Atypical Fibroxanthoma Invading the Parietal Bone

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History & Clinical Course

Eighty-six year-old male with multiple medical co-morbidities, presented from an outside dermatologist with biopsy-proven atypical fibroxanthoma (AFX) of the left parietal scalp. The lesion was treated with Mohs micrographic surgery and the tumor was cleared after two stages. The final defect measured 4.0 x 3.5 cm, and it was left to heal by secondary intention (Figure 3).

Wound check six months after Mohs micrographic surgery revealed a new firm nodule. Biopsy was consistent with a hypertrophic scar with scattered atypical epithelioid cells, present on the margins, concerning for AFX. He returned for a second Mohs micrographic surgery, and the surgery was stopped after two stages given AFX extension into bone. The defect at this point measured 5.5 x 5.0 cm. The patient was sent to ENT for evaluation. The patient underwent radical resection of the outer cortex of the scalp and the defect was repaired with a perstring closure, as well as a full-thickness skin graft from the left supraclavicular fossa. The patient followed-up with ENT one month after surgery, with a 3.0 x 4.0 cm new growth at the surgical site. Repeat biopsy was consistent with AFX recurrence. The preliminary plan is resection and radiation therapy.

Physical Examination

Initial clinical visit revealed a well-appearing Caucasian male with an approximately 2.5 x 2.5 cm ulcerated red nodule and surrounding indurated erythematous plaque of the left parietal scalp.

Radiology

CT of the head without contrast at initial ENT consultation revealed a subacute or chronic left subdural hematoma with mild midline shift but no mass effect. Follow-up CT of the head without contrast approximately one month later, after discovery of second recurrence, showed diffuse volume loss and periventricular white matter changes consistent with age-related atrophy and no evidence of hemorrhage.

Histology

Histology

[Images of histological slides labeled 2A-2D and 4X]

Clinic Images

Clinical Images

[Images of clinical images labeled 3A-3B]

Figure 1. Punch biopsy from left parietal scalp showed an ulcerated proliferation of atypical spindled cells in a fascicular arrangement extending from the epidermis into the underlying dermis, with scattered atypical mitoses.

Figure 2A-D. Immunohistochemical stains as noted. Stains not pictured: CD31, Sox10.

Discussion

AFX is an uncommon fibrohistiocytic tumor, regarded as a less aggressive, superficial variant of undifferentiated pleomorphic sarcoma (UPS). Clinically, AFX presents as a pink/red firm, asymptomatic papule/nodule with ulceration, typically less than two centimeters in diameter on the sun-exposed areas, especially the head and neck of older males. AFX of the extremities is usually larger, slower-growing, and with less well-defined borders. The pathogenesis of AFX remains largely unknown with contributing etiologies including ultraviolet radiation, p53 mutations, cyclobutane pyrimidine dimers, prior radiation therapy, and immunodeficiency. There is considerable controversy regarding the differentiation between UPS and AFX. Some authors suggest that tumors larger than two centimeters in diameter, involving the fascia or muscle, exhibiting necrosis, vascular invasion, or metastasis should be regarded as UPS, given its more aggressive nature.

Immunohistochemistry plays an important role in differentiating AFX from its histological differential, including spindle cell squamous cell carcinoma (SCC), desmoplastic melanoma, and UPS. Cytokeratin markers and S-100 are negative in AFX but positive in spindle cell SCC and desmoplastic melanoma, respectively. CD10 is a cell-surface glycoprotein that is strongly positive in AFX and much weaker and focal in both desmoplastic melanoma and spindle cell SCC. (Figures 2A-D) LN-2 is a promising marker in delineating the difference between AFX and UPS. Studies show that 90% of UPSs stain strongly for it and 90% of AFXs have weak or no staining.

Treatment with Mohs micrographic surgery has a lower recurrence rate than excision. Radiation may be used as an adjunctive treatment for recurrent or metastatic disease. Induration, ulceration, or poor wound healing predict recurrence, which typically occurs within one to three years of treatment. Depending on modality of treatment, recurrence has been reported in up to 16% of cases, and it often foreshadows metastasis. Metastasis to lymph nodes, liver, lung, and subcutaneous tissue has been described in up to four percent of patients. While AFX has been noted in unusual sites such as the ethmoid sinus, eyelid, and cornea, this is the first case of parietal bone involvement to our knowledge.

References
