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Atypical Fibroxanthoma of the Foot

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Atypical fibroxanthoma (AFX) of skin usually occurs in the elderly sun exposed head and neck area as a nodular lesion. The author presents a case of a AFX developed on young female patient's foot with bleeding crust and itching sensation.

The histopathologic finding and various immunohistochemical studies were in good agreement with the diagnosis of AFX. The author also discusses differential diagnosis from other histologically similar neoplasms utilizing immunohistochemistry.

Key Word: Atypical fibroxanthoma, Foot

Atypical fibroxanthoma(AFX) usually occurs as a small nodular or ulcerated nodule in the elderly patient predominantly on sun exposed head and neck area. Occasionally, it may be mistaken for a carcinoma or a pyogenic granuloma. Inspite of histological anaplastic appearance with multiplicity of cell types, marked cytological atypia, bizarre giant cells and abundant mitotic activity, the atypical fibroxanthoma behaves usually as a benign neoplasm.

Distant metastasis of AFX cases have rarely been reported, particulary in younger patient with histologic factors of vascular invasion, recurrence, deep tissue invasion, tumor necrosis and possibly defective or depressed host resistance[1].

Histologically, it is indistinguishable from pleomorphic form of malignant fibrous histiocytoma, except AFX is situated more superficially than malignant fibrous histiocytoma.

Thus, AFX is regarded as a superficial form of malignant fibrous histiocytoma that usually carries more favorable prognosis. This author presents a case of atypical fibroxanthoma of foot and discusses immunohistochemical studies.

CASE REPORT

A 34 year old female patient with biopsy report to be atypical fibroxanthoma of foot was presented to me for further surgical treatment. The initial tumor size was not obvious, but the patient had a 1.5×1.5 cm previously biopsied open wound on the middle of dorsal surface of her left foot. Patient stated that about 2 month before this biopsy she noted an area of itiching which easily bled and was covered with crust, and gradually increased in size. According to the pathologist, the initial biopsy specimens were three pieces of skin ranging from 1.3 to 0.8 cm. Surgical specimen was diagnosed as "malignant spindle and epitheloid cell tumor consistent with atypical fibroxanthoma (Superficial cutaneous malignant fibrous histiocytoma), tumor cells extended to all margins of the biopsy." Immunohistochemical marker study revealed that the tumor cells stain strongly positive for Vimentin and moderately positive for CD68 (KP1), negative for Cytokeratin, S100 protein and Desmin. Patient underwent wide exicision of AFX of foot with 0.5 cm margin. The intraoperative frozen section failed to show any residual tumor, but a permanant section showed residual tumor very close to the deep resection margin and vascular invasion in one vessel. Additional 3 cm margin and deeper reexicision down to the paratenon on the dorsum of foot was performed. Final pathological report showed that the resection margin was free from tumor. The histopathologic slide showed ill defined tumor present within the superficial and deep dermis of the skin. The tumor was composed of spindled and epithelioid malignant cells arranged in a fascicular and storiform pattern. Nuclei were large, oval to spindle-shaped, with prominent central nucleoli. Mitotic

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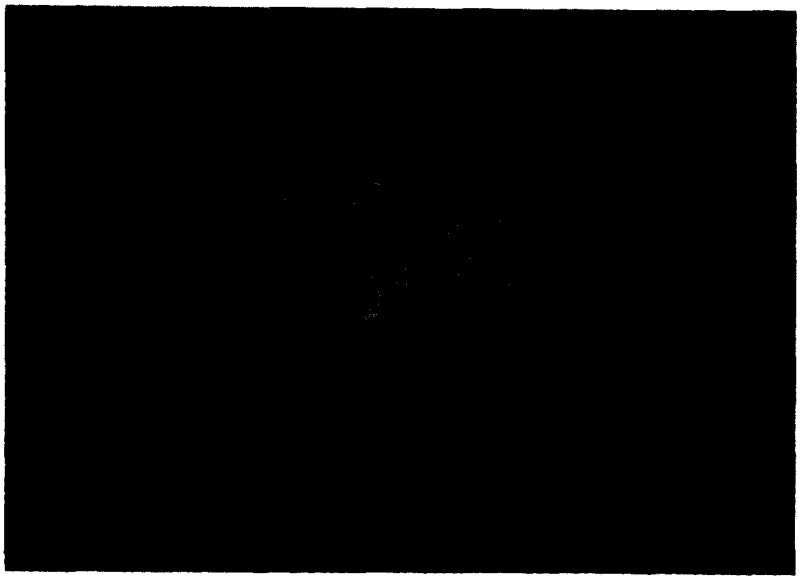

Fig. 1. Photomicrograph showing characteristic pleomorphic polyhedral cells arranged in a fascicular and storiform pattern (H & E stain, ×100).

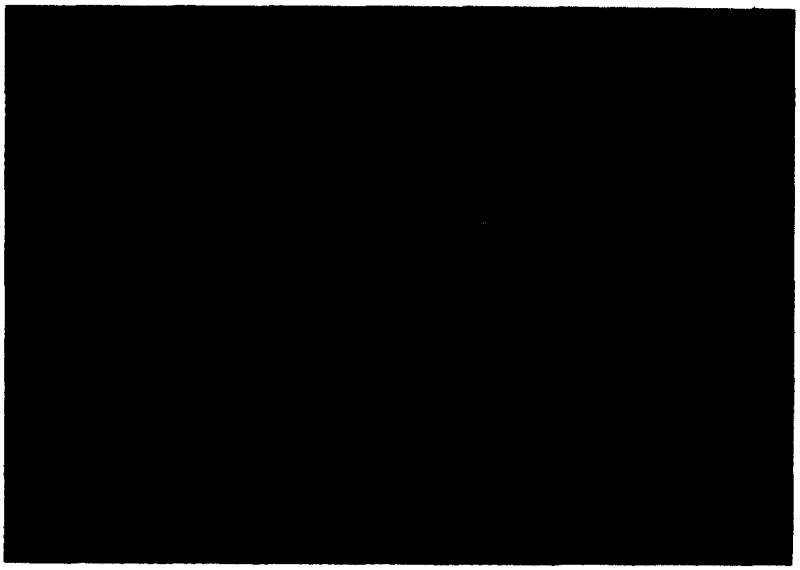

Fig. 2. Area of multiple focal necrosis in the middle of tumor (H & E stain, ×40).

figures are seen in up to 5 of 10 hpf. Vascular invasion was seen in one vessel in the middle of the tumor. The patient underwent screening test of brain computerized tommography (CT), chest Xray and abdominal CT scan including liver which were all reported to be negative for possible metastasis. She did not have clinically palpable popliteal nor femoral lymphadenopathy. The widely resected open wound was reconstructed with skin graft.. One week after the surgery, the patient developed right sided facial palsy which rushed us to do facial CT and repeated brain CT which again turned out to be negative indicating this patient developed simple Bell's palsy probably of viral origin. Presently, she is about 3 years after the wide excision of AFX and is free of any clinical evidence of recurrence or distant metastasis.

DISCUSSION

Atypical fibroxanthoma is a term first used by Helwig to describe a tumor of the fibrous histiocytoma group which appears cytologically malignant but clinically[2] behave in a benign manner.

The most common clinical presentation is a firm solitary

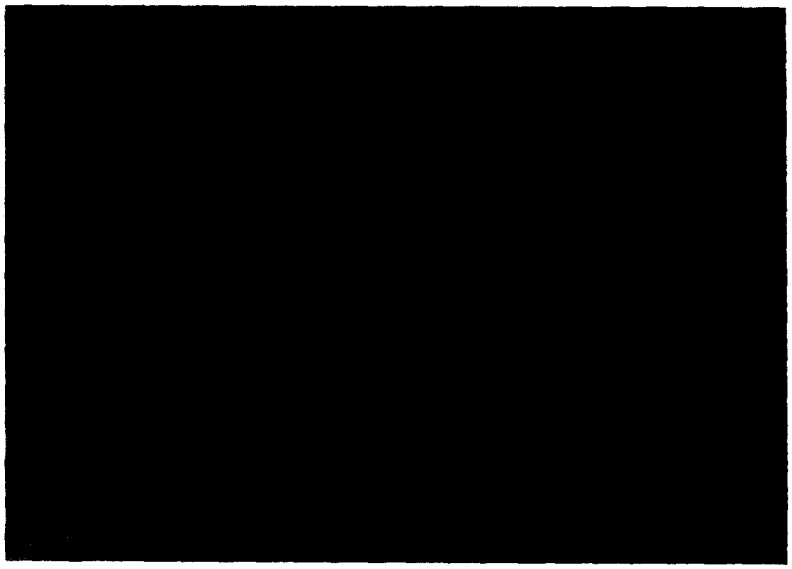

Fig. 3. Photomicrograph showing vascular invasion of tumor (H & E stain, ×100).

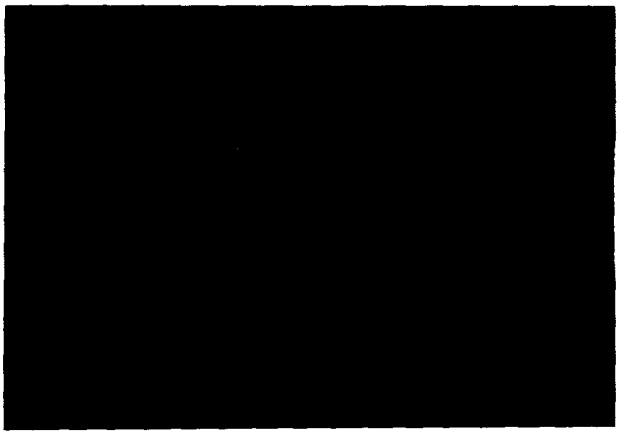

Fig. 4. After initial excision of atypical fibroxanthoma of foot at local dermatologic clinic.

nodule or ulcerated nodule usually less than 2 cm in dimension for the most on elderly sun exposed head and neck area. In the present case, the initial tumor in young female patient presented as a bleeding crust on the dorsum of foot with itiching sensation. Fretzin[3] reported that only three out of 140 cases of AFX developed on the foot indicating rarity of specific anatomic site for AFX.

This tumor (AFX) should be differentiated from malignant fibrous histiocytoma (MFH), amelanotic spindle cell malignant melanoma, spindle cell squamous carcinoma and sometimes from other mesenchymal neoplasm such as leiomyosarcoma and undifferentiated angiosarcoma[4]. Malignant fibrous histiocytoma may resemble AFX histologically and ultrastructurally. However, malignant fibrous histiocytoma occurs principally in the extremities or retroperitoneum of younger people, in contrast to AFX which has predilection for the skin of the head and neck area of elderly. Furthermore, it is a soft tissue neoplasm originating in the facial or subcutaneous tissue and, if the dermis is involved, it does so secondarily[1]. Clinically the duration of AFX is rather short, varying from weeks to months, while MFH has a much longer clinical course (months to years).

Amelanotic spindle cell malignant melanoma shows cells that are strongly reactive for S-100 protein by immunohistochemical methods. The tumor cells of AFX is negative for S-100 protein[5]. Spindle cell squamous carcinoma can be distinguished from AFX by positive reactivity of tumor cells to antikeratin[6] and by the presence of tonofilaments in its cytoplasm by electronmicroscopy. Dermatofibrosarcoma protuberance can be distinguished from AFX by its uniform spindle-shaped fibroblast, often arranged in a cartwheel or storiform manner, and by the paucity of mitotic figures and absence of pleomorghic giant cells[7].

A leiomyosarcoma primarily in the skin is rare and the extremities are the most common site. Differentiated myomatous areas may be distinguished by the presence of bundles of spindle-shaped cells with elongated blunted nuclei and intracellular myofibrils, and also with positive reactivity to desmin in immunohistochemical staining.

Immunohistochemical study of the presently described tumor showed strong positive reaction for Vimentin and

moderately positive for CD68 (KP1) and negative for antibodies to Cytokeratin, being consistent with fibrohistiocytic origin. Thus, the histologic findings and results of immunohistochemical study were consistent with the diagnosis of AFX. After review of eight examples of metastasizing AFX, Helwig suggested factors that portend aggressive behavior and metastasis. They are vascular invasion, recurrence, deep tissue invasion, tumor necrosis and possibly defective or depressed host resistance[1]. Since this tumor recurred in a young woman and showed aggressive nature of vascular invasion and focal area of necrosis, it is imperative that the present case should be followed carefully although she is free of disease for 3 years after the surgical treatment.

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REFERENCES

1. Helwig EB and May D: Atypical fibroxanthoma of the skin with metastasis. Cancer 57: 368~376, 1986
2. Helwig EB: Atypical fibroxanthoma. Tex. med. 59: 644~667, 1963
3. Fretzin DF, Helwig EB. Atypical fibroxanthoma of the skin. A clinicopathologic study of 140 cases. Cancer 31: 1541~1552, 1973
4. Enzinger FM and Weiss SW: Malignant fibrohitiocytic tumors in soft tissue tumors St. Louis CV Mosby, 1983 pp166~198
5. Nadjem MA: Case of diagnosis. Multiple atypical fibroxanthoma. Mil. Med 151(12): 666, 668~9, 1986
6. Kuwano H, Hashimoto H and Enjoji M: Atypical fibroxanthoma distinguishable from spindle cell carcinoma in sarcoma-like skin lesion. Cancer 55: 172~180, 1985
7. Taylor HB and Helwig EB: Dermatofibrosarcoma protuberans, a study of 115 cases. Cancer 15: 717~725, 1962
8. Gabbiani G, Kapani Y, Barazzone P and et al. Immunochemical identification of intermediate-sized filaments in human neoplastic cells: A diagnostic aid for surgical pathologist. Am J pathol. 104: 206~216, 1981