Excellent Outcomes of Atypical Fibroxanthoma Warrant Specific Diagnosis

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BACKGROUND:
Atypical fibroxanthomas (AFX) are rare malignant cutaneous neoplasms typically found on the head or neck of elderly Caucasian men. Limited clinicopathologic and outcomes data on this cancer exists. We report the clinical, pathologic and treatment characteristics, as well as oncologic outcomes in this dataset which represents the largest, single-institution retrospective analysis within the past decade.

METHODS:
Clinical, pathologic, treatment, and outcome data were compiled for all patients with AFX diagnosed, evaluated, and treated at a single institution between 2000–2020. Descriptive statistics were used to evaluate clinical and pathologic characteristics. Kaplan-Meier method and Cox proportional-hazards models were completed to assess overall survival and recurrence free survival.

RESULTS:
87 patients with AFX were identified. Median age was 75 years (range 38-89 years). The majority were men (n=67, 78%), immunocompetent (n=79, 91%), and Caucasian (n=85, 98%). 84% (73/87) were located on the head and neck. Table 1 describes the descriptive clinicopathologic variables seen in this series.

38% (33/87) patients were diagnosed with AFX on initial biopsy. 62% (54/87) were diagnosed with AFX only after complete resection; the differential diagnosis in these cases included pleomorphic dermal sarcoma (PDS) in 32% (28/87), undetermined spindle cell malignancy in 15% (13/87), melanoma in 5% (4/87), malignant fibrous histiocytoma in 5% (4/87), and poorly differentiated squamous cell carcinoma in 5% (4/87). Patients treated with surgical resection (n=85, 98%) had a median surgical margin of 1.0 cm (range 0.2-3.0 cm). 2% (n=2) patients were treated primarily with definitive radiation, and 7% (n=6) patients underwent adjuvant radiation: 5 for positive margins and 1 for perineural invasion, seen on final pathological evaluation. Over a median follow up of 14 months (range 8 months-not estimable), 2.3% (n=2) of patients developed recurrence. One patient had local and regional recurrences 5 months after surgery for a lesion confined to the dermis. Recurrences were treated with surgery and adjuvant radiation. The other patient developed multiple local recurrences 99 months post-operatively. All 3 of this patients' recurrences were treated surgically. These patients were reclassified and treated as PDS based on behavior and ultimately died of other causes. No patients developed a distant recurrence or died of disease.

CONCLUSIONS:

In the majority (64%) of patients complete resection is required to yield a definitive diagnosis of AFX. Median resection margins of 1 cm achieve excellent local control with ~98% recurrence free survival. Immunohistochemical staining is frequently needed to exclude other higher-risk tumor types, and thus is an integral component of the work up and diagnosis. In our series, only 2% of tumors diagnosed as AFX after complete excision subsequently demonstrated higher grade behavior compatible with pleomorphic dermal sarcoma.