Pleomorphic Malign Fibrous Histiocytoma of Auricle: Can size be a criteria in distinguishing pleomorphic malign fibrous histiocytoma and atypical fibroxanthomas

KULAK KEPÇESİNİN PLEOMORFİK MALİGN FİBRÖZ HİSTİYOSİTOMASI: Pleomorfik malign fibröz histiyositoma ile atipik fibroksantomun ayrımında tümör boyutu kriter olabilir mi?

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Summary

Differential diagnosis of pleomorphic malign fibrous histiocytoma and atypical fibroxanthoma sometimes may cause a diagnostic dilemma, because of their similar histopatologic and immunohistochemical features. Atypical fibroxanthomas rarely recur and metastasize. Tumor necrosis, depth of invasion, vascular invasion and high recurrence rate are important features which were detected in metastasizing case. A 65-year-old male patient who had a rapidly growing ulcerated nodular tumor on his right auricle exceeding 3 cm in diameter was presented here. In these tumors, to which were applied various therapeutic approaches according to the patients' condition, we discussed the importance of tumor size in differential diagnosis and giving appropriate name.

Key Words: Pleomorphic malign fibrous histiocytoma, Atypical fibroxanthoma

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Özet

Pleomorfik malign fibröz histiyositoma ile atipik fibroksantomun ayrımı her iki lezyonun da benzer histopatolojik ve immunohistokimyasal özelliklerinin olması nedeniyle bazen güçlük yaratabilir. Atipik fibroksantomada nüks ve metastaz nadiren görülür. Metastaz; tümör nekrozu, invazyon derinliği, vaskuler invazyon ve sık tekrarlama ile ilişkili bulunmuştur. Burada, 65 yaşında erkek hasta, kulak kepçesinde hızla büyüyerek 3 cm' yi aşan ülsere noduler kitle oluşması nedeniyle sunuldu. Hastanın durumuna göre değişen tedavi yaklaşımlarının uygulandığı bu tümörlerde, tümör boyutunun ayırıcı tanıda ve isimlendirmede faydalı olup olmayacağı tartışıldı.

Anahtar Kelimeler: Pleomorfik malign fibröz histiyositoma, Atipik fibroksantoma

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Malign fibrous histiocytoma and atypical fibroxanthomas (AFX) have similar histopathologic and immunohistochemical features. AFX is now accepted as a superficial form of malignant fibrous histiocytoma. It occurs most commonly in the sun damaged skin of elderly patients. Clinically, it is a pearly, erythematous, ulcerated nodule or plaque measuring less than 2 cm in diameter. It usually develops over the course of several weeks. AFXs recur infrequently and rarely metastasize (1). Helwig and May reported 8 cases of metastasizing AFX. In these cases, tumor necrosis, depth of invasion, vascular invasion and high recurrence rate are the

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important differantiating features found in metastasizing cases (2). Worrell et al. demonstrated that 13 of the 14 cases of AFX had diploid distribution of nuclear DNA, in contrast to this finding, the vast majority of malign fibrous histiocytoma (MFH) had been reported as aneupleuid (56 to 100 percent of cases) (3-5).

We reported an atypical tumor with fibrohistiocytic origin of auricle which had an unusual rapid growth pattern.

Case Report

A 65-year-old male patient was admitted to our hospital with a rapidly growing nodular lesion on his right auricle. The lesion occurred 6 months ago; however, it grew from 1.5 cm to 3 cm in diameter within 4 months. It was ulcerated and had heamorrhagic appearance. The tumor was totally excised, and 6 months later there was no recurrence. Histopathologically, epidermis was atrophic and ulcerated, and there was no grenz zone. The tumoral tissue was composed of pleomorphic histiocytelike cells and atypical giant cells with bizarre nuclei (Figure 1). Numerous mitotic figures including abnormal forms were observed, and storiform pattern was seen in some areas. There was dense hemosiderin deposition in the histiocytelike cells (Figure 2). In immunohistochemical staining, CD 68 was positive (Figure 3), S-100 and HMB-45 were negative.

Material and Method

Specimen was fixed in neutral buffered formalin and sections cut at 4μm were stained with hematoxylin and eosin. Immunohistochemical staining using the streptavidin-biotin-peroxidase complex technique with diamino benzidine chromogen substrate. Primary antibodies directed against CD 68 (ZYMED), anti S-100 (ZYMED), HMB-45 (DAKO) were used.

Discussion

AFXs have been reported under many different names including pseudosarcoma of the skin and paradoxical fibrosarcoma. Enzinger and Weiss have suggested that atypical fibroxanthoma can be regarded as superficial form of malignant fibrous histiocytoma (1). MFHs of soft tissues due to their

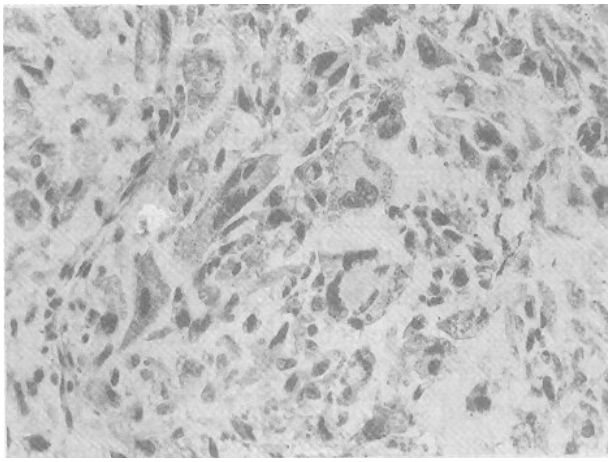

Figure 1. Tumoral tissue composed of mainly large pleomorphic histiocytelike cells rather than spindle cells. (This pigment was not stain with S-100 and HMB-45.)

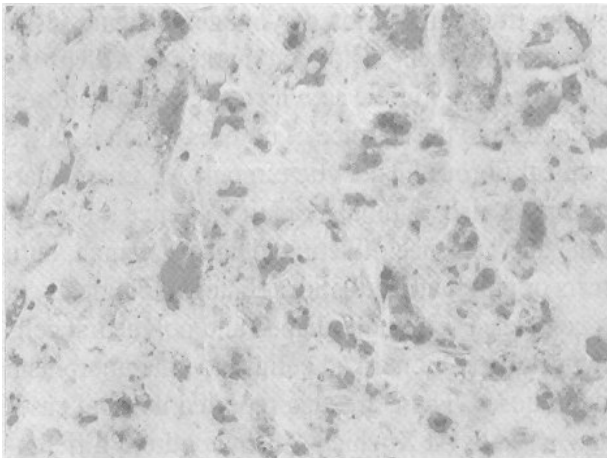

Figure 2. Dense hemosiderin pigment deposition with Prussion blue staining.

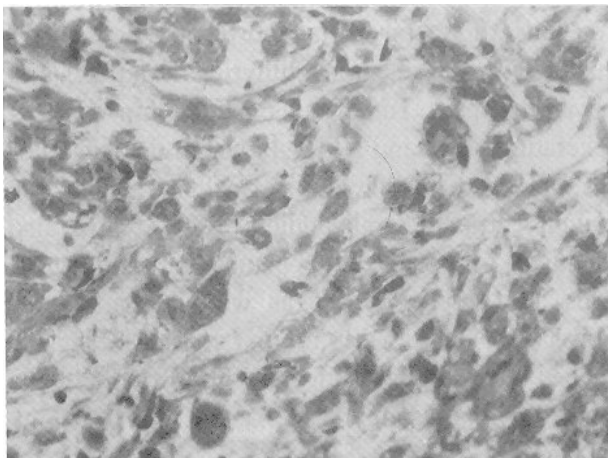

Figure 3. These bizarre multinucleated cells show CD-68 positivity. Some atypical mitoses are also evident.

deep localization and having bigger size have a recurrence rate of 50% and metastasize in 23 to 50% of cases (6).

Regardless of their different histogenesis, cutaneous fibrohistiocytic tumors when categorized according to their histopathologic features can be named as cutaneous fibrous histiocytoma, atypical fibrous histiocytoma and AFX (7). All of them generally behave in a benign fashion, except for only a small number of AFX which may produce metastatic disease and show infrequent recurrence. In one series of 140 cases there were 9 recurrences (8). This fact suggests a dillemma in diagnostic terminology regarding AFX.

Characteristically, AFXs appear on the head and neck of elderly men. In Fretzin and Helwig's report; however, a small group of lesions occured on the trunk and limbs of younger patients (8-9). The most frequent histologic marker of AFX is the high degree of mitotic activity and atypical mitoses, in contrast to the absence of mitoses which is called atypical fibrous histiocytic tumour. AFXs are characterized also with ulceration, absence of grenz zone and presence of actinic damage, trauma, burn scars or previous irradiation. Histopathologically, it is composed of spindle shaped and polyhedral cells characterized by marked cellularity, pleomorphism and mitoses. There are also reported unusual cases diagnosed as AFX which show chondroid differentiation and contain osteoclast-like multinucleated giant cells (6,10-12).

AFX is accepted as a tumor of fibrohistiocytic origin which is supported by the results of immunohistohemical stains. Tumor cells are positive for vimentin, factor XIIIa, and histiocytic markers such as lyzozyme, α-1-antithyripsin and α-1-anti chymothyripsin. Tumor cells fail to stain with cytokeratin, S-100 and HMB-45 which demonstrates that AFX do not have epidermal or melanocytic origin. Ultrastructural studies of AFX have also identified cells with the appearance of mesencymal cells and myofibroblasts as well as transitional forms (11). Our patient's tumour consists of bizarre shaped pleomorphic histiocytlike cells which were stained with a histiocytic marker, CD 68.

The histopathologic differential diagnosis of AFX includes all pleomorphic spindle cell tumors of the skin, such as the spindle cell variant of squamous cell carcinoma, malignant melanoma and leiomyosarcoma (1). We differentiated our case from malignant melanoma with negative staining of S-100 and HMB-45. It is also easy to distinguish our case from the other spindle cell tumors because spindle cell component had formed small areas.

Treatment modalities that have been used for AFX include excision, Moh's surgery, electrosurgery, radiation, wide excision followed by radical lymph node dissection. The combination of these with chemotherapy is optional, and is reserved for patients with recurrent and metastatic disease. Local excision is now considered as preferred treatment type. Following surgical excision, clinical evaluation of the site of primary neoplasm and palpation of draining lymph nodes are recommended at 6-month intervals to detect recurrences and metastases (10).

In our patient, localization of the lesion and defined features were compatible with AFX; the rapid growth pattern and the size of lesion (3 cm) made us think that the tumour may be potentially malign and we diagnosed it as pleomorphic malign histiocytoma. In our case, because the size of lesion exceeded 2 cm, we experienced a dilemma in diagnosis. The accurate recognition and differential diagnosis of AFX are important, because underdiagnosis may lead to inedequate treatment and overdiagnosis may lead overly agressive, potentially mutilating surgery (10). In spite of the aggresive histopathologic appearance of AFX, it is a borderline tumour. If any criteria which is known to be found in a metastasizing case were found in a tumor to give a diagnosis as AFX may be inadequate, because of its being known as benign course despite the use of various treatment modalities. We thought our case as pleomorphic malign fibrous histiocytoma rather than AFX and followed-up the patient after complete surgical excision. Six month later, recurrence was not observed. The follow-up of this case may show us the importance of size in assigning size criteria in the diagnosis of AFX.

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