcoma that occurred in the external ear of a 36-year-old Caucasian man. He was otherwise healthy without a history of any predisposing factors. He had a nodular lesion in the left ear, of three-month duration. The lesion was excised completely. Histologic and immunohistologic findings were consistent with a diagnosis of Kaposi's sarcoma. Serologic analyses were negative for anti-HIV antibody and anti-cytomegalovirus IgM and IgG and blood count was normal. Differential count of leucocytes and immunoglobulin electrophoresis were normal. During a two-year follow-up, no recurrences, development of new lesions, or HIV seroconversion were detected. To our knowledge, this case is the first to report a solitary lesion of Kaposi's sarcoma occurring in the helix of the ear in a healthy young patient.

Key Words: Ear, external; ear neoplasms; sarcoma, Kaposi/diagnosis/pathology.

Bu makalede, 36 yaşındaki sağlıklı bir erkek hastada kulak kepçesinde saptanan Kaposi sarkomu sunuldu. Hastanın sol kulağında üç aydır var olan nodüler lezyon için predispozan herhangi bir faktör belirlenemedi. Lezyon tümüyle çıkarıldı. Histolojik ve immünhistolojik bulgular Kaposi sarkomu ile uyumluydu. Serolojik analizlerde anti-HIV ile anti-sitomegalovirüs IgM ve IgG antikorları negatif bulundu. Kan sayımı normaldi. Lökosit altgruplarının sayımı ve immünglobulin elektroforezi normal sonuçlar verdi. İki yıllık izlem dönemi içinde hastada lezyon nüksü, yeni lezyon gelişimi, ya da HIV serokonversiyonu gözlenmedi. İlgili literatürlerin taranmasında, kulak kepçesinde Kaposi sarkomuna ait soliter lezyonun bildirildiği genç sağlıklı bir olguya rastlamadık.

Anahtar Sözcükler: Kulak, dış; kulak neoplazmları; sarkom, Kaposi/tanı/patoloji.

Kaposi's sarcoma (KS), first described by Moritz Kaposi in 1872, was an unfamiliar entity to most physicians in the past.[1] It has gained popularity since acquired immunodeficiency syndrome (AIDS) became a widespread concern throughout the world. The classic form of the disease occurs mainly in elderly people in Europe and Mediterranean countries, without any known association with immunodeficiency.[2] Other types are endemic (African), iatrogenic (related to immunosuppression), or epidemic (AIDS-related).[1] In this case report, a young Caucasian male patient with KS is described, that presented as a solitary lesion in the ear.

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CASE REPORT

A thirty-six-year-old man presented with a nodular lesion in the left ear, of three-month duration. He did not have complaints of any suspicious lesions in other parts of the body, nor a history of blood transfusion, transplantation, or immunosuppressive therapy. He was married and refused any inappropriate sexual practices. On physical examination, there were no abnormal findings except for the presence of the nodule in the scapha of the left ear. The lesion was pinkish-brown in color, measured 6 mm in diameter, and was surrounded by a wide brownish hue (Fig. 1). A complete excision was performed with a two-millimeter margin. Histologic examination revealed a well-defined nodular lesion in the dermis (Fig. 2), with a discernible spindle-cell component, that initially appeared around the proliferating vascular channels. At a higher magnification (X40), slit-like vascular spaces containing erythrocytes were typical in longitudinal sections. There were ectatic vessels and chronic inflammatory cells composed of lymphocytes and plasma cells located at the periphery (Fig. 3 and 4).

Immunochemistry was performed using the standard three-step streptavidin-biotin complex technique. The tumor cells were positive for CD34 (QB End/10, Neomarkers, Fremont, CA, USA), but negative for F8 (Factor VIII-Releated Antigen-VWF, N1505, Dako, USA), cytokeratin (AE1/AE3, N1590, Dako), actin (Anti-Human Muscle Actin, N1567, Dako), and S100 (N1573, Dako). These findings were consistent with an immunohistologic diagnosis of KS. Serologic analyses were negative for anti-HIV antibody and anti-cytomegalovirus IgM and IgG and blood count was normal. Differential count of leucocytes and immunoglobulin electrophoresis yielded normal results. During a two-year follow-up, no recurrences, development of new lesions, or HIV seroconversion were detected.

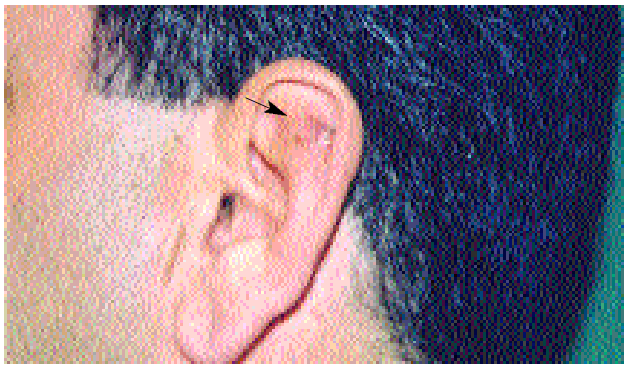

Fig. 1 - *The lesion was localized in the scapha of the left ear. It was pinkish-brown in colour and was surrounded by a brownish discoloration.*

DISCUSSION

The association between Kaposi's sarcoma and AIDS is well-documented. It is endemic in African regions. It is also encountered among elderly individuals in Europe and Mediterranean countries, without any known association with AIDS. Kaposi's sarcoma-

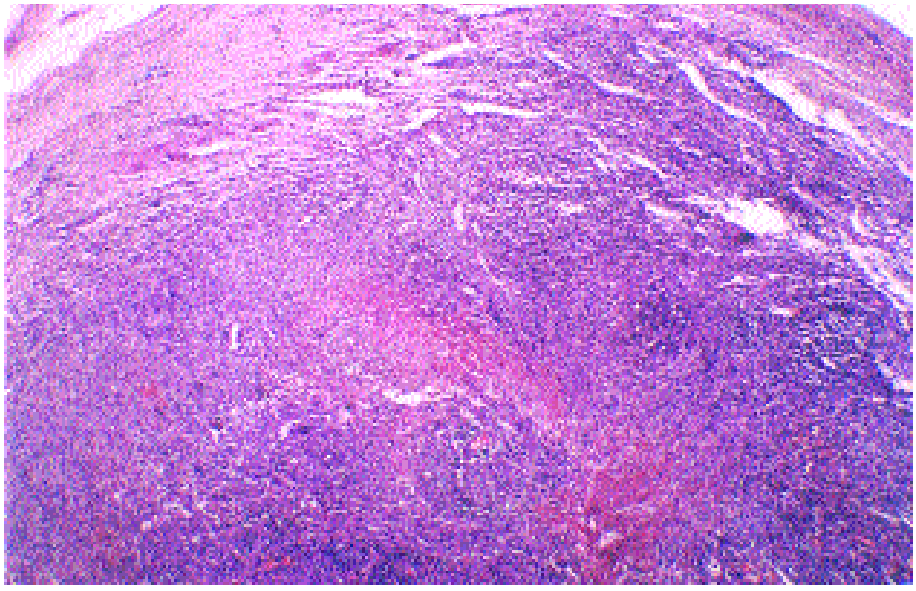

Fig. 2 - *The lesion was distinctly nodular. Crescentic vessels and chronic inflammatory cells were observed at the periphery (H-E x 10).*

associated herpesvirus (KSHV) or human herpesvirus 8 (HHV-8) have been identified in both HIV-associated and classical KS.[1,3,4] Human herpesvirus 8 is epidemic among people of Mediterranean origin, including Turkish population,[5,6] but its presence may not necessarily cause any disease in immunologically competent individuals.[7] Aside from HHV-8, cytomegalovirus has also been identified as an etiologic factor for KS, which is commonly seen in normal population.[8,9] Environmental and occupational factors may also play a role in the development of soft tissue sarcomas including KS.[10,11]

To our knowledge, the case presented here is the first to report a solitary lesion in the helix of the ear in a healthy young white man. The age of the patient was below that reported for those with classical KS, ranging from 40 to 70 years of age.[1] Although other types of KS occur at younger ages, the disease usually coexists with several predisposing factors, including HIV seropositivity, a history of transplantation, or having an African origin, none of which was seen in our patient. In addition, the characteristics and the site of the lesion were different from those typically observed in the lower

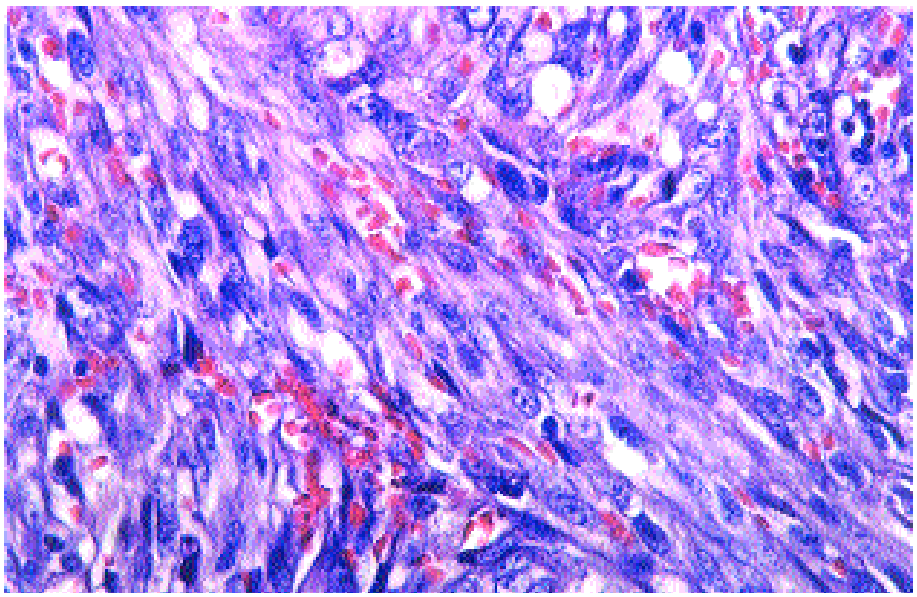

Fig. 3 - *Spindle cells separated by slit-like vascular spaces containing erythrocytes and spindle cells with a few hyaline globules (H-E x 40).*

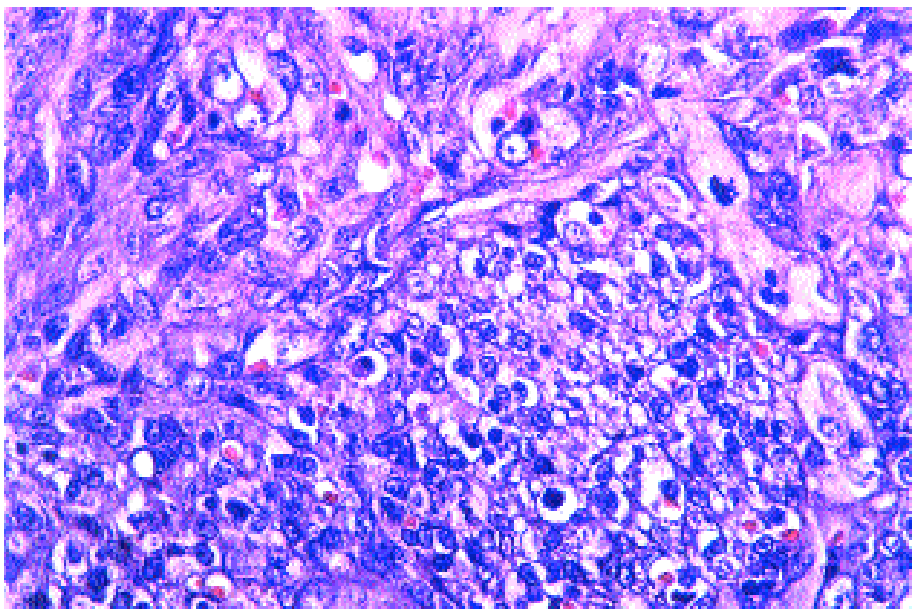

Fig. 4 - *Pleomorphic spindle cells with mitotic figures (H-E x 40).*

extremities of older individuals in classical KS, and from disseminated forms occurring in immunosuppressed patients. So far, only three cases of KS have been reported in the helix of the ear without a history of immunodeficiency,[12-14] but those patients were at older ages. The appearance of a solitary lesion in the helix of the ear in a healthy young patient may be explained by a sporadic cutaneous involvement caused by a viral infection or environmental factors.

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