

# DERMATOPATHOLOGY UNIT

## UPP | Department of Dermatology

### UPMC Dermatopathology "Case of the Month" Presentations

#### UPP - Department of Dermatology, Dermatopathology Unit

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**Case Authors: Peggy Lin MD, Leena T. Lourduraj MD, Drazen M. Jukic MD PhD**

### MAY 2005 CASE OF THE MONTH

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#### CLINICAL FINDINGS

#### CLINICAL HISTORY

An outside dermatologist sent a skin specimen to the UPMC Department of Dermatopathology for a second opinion/consultation. The specimen was removed from the left arm of a 19-year-old female and a request was made to evaluate for a possible smooth muscle tumor of uncertain malignant potential. No clinical history was submitted with the case, which was submitted as a consult slide and consult block.

#### HISTOPATHOLOGY

H&E sections reveal a plaque-like proliferation of plump spindle cells arranged in a plexiform formation (Figure 1). Groups of cells intersect between collagen bundles and envelope individual adnexal structures (Figure 2). Some of the cells have a foamy cytoplasm. A good Grenz zone between this neoplasm and the epidermis is appreciated (Figure 3). The neoplasm extends focally into the subcutis, in a somewhat pushing fashion. Mitotic figures are observed, approaching 1-2/10 HPF (Figure 4)

Immunohistochemistry studies demonstrate that the tumor cells are positive for vimentin, smooth muscle actin, focally with CD68, and there is increased proliferation rate seen with interpretation of Ki67, that focally reaches up to 15%. Other stains performed were myoglobin, myosin heavy chain, S100, CD56- NCAM, CD57, desmin, Factor XIIIa, calponin, smooth muscle myosin heavy chain, CD34, CD31, and tyrosinase were all negative. (Figures 5 and 6)

The histological features and special studies are consistent with atypical myofibroblastic tumor due to the positivity for smooth muscle actin and negativity for desmin. This is a neoplasm of either fibrohistiocytic or myofibroblastic lineage (atypical plexiform histiocytic tumors could somewhat stain positive with SMA).

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## DISCUSSION & DIAGNOSIS

### Diagnosis

Atypical plexiform fibrohistiocytic tumor

### Clinicopathologic

**Epidemiology:** Most patients have been reported to be between 2 months to 71 years of age.<sup>1,4</sup> Females being affected more than men (3:1),<sup>1,5,6</sup> though some report equal frequency between females and males.<sup>4</sup> Lesions of atypical plexiform fibrohistiocytic tumors usually occur on the upper extremities (63.1%)<sup>1,3,4</sup> of patients younger than 20 years. <sup>1,3,4</sup> Median age in one study was 14.5 years. <sup>1</sup> There is no reported racial predilection.<sup>1,3</sup>

Lesions of the upper extremity were reported as elbow, forearm, wrist, hand, and shoulder. <sup>1,5</sup> Other locations of the body of lower frequency were the head and neck and the back and chest wall. <sup>1,5</sup> The chin was the affected location in one 71 year old patient <sup>1</sup> and one 10 year old patient. <sup>6</sup> Most lesions are solitary, though multiple lesions have been associated in a case of recurrence. <sup>1</sup>

The neoplasm typically presents as a solitary, firm, rubbery, and deep-seated nodule. (1) Overlying skin is not discolored or ulcerated, but may be slightly raised or show a slight central depression. (1,4) Growth of the tumor is most often slow, though both growth spurts of the

lesion or minimal to no growth have been reported. (1) A history of preceding trauma is reported in only a few cases. (1) There is often no pain nor tenderness associated with the lesions.(1,2)

**Prognosis:** Biologic behavior is unpredictable(5) with recurrence rates are reported as 23%,2 37.5%, (1,2) and 40%, (4) usually in the first 2 years of diagnosis. 1,4 This tumor has also been reported to metastasize to the lymph nodes in a few cases (3-6.2%). 1,2 Recurrence has been reported to occur between 3 to 24 months 4 and up to 36 months in one study.( 1) The tumor has even been reported to occur twice in one case(.4) Recurrence was observed to occur prior to metastasis. Metastasis is not reported to go to the lung or other organs.( 1) Because of the possibility of recurrence and metastasis, it is important to discern these tumors from benign fibrohistiocytic tumors, such as dermatofibromas and fibrous xanthomas.( 6)

**Histology:** Tumors are mostly located in the lower dermis(1) at the dermal-subcutaneous junction4, although one case of recurrence occurred in the superficial dermis(.4) Neoplasms of this type have also been reported to extend into the subcutis or muscle. (1) On section, the lesion is reported to have a gray-white to gray-tan color and the surface having irregular, dense, gray-white trabeculae.(1) Reported lesions ranged in size from 0.3 to 6 cm. (1,2)

The neoplasm is composed of nonencapsulated4 and poorly demarcated1 nodules with slightly lobulated and irregular borders. (1) Cellular composition varies1 and fascicles are composed of mononuclear and multinucleated plump spindle (4) cells with histiocytic (43%) (1,4) and fibroblastic (17%) (1) features. Some report cell types being round or oval cells with granular cytoplasm and a vesicular nucleus or as elongated spindle cells. (3) Cells arrange in a plexiform fashion, with some tumors reported to infiltrate into skeletal muscles. (1,2,4) Vascular invasion has been reported in recurrent lesions. (1)

Tumor fascicles are of various sizes, which anastomose randomly, and trap normal tissue in between them, giving the plexiform or reticulated appearance. (4) Histiocytic nodules have been reported to appear as confluent nodular aggregates(4) or as multifocal histiocytic nodules mixed with fibroblastic trabeculae. 4 Mild atypia and mitoses may be observed. (1,3) Multinucleated and osteoclast-like giant cells have been reported by some. (1) Other features that may be seen are microhemorrhage,(1) dense fibrosis with hyalinization at the periphery,1 hemosiderin deposition,(1,2) and a chronic inflammatory infiltrate.(1) The tumor mostly affects the mid to deep dermis and the upper subcutis, and generally does not affect the upper dermis, though it has been reported.(1) Overlying epidermis is usually not affected, but may display acanthosis.(1) Extension to muscular tissue may occur.(1) Vascular invasion has been described.(4) Tumor necrosis is generally not present.(1) Recurrent lesions usually show the same pattern as the originally excised tumor.(1)

Immunohistochemistry reveals myofibroblastic differentiation, expressing positivity for smooth muscle actin 1,2 and vimentin1 and negativity for S-100 protein, (1,2,4) desmin, (1,2,4) cytokeratin,1 or Factor VIII-related protein.1 Positivity for CD 68 is also seen.(2-4) Expression of a-1-antitrypsin and a-1-antichymotrypsin is seen in one-third of cases, which supports

fibrohistiocytic lineage.(1)Factor XIIIa positivity was not reported in all cases.(4)

**Differential diagnosis:** Includes benign fibrous histiocytoma (1,3,) plexiform neuroma, (1) fibromatosis, (1) benign and malignant giant cell tumor, 1 and fibrous hamartoma.(1) Benign fibrous histiocytoma has a more cohesive and uniform appearance, rather than the characteristic plexiform and multinodular pattern and is usually located on the lower extremities. 4 Myofibroblastic differentiation is suggested with positivity for smooth muscle actin.(3( Negativity for factor XIIIa suggests that this tumor is not of dermal dendrocytic lineage. (3) Fibrous histiocytoma and giant cell tumor usually affects adults and exhibit solid, rather than plexiform or nestlike pattern.(1) Fibrous histiocytoma also shows a more storiform pattern and may exhibit xanthoma cells and siderophages.(1) Fibrous hamartoma usually occurs in the first 2 years of life and does not contain multinucleated giant cells.(1) It has dense fibroblastic trabeculae and mature fat with a cellular myxoid component.1 Plexiform neurofibroma demonstrates positivity for S-100 protein.1 Giant cell form of malignant fibrous histiocytoma and giant cell tumor of tendon sheath usually shows cellular pleomorphism and giant cells with abnormal nuclei and in most cases, affects older patients.1

**Treatment:** Biologic behavior is not correlated with any specific clinical or histologic finding.( 1) Since recurrence is unpredictable, but reported to be as high as 40%,(4) the treatment for atypical plexiform fibrohistiocytic tumors is surgical excision (wide excision preferable to local), with frequent interval monitoring for local recurrence and for metastasis to the lymph nodes.(1-6) X-ray irradiation and has also been reported as an adjunct to surgical therapy in recurrent cases.4 Chemotherapy has also been reported to be used after radical lymph node dissection in one 5 year old girl, with no evidence of pulmonary metastasis 10 months later. (1)

## References

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