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# Recurrent Angiomatoid Fibrous Histiocytoma: A Case Report and Review of the Literature

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## ABSTRACT

**Background:** Angiomatoid fibrous histiocytoma (AFH) is a rare soft-tissue tumor with intermediate malignant potential. It most commonly occurs in children and young adults.

**Case Report:** We report a case of a recurrent AFH of the back, locally recurring within 14 months of the original operation. We also review the literature for this uncommon entity. The patient underwent a wide resection of the recurrent AFH, obtaining negative surgical margins. Postoperatively, he has done well.

**Conclusion:** Twelve months since his re-excision, the patient shows no evidence of recurrence to date. He is seen every 6 months for continued clinical examinations.

## INTRODUCTION

Angiomatoid fibrous histiocytoma (AFH) is a rare soft-tissue tumor with intermediate malignant potential and a median age of presentation of 14 years. It accounts for approximately 0.3% of soft-tissue neoplasms.[1,2] The majority of cases occur in the extremities, are slow growing, and are typically painless.[3,4] We present the case of a young male with a local recurrence of an AFH.

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## CASE REPORT

A 21-year-old male presented to an outside hospital with a painless well-circumscribed mass located on his left upper back. An excisional biopsy was performed, and the final pathology revealed an AFH of intermediate malignancy with all surgical margins that were involved on the biopsy specimen. These findings were discussed with the patient and he was advised to undergo re-excision of this area to obtain negative surgical margins. However, the patient was lost to follow-up and re-excision was never performed. After 14 months, the patient presented to our institution with a return of the mass within the previous scar.

He stated that the mass had recently grown quite rapidly and was enlarging beyond its original size. Physical examination was significant for a firm, raised mass measuring $4 \times 4 \times 3$ cm along the patient's left upper back (Figure 1). No erythema, induration, or discharge was associated with the painless mass. The patient underwent wide local excision of the recurrent mass. We placed surgical clips at the periphery of the resulting defect in anticipation of the possible need for adjuvant radiation therapy to the area (Figure 2). The defect was closed primarily, with minimal tension along the midportion of the incision (Figure 3).

On pathologic examination, the lesion was 4.5 cm in its greatest dimension, and margins were negative for tumor invasion (Figure 4). Gross analysis of the specimen revealed an ovoid fragment of darkly pigmented skin weighing 79.5 g. The epidermal surface showed a raised nodule measuring 4.5 cm in diameter that extended above the surrounding epidermis by 2.5 cm. The cut surface revealed a bulging, deep red-brown, well-circumscribed mass. The histologic analysis revealed a well-demarcated mass without a capsule extending into the dermis, focally extending into the subcutaneous space. The overlying epidermis was flat with epithelial ridges disappearing.

We identified a few accessory, but hyperplastic, blood vessels present within the superficial dermis.

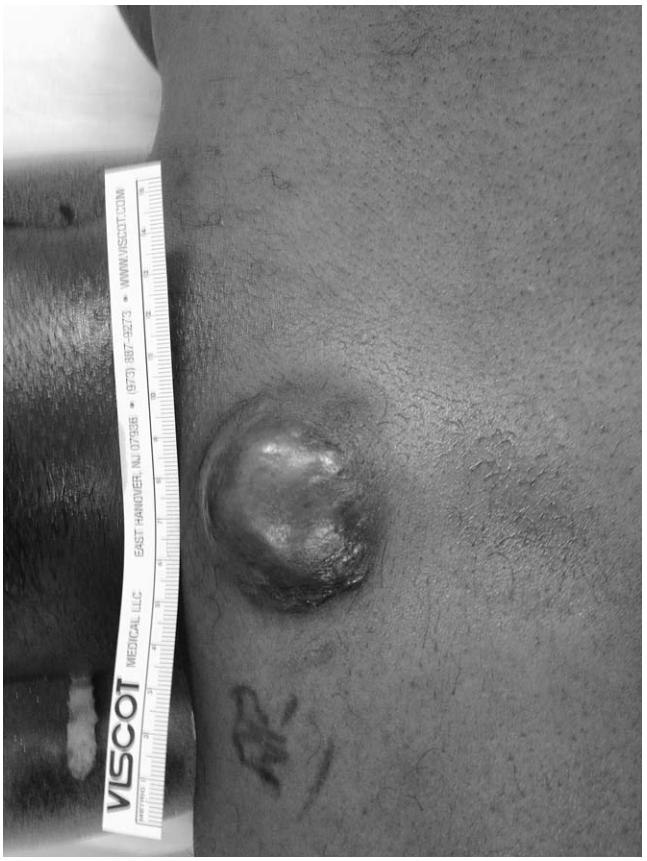


**Figure 1. Clinical examination revealing firm raised mass along left upper back.**

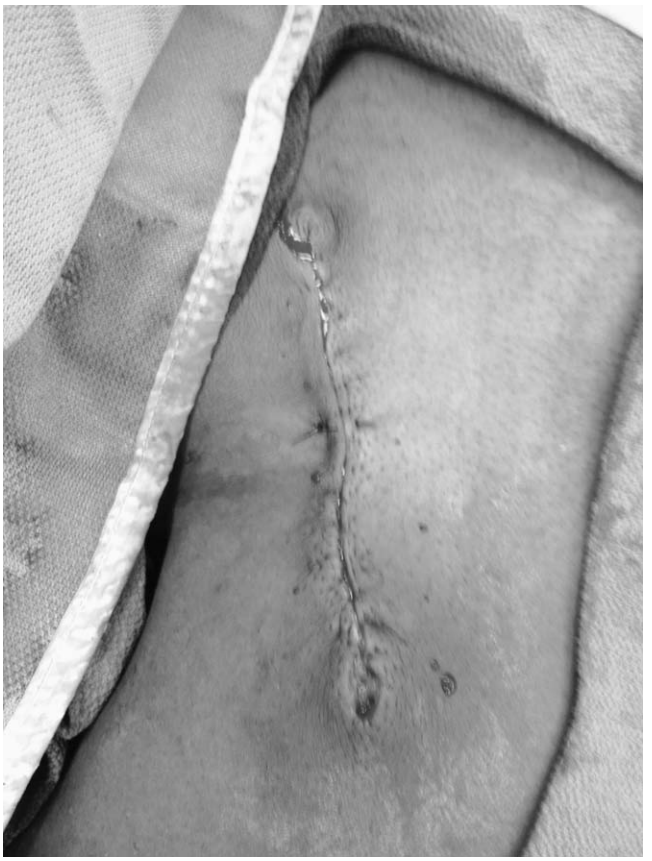

**Figure 3. Intraoperative view of primary closure after mass excision.**

On low-power magnification, dilated and congested irregular-shaped blood vessels within the tumor mass were visible, which were lined not by endothelial cells but rather by flattened tumor cells (Figure 5). Multiple foci of large hemorrhage were present, resulting in cystic changes with hypocellular areas of hyalinization seen around the dilated vessels. In the hypercellular areas, the tumor cells showed typical features of fibrous histiocytoma, which are spindle to ovoid

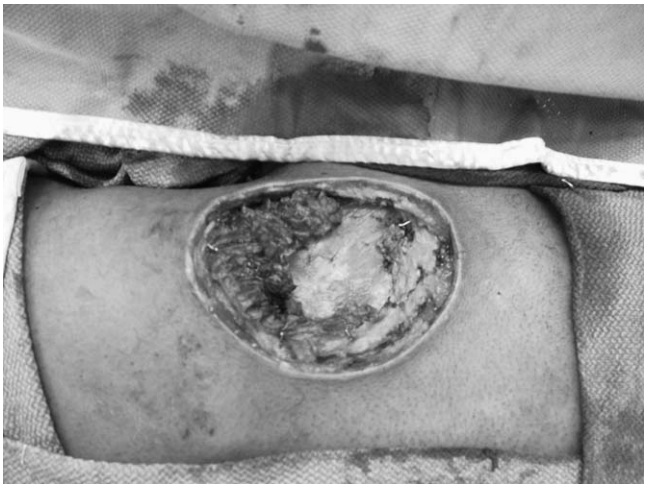

**Figure 2. Intraoperative view of wound base after mass excision with surgical clips placed for possible adjuvant radiation therapy.**

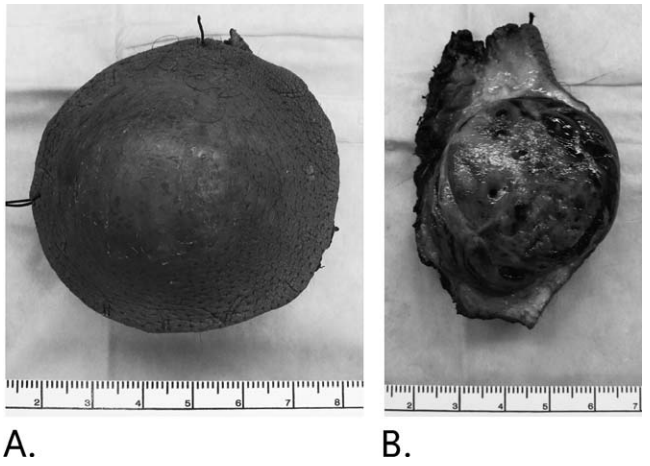


**Figure 4. A: Gross specimen of excised mass. B: Cut surface of gross specimen revealing a bulging, deep red-brown, well-circumscribed mass.**

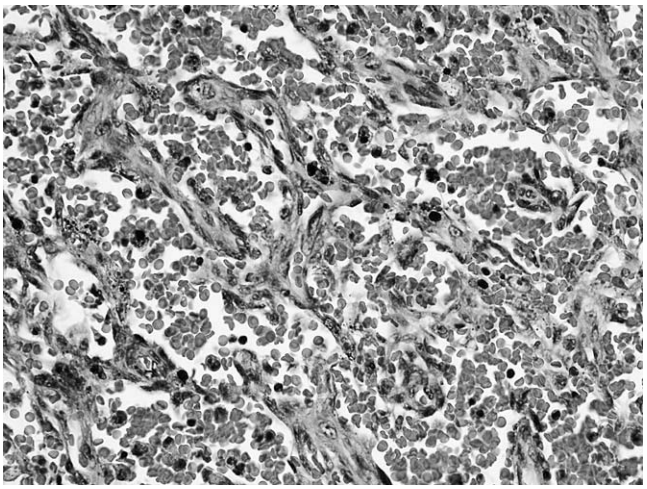

**Figure 5. Hematoxylin and eosin stain demonstrating ovoid to spindle and epithelioid atypic cells with vesicular nuclei as well as microscopic hemorrhage.**

shaped with mild atypia and arranged in a whorled pattern. Scattering histiocytes containing plenty of hemosiderin were identified, with up to 15 mitoses identified per 10 high-power fields. No necrosis was present, and final histologic grade was 1.

## DISCUSSION

Enzinger first described malignant AFH in 1979 with a series of 41 cases.[5] AFH was initially described as a variant of malignant fibrous histiocytoma; however, a large case review that demonstrated its favorable prognosis ultimately led to its recognition as a distinct entity.[6] The preponderance is slightly higher in females than in males, and systemic symptoms such as anemia, malaise, and fever have been reported, suggesting a possible connection with cytokine release.[5,7]

The World Health Organization formally removed AFH as a subtype of malignant sarcoma in 2002 and placed it in a category of tumors with uncertain differentiation.[3] Despite these developments, the exact line of differentiation for AFH remains unknown.[8] Immunohistochemistry demonstrates positivity for desmin, CD68, and CD99, whereas cytogenetic evaluation demonstrates that the EWSR1-CREB1 fusion gene is present in a majority of cases.[1,2,7,9]

In an informative series of 158 cases from 1979 to 1995, Fanburg-Smith and Miettinen suggest myoid cells of lymphoid tissue as a potential origin of AFH, given the findings of desmin-positive cells in the adjacent lymphoid infiltrate as well as the high percentage (66%) of their cases found in sites of normal lymphoid tissue.[10] Other translocations have also been described, but a correlation with tumor behavior and type of gene fusion has not yet been demonstrated.[11-18] Inconsistent and nonspecific findings have been described with imaging of the masses, either with x-ray or magnetic resonance imaging.[19]

Metaanalysis findings demonstrate that the majority of patients are disease free after local excision.[20,21] Fanburg-Smith and Miettinen reported a 1% frequency of metastasis in their series.[10] However, other studies have reported that up to 23% of patients may develop recurrence and 8.7% may develop metastasis within 24 months postexcision.[6,21] These factors contribute to the classification of AFH as having intermediate malignant potential.[21,22] Costa et al described successful management of local recurrence with radiation, which is an important consideration for our patient.[23] Local recurrence and metastasis have been found to correlate with invasion into the deep fascia or muscle.[6]

## CONCLUSION

We report a case of AFH that recurred within 14 months of its original excision. The prevention of a recurrence is essential for long-term survival. Consequently, the patient will continue to require long-term follow-up. Twelve months after re-excision of this recurrent tumor, he shows no evidence of tumor recurrence as of the writing of this article.

Teaching points for this rare tumor are to perform the appropriate surgical excision to obtain negative surgical margins and to continue close postoperative surveillance with clinical examinations to ensure no signs or clinical evidence of tumor recurrence exist.

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