Atypical Fibroxanthoma Metastasis: a Mystery for One Thousand and One Nights

Nima Amini, MD; Maxwell A. Fung, MD

Departments of Pathology & Laboratory Medicine and Dermatology, University of California Davis, Sacramento, CA

Introduction

Atypical fibroxanthoma (AFX) is a spindle cell tumor classified as a low-grade sarcoma associated with chronic sun exposure particularly in the head and neck area. The term “AFX” was first used by Helwig in 1961 for a skin tumor that shows high grade of nuclear pleomorphism but with a great prognosis and only exceptionally rare metastases.[3] AFX is a diagnosis of exclusion. Metastases occur in less than 1 percent of AFX cases and less than 40 examples of metastatic AFX have been published. The most common location for metastases is lymph node, followed by visceral organs and parotid glands.[2,4]

Case History

A 75-year-old man presented with a 2 year history of a lesion on the left temple. A shave excision was performed on a 1.1 cm well-demarcated, ulcerated, brown pigmented plaque. Microscopically, sections showed a nodular dermal proliferation of atypical spindle cells without infiltrative growth or subcutaneous involvement (Figure 1). The tumor cells were negative for S100, Melan A, AE1/AE3, Desmin and CD34. The tumor was strongly immunopositive for CD10 (Figure 3) with focal expression of smooth muscle actin (SMA), consistent with AFX. Two and a half years later, he presented with a left neck mass. Tumor cells from a fine needle aspiration biopsy showed an identical morphology and immunophenotype as the skin tumor (in addition, 34BE12, MNF116 and p63 were all negative), consistent with metastatic AFX. (Figures 4,5,6). In comparison, the metastasis appeared to be of a higher grade. (Figures 2,4)

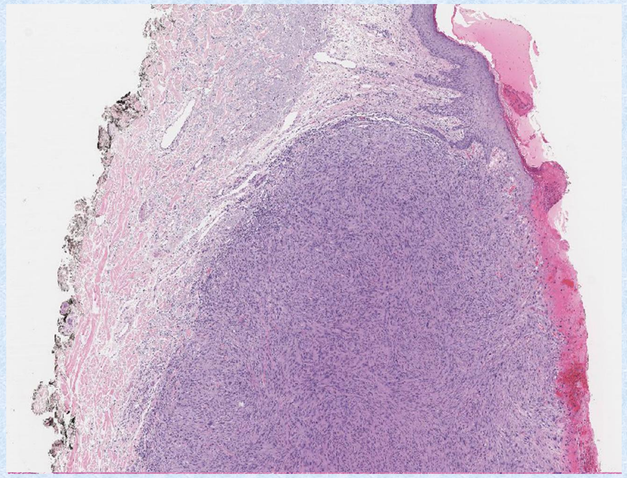

Figure 1. The well-demarcated, ulcerated primary lesion on H&E stain (20X)

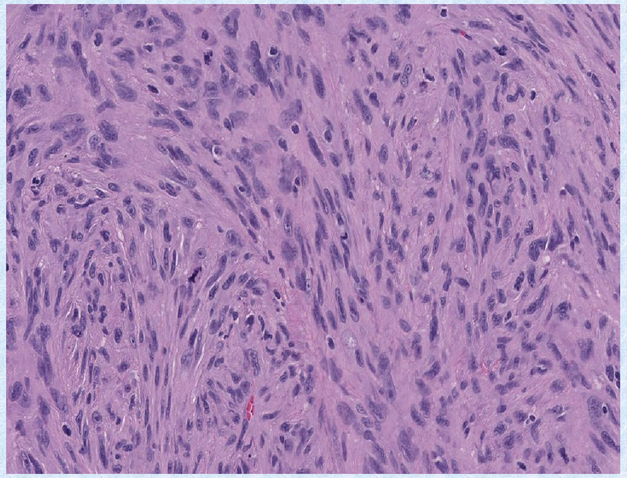

Figure 2. Bizarre multinucleated tumor cells in hypercellular stroma with frequent mitotic figures,on H&E stain (200X)

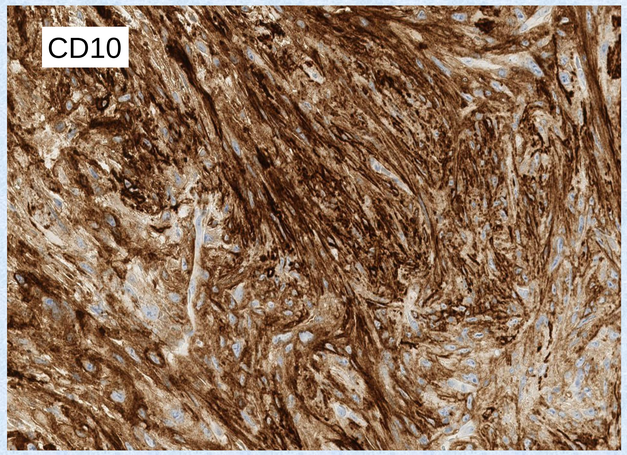

Figure 3. Primary lesion, tumor cells are strongly positive for CD10 (200X)

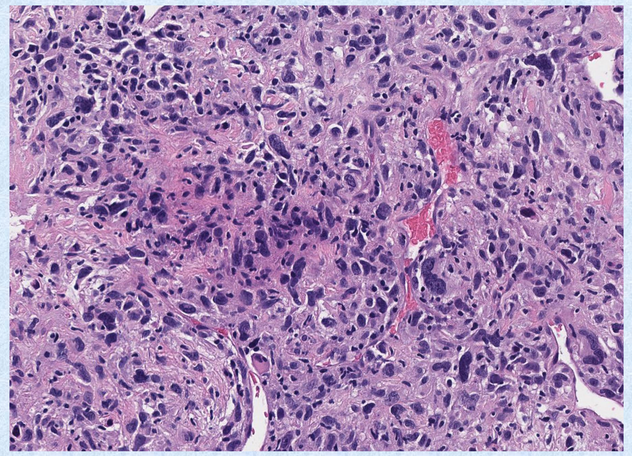

Figure 4. Identical histologic features seen in the secondary lesion on H&E stain (200X)

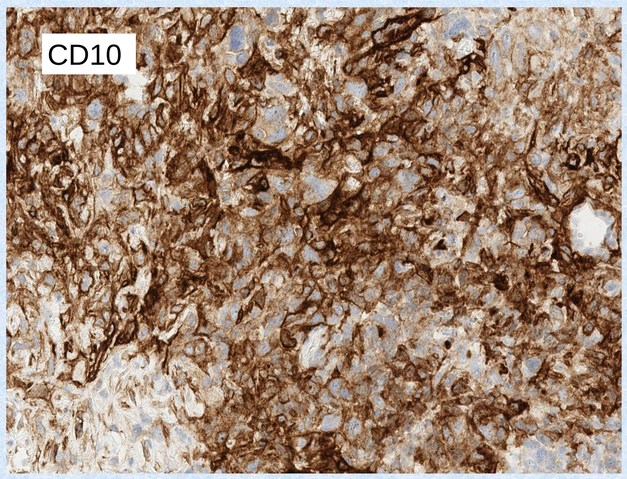

Figure 5. Secondary lesion, tumor cells are strongly positive for CD10 (200X)

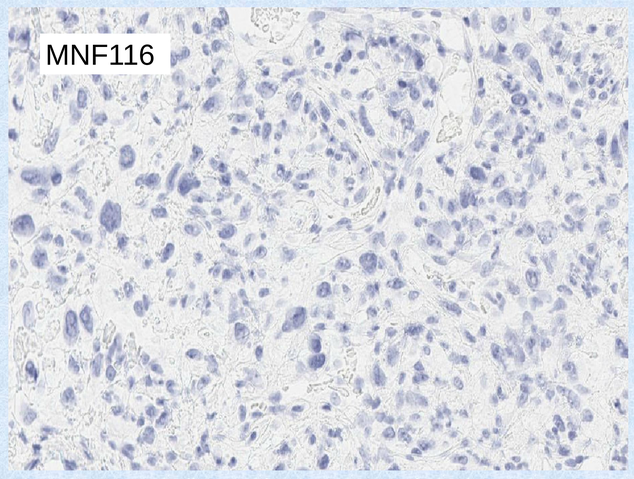

Figure 6. Tumor cells negative for MNF116 (200X)

Discussion

Since metastatic AFX is so rare, the main challenge is to determine whether a new tumor represents metastatic AFX versus another primary tumor or metastasis. Moreover, AFX is a diagnosis of exclusion (akin to undifferentiated pleomorphic sarcoma). Vascular invasion, tumor necrosis and deep tissue invasion are known factors contributing to increased risk of AFX metastasis.[1] None of these factors were present in of our case. As for immunohistochemistry, there is no AFX-specific immunostain.[1] In addition to identical morphology, the neck mass was strongly positive for CD10 and vimentin and negative for S100, Melan A, multiple cytokeratins (AE1/AE3, 34BE12, MNF116), p63, and CD34. Thus, our case is a genuine AFX metastasis and highlights a rare but important potential behavior of AFX. In conclusion, although AFX is not considered to be an aggressive form of sarcoma, metastasis can occur rarely. Immunohistochemical stains play a critically important role in distinguishing AFX and metastatic lesions and differentiating these tumors from other spindle cell neoplasms such as melanoma, spindle cell squamous cell carcinoma, poorly differentiated angiosarcoma, leiomyosarcoma, and, rarely, dermatofibrosarcoma protuberans, among others. The diagnosis of AFX must be based on a composite assessment of the histological features, immunophenotype, and clinical setting. It is important to report rare metastatic cases of AFX as a reminder of the small but significant potential for aggressive behavior.

References

1. Lum, D J. Peritoneal metastases from an atypical fibroxanthoma. The American Journal of Surgical Pathology. 2006 August;30(8): 1041-1046
2. Satter KE. Metastatic atypical fibroxanthoma. Dermatology Online Journal. 2012 18 (9): 3
3. Helwig EB. Atypical fibroxanthoma. In: Seminar Proceedings of 18th Annual Seminar of San Antonio Society of Pathologists, 1961. Tex State J Med. 1963; 59: 664-667.
4. Helwig EB, May D. Atypical fibroxanthoma of the skin with metastasis. Cancer. 1986 Jan 15;57(2):368-76.

Contact info: nimamini@ucdavis.edu