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Dermatofibrosarcoma Protuberans of the Scalp: A Relatively Unusual Site of Occurrence

Kafa Derisinde Dermatofibrosarkoma Protuberans: Oldukça Nadir Bir Yerleşim Bölgesi

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ABSTRACT

Dermatofibrosarcoma protruberans is an uncommon low grade soft tissue tumor comprising of only 5% of adult soft tissue sarcomas. Scalp is a relatively uncommon site of occurrence for this tumor. Its identification at this site and differentiation from benign fibrous histiocytoma is important due to the propensity of recurrence and metastasis in the former unless a wide, optimal surgical margin is ensured during excision.

Key Words: Dermatofibrosarcoma protruberans, Scalp

ÖZ

Dermatofibrosarkoma protruberans yetişkin yumuşak doku sarkomlarının sadece% 5'ini oluşturan nadir bir düşük dereceli yumuşak doku tümörüdür. Kafa derisi bu tümör için oldukça nadir bir oluşum bölgesidir. Eksizyon sırasında geniş, optimal cerrahi sınır sağlanmadığı taktirde rekürens ve metastaz eğilimi göstermesi nedeniyle, bu bölgede tanınması ve benign fibröz histiositomadan ayırt edilmesi önemlidir

Anahtar Sözcükler: Dermatofibrosarkoma protruberans, Kafatası derisi

Sir,

Dermatofibrosarcoma protuberans (DFSP) is a relatively uncommon, fibrohistiocytic low-grade cutaneous tumor, characterized by a slow, infiltrative growth pattern. It accounts for approximately less than 5% of adult soft tissue sarcomas and less than 1% of all malignant tumors of the head and neck (1). It has a marked tendency to recur locally after surgical excision. Rarely, they metastasize to regional lymph nodes or to distant sites.

A 24-year-old male presented with a 4 cm diameter nontender, firm, nonpulsatile, slowly growing cutaneous nodule of 10-month duration in the left parietal region of the scalp. No history of trauma could be elicited. The nodule was fixed to the overlying skin but was movable over the deeper structures. Transillumination and impulse on cough was absent. Skull X-ray did not reveal any involvement of the underlying bone.

Grossly, the excised nodule was composed of multiple skin-covered greyish-white tissue bits. On histological examination, the tumor was located in the dermis with involvement of the deeper subcutaneous tissue. The epidermis overlying the tumor was unremarkable. The tumor was highly cellular consisting of spindle-shaped cells arranged in the characteristic storiform pattern of growth resembling a "straw mat". Nuclear hyperchromasia and few mitotic figures were noted. Pleomorphism, giant cells and foamy or hemosiderin-laden cells were absent. Skin adnexal structures such as hair follicles, sweat and sebaceous glands were seen entrapped within the tumor (Figures 1, and 2).

First named by Hoffmann as DFSP (2), this relatively rare tumor usually appears between 6-65 years of age while the maximum incidence is in the 2nd and 3rd decades (3). The most common site of origin is the skin of the trunk (50% to 60%) followed by the proximal extremities (20% to 30%). The head and neck region is again a relatively rare (10% to 15%) site of origin. However, no anatomical region is spared except for hands (4). Although regional lymph node metastases and distant metastases are rare, the tumor may have aggressive local invasion. A high recurrence rate of approximately 43% and 18% have been reported with conventional and wide (more than 2 cm margin) excisions respectively (4). The scalp is an uncommon site for this rare tumor. Recognition of this tumor in this site is important

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because of its propensity of recurrence and the chance of intracranial invasion as well. The ideal treatment modality for DFSP in this location is controversial. However, Sinha VD et al[5] described a case of DFSP in the scalp with underlying bone erosion and without any intracranial extension which was operated with a wide 3 cm safety margin, removal of the underlying bone along with a 3 cm margin and covering the skin deficit with a transposition flap. The case did not show any recurrence in the four-month follow-up (5).

DFSP should be differentiated from fibrous histiocytoma; the latter is a symmetric, small dermal tumor composed of a heterogenous population of mononuclear cells admixed with foamy, lipid-laden cells and/or hemosiderin. Moreover, fibrous histiocytomas rarely involve the subcutis, do not usually entrap the cutaneous adnexa and contain intralesional polarisable collagen. Immunohistochemical staining with CD34 and reciprocal chromosomal translocation, t(17;22), may prove helpful in difficult cases.

To conclude, we describe a relatively uncommon cutaneous tumor, in an also uncommon site, the scalp. Awareness of this tumor in rarely described and the site is important, given the propensity of recurrence and metastasis unless a wide, optimal surgical margin is ensured during excision.

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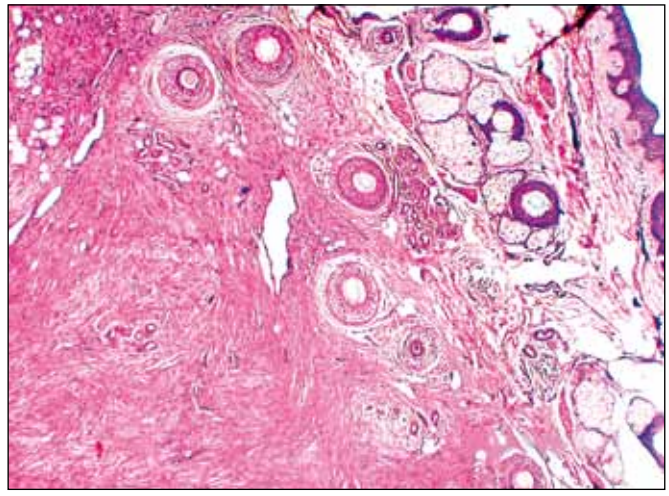

Figure 1: Tumor seen in the dermis with entrapment of skin adnexal structures; uninvolved epidermis seen in the upper right (H&E; x100).

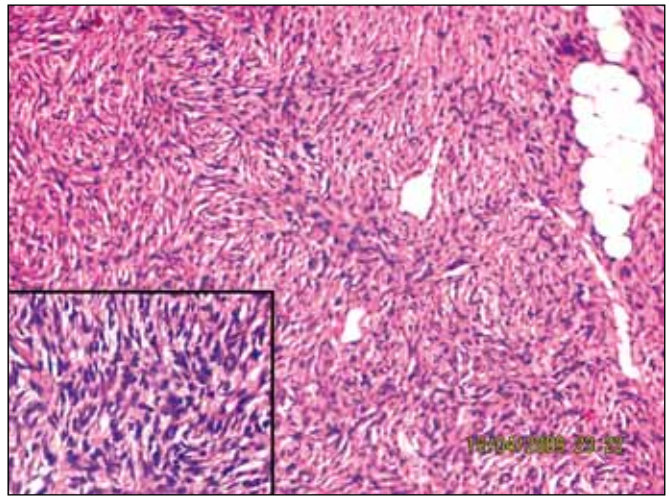

Figure 2: Tumor with characteristic storiform pattern, invading subcutaneous fat (seen on the right) (H&E; x200), inset shows monomorphic cells in another focus (H&E; x400).