

Aneurysmal Fibrous Histiocytoma, a Deceptively Pigmented Lesion of Skin: Study of Histopathologic Characteristics with Review of Literature

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DOI: <https://doi.org/10.36348/sjpm.2026.v11i08.004>

| Received: 12.08.2026 | Accepted: 03.10.2026 | Published: 13.10.2026

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Abstract

Background: Aneurysmal fibrous histiocytoma (AFH) is a rare variant of benign fibrous histiocytoma characterized by fibrohistiocytic spindle cells, blood-filled pseudovascular spaces, and hemosiderin deposition. Clinically, it may mimic vascular or melanocytic lesions, posing a diagnostic challenge. We describe the clinicopathological features of 18 cases of AFH and discuss its differential diagnosis. **Methods:** Archival records from a tertiary institution were searched for cases diagnosed between January 2009 and December 2021. Hematoxylin and eosin-stained sections and immunohistochemical preparations were independently reviewed by two soft tissue pathologists, and clinical, histopathological, and follow-up data were systematically analyzed. **Results:** The 18 patients had a mean age of 37.1 years (range, 14–64 years), with a slight male predominance M:F = 5:4 (10:8). Preoperative diagnoses were documented in 7 cases (38.9%), most commonly hemangioma (n=3). The back/scapular region was the most frequent site (33.3%), followed by the lower limb (22.2%). Grossly, lesions demonstrated a solid-cystic appearance with blood-filled spaces. Histologically, non-endothelialized blood-filled spaces were surrounded by bland spindle and histiocyte-like cells, most commonly arranged in a storiform pattern (55.6%). Mean hemosiderin deposition was 40.6%. Atypical/monster cells and Touton giant cells were each identified in 35.3% of cases, collagen trapping in 52.9%, and staghorn vessels in 23.5%. Resection was performed in 72.2% of cases. Among 12 patients with available follow-up (4–20 years), local recurrence occurred in 2 (16.7%). **Conclusions:** AFH can closely mimic vascular and melanocytic tumors. Recognition of its characteristic histopathological features, supported by targeted immunohistochemistry, is essential for accurate diagnosis and avoidance of overtreatment.

Keywords: Aneurysmal Fibrous Histiocytoma, Dermatofibroma, Benign Fibrous Histiocytoma, Pseudovascular Spaces, Hemosiderin Deposition, Cutaneous Soft Tissue Tumors, Immunohistochemistry, Local Recurrence.

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BACKGROUND

Aneurysmal Fibrous Histiocytoma (AFH) is a rare but distinct variant of benign fibrous histiocytoma (BFH)/dermatofibroma, accounting for approximately 1-2% of all cutaneous fibrous histiocytoma cases [1]. It predominantly affects middle-aged adults with a slight female predilection and a predilection for the lower extremities [2].

Clinically, extensive intralesional hemorrhage with hemosiderin deposition imparts a characteristic blue-black discoloration, mimicking hemangioma, melanocytic lesions, or cysts, while simultaneously driving rapid tumor enlargement, both features that heighten clinical concern for malignancy [2].

Histologically, AFH is characterized by blood-filled spaces devoid of an endothelial lining, surrounded by bland spindle cells, histiocyte-like cells, hemosiderin-laden macrophages, multinucleated giant cells, and a variable chronic inflammatory infiltrate [3]. Despite its benign cytology, AFH carries a local recurrence rate of up to 19%, far exceeding the less than 2% seen in conventional cutaneous fibrous histiocytoma, with isolated reports documenting repeated recurrences [2-4].

Its morphological complexity necessitates distinction from several overlapping entities, including angiosarcoma, angiomatoid fibrous histiocytoma (a neoplasm of intermediate malignant potential), and malignant melanoma [5,6]. Accurate histopathological

diagnosis is therefore essential to avoid misclassification and guide appropriate management.

Given the paucity of large-scale studies, this study aims to evaluate the clinicopathological characteristics of AFH and review the existing literature to further define its diagnostic criteria and clinical behavior.

MATERIALS AND METHODS

This descriptive observational study was conducted after obtaining approval from the institutional Ethical Review Committee. All patients diagnosed with AFH between 1st January 2009, and 31st December 2021 were included. Cases were identified electronically through the Integrated Laboratory Management System (ILMS). All cases of aneurysmal fibrous histiocytoma were selected using non-probability purposive sampling method.

Hematoxylin and eosin (H&E) stained slides, along with immunohistochemical preparations, were retrieved and independently reviewed by two pathologists with expertise in soft tissue pathology. Data were collected using a structured proforma, including patient demographics (age and gender) and tumor characteristics such as site, size, architectural patterns, cytological features, and immunohistochemical findings. Information regarding treatment (surgical management and adjuvant therapy) and follow-up outcomes, particularly recurrence, was also recorded.

Follow-up data were supplemented through telephonic communication while maintaining strict ethical standards and patient confidentiality. Data analysis was performed using Microsoft Excel (Version 16.85). Categorical variables, including gender, tumor site, and morphological patterns, were summarized using frequencies and percentages. Continuous variables, such as age and tumor size, were analyzed using measures of central tendency. The results were presented in tables and figures to highlight key findings.

RESULTS

A total of 18 patients with aneurysmal fibrous histiocytoma were included in the analysis. The mean age at presentation was 37.1 years (median 36 years; range 14–64 years). The cohort comprised 10 males and 8 females (M:F ratio 5:4).

Clinical impression was not documented in 11 of 18 patients (61.1%). Among the remaining 7, hemangioma was the most common preoperative diagnosis (n=3), followed by sebaceous cyst (n=2), pigmented nevus (n=1), and skin papilloma (n=1). Specimen size was available in 13 cases, with dimensions ranging from 2.0 to 13.9 cm (mean 5.3 cm; median 3.6 cm). On gross examination, the lesion had a solid-cystic appearance, with solid areas showing a tan white to variably yellow color and blood-filled cystic spaces imparting a dark red cut surface, as depicted in Figures 1a and 1b.

The most common anatomic site was the back/scapular region (n=6, 33.3%), followed by the lower limb (knee and thigh n=4, 22.2%), upper limb (arm and forearm n=3, 16.6%), and head/neck and chest region (scalp, behind the ear and chest n=3, 16.6%). Two cases had no documented site (11.1%). Of the total 4 PBCR cases (those that came for second opinion), 3 had diagnosis made, 1 was inconclusive and just reported as neoplastic. Of the remaining 3 the diagnoses were recorded as Aneurysmal Dermatofibroma, Dermatofibrosarcoma protuberans and Synovial sarcoma respectively.

All tumors demonstrated blood-filled spaces centered in the superficial tumor in most cases, but also in the center and deeper aspects in some (Figures 2a and 2b). These spaces lack an endothelial lining and are surrounded by bland spindle cells (Figure 3a), histiocyte-like cells which are arranged in storiform pattern (55.6%) (Figure 3b), followed by fascicular (22.2%) (Figure 3c) and mixed storiform-fascicular patterns (22.2%). The mean hemosiderin deposition was 40.6% (median 45%; range 5–80%) (Figure 3d), demonstrating considerable variability across cases. Atypia/monster cells were present in 35.3% of cases (Figure 4a) and absent in 64.7%. Touton giant cells were identified in 35.3% of cases, while 64.7% showed no such features (Figure 4b). Staghorn vessels were identified in 23.5% of cases, while 76.5% showed no such vascular pattern (Figure 4c). Collagen trapping was observed in 52.9% of cases (Figure 4d), while 47.1% showed no evidence of collagen trapping. Cholesterol clefts were observed in 22.2% of cases, while 77.8% showed no such features. Inflammatory infiltrate was absent in 29.4% of cases, minimal (rare/few) in 58.8%, and moderate in 11.8%, indicating generally low levels of inflammation across the cohort.

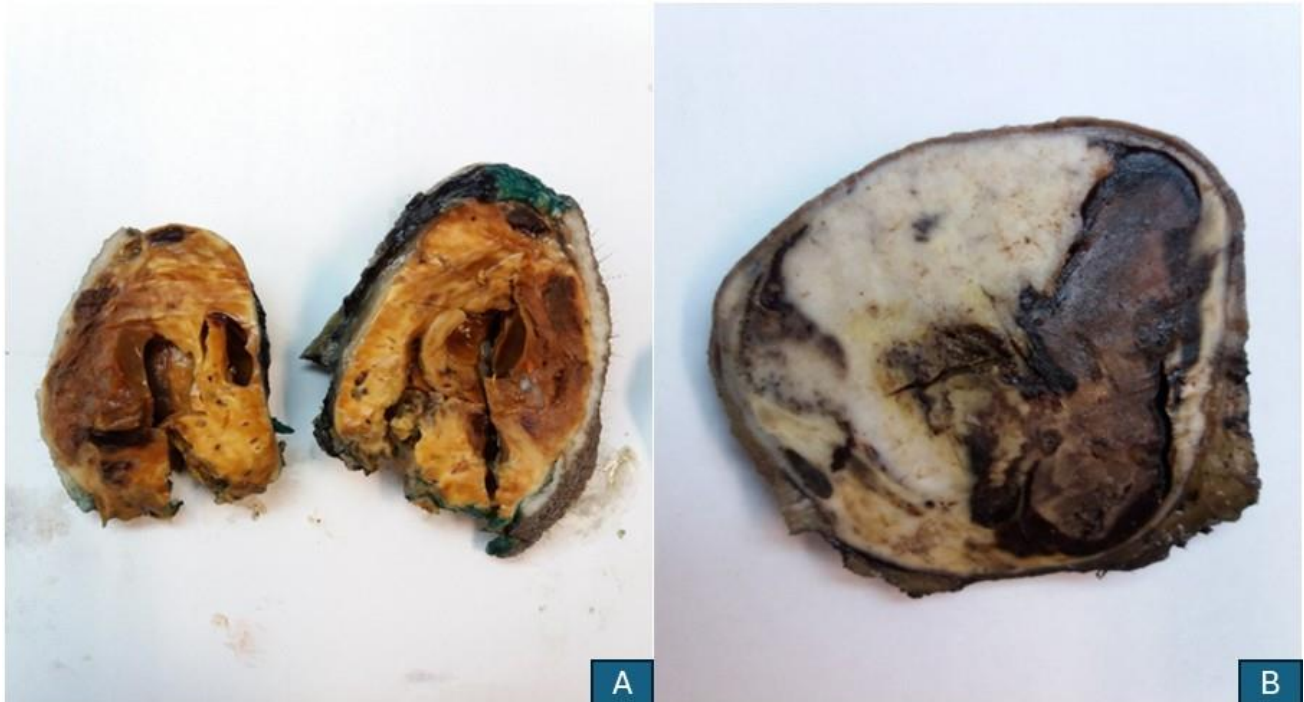


Figure 1: Figure 1a shows a solid cum cystic lesion with hemorrhagic areas and scattered vascular spaces. Figure 1b. Another gross example with a large hemorrhagic area surrounded by tan solid area

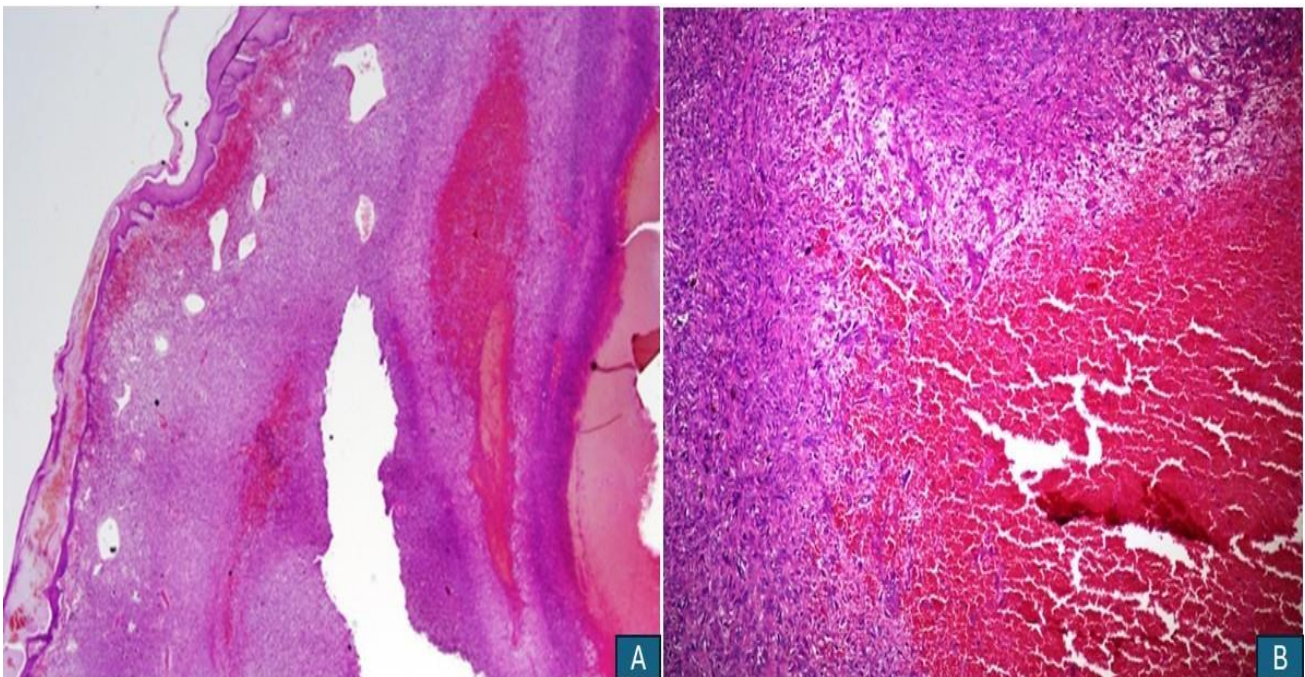


Figure 2: Figure 2a shows a solid-cystic lesion with areas of hemorrhage located in the dermis. Scattered thin-walled, ectatic vascular spaces are interspersed throughout, imparting a sinusoidal architecture, Figure 2b shows towards (left) solid component composed of sheets of tumor cells with moderate nuclear pleomorphism and indistinct cell borders. The cystic region (right) is characterized by large pools of extravasated erythrocytes

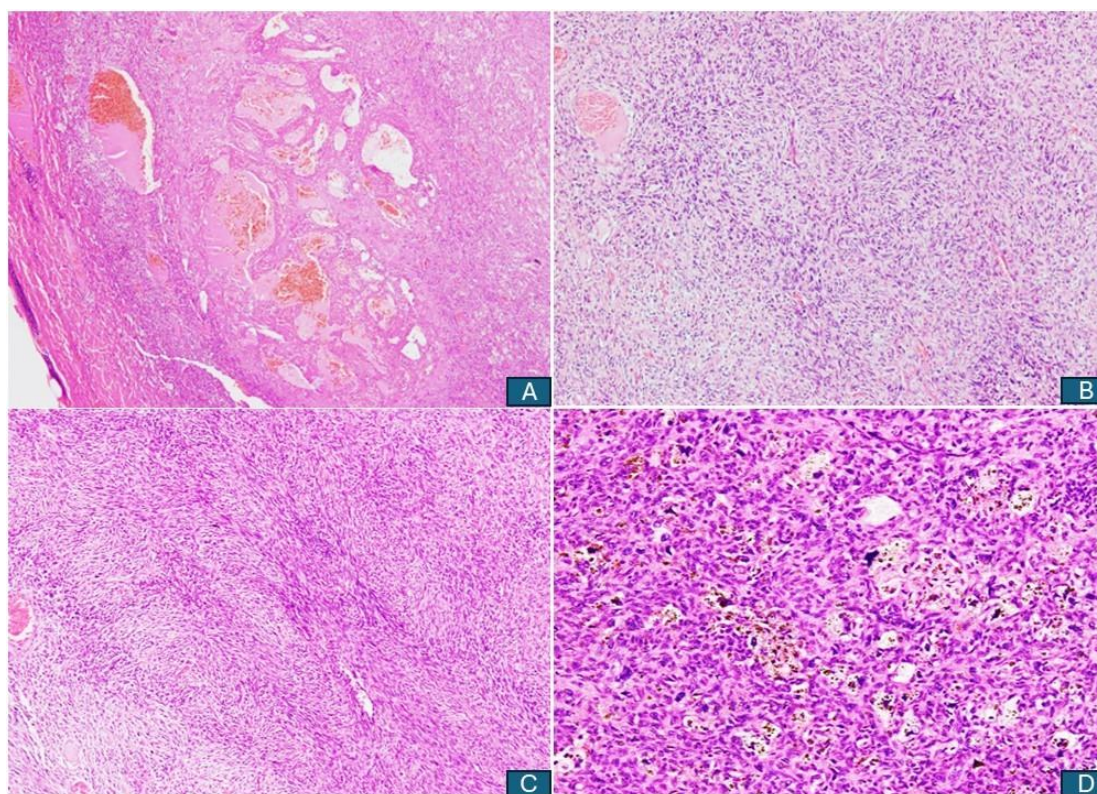


Figure 3: Figure 3a: Low power view showing spindle shaped cells admixed with blood filled spaces. Figure 3b: Low power view showing spindle shaped cells displaying storiform growth pattern. Figure 3c: Low power view showing fascicular arrangement of cells. Figure 3d: Lesion showing prominent hemosiderin deposition in the form of intracytoplasmic granules within histiocytes

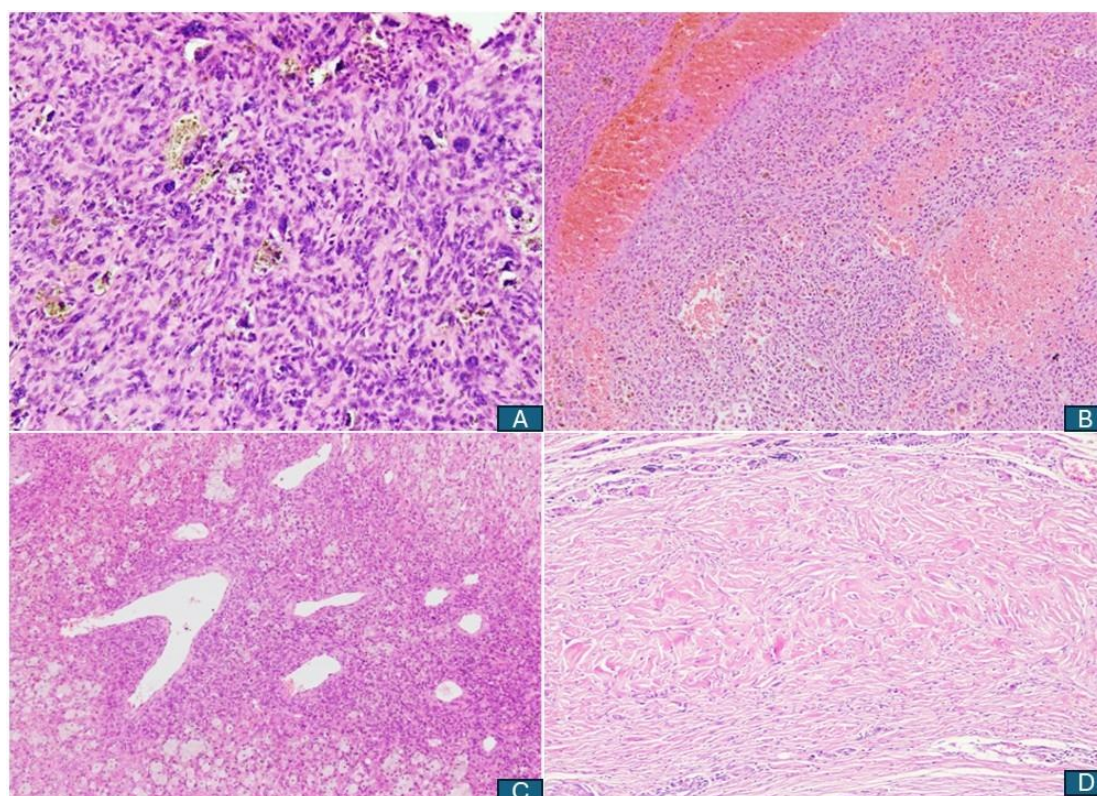


Figure 4: Figure 4a: showing scattered atypical/monster cells. Fig 4b: Lesion showing occasional touton type giant cells. Fig 4c: showing prominent staghorn vessels and aggregates of foamy histiocytes. Fig 4d: showing collagen trapping

Epidermal changes included normal epidermis in 7 cases (38.9%), hyperplastic changes in 7 cases (38.9%), absent epidermis in 3 cases (16.7%), and ulceration in 2 cases (11.1%); one case showed both hyperplastic and ulcerated epidermis and is counted in both categories.

Surgical resection was performed in 13 patients (72.2%), while 5 (27.8%) were lost to follow-up prior to treatment documentation. Follow-up data on recurrence were available in 12 of 18 patients, with a duration ranging from 4 to 20 years based on the earliest case identified in 2005. Local recurrence was observed in 2 of these 12 cases (16.7%); one patient experienced multiple recurrences, and another had a recurrence one year after initial resection. One patient died of lung carcinoma unrelated to AFH. Six patients (33.3%) were lost to follow-up and could not be assessed for recurrence.

DISCUSSION

Aneurysmal fibrous histiocytoma (AFH) is a rare variant of benign fibrous histiocytoma, and the existing literature is largely limited to isolated case reports and small series. It mimics melanocytic and vascular malignancies both clinically and histologically, making accurate recognition essential to avoid overtreatment. Aneurysmal (“angiomatoid”) fibrous histiocytoma was first recognized and described in 1981 by Santa Cruz and colleagues in a series of 17 cases where they described this lesion as a dermal based, larger, pigmented and fluctuating swelling mostly on the extremities, that may have acute onset of pain and rapid growth [9]. Clinically these lesions resemble melanomas or other melanocytic nevi, hemangiomas, neurofibromas or a non-specific cyst. Thus, these lesions were different both on clinical grounds as well as histologically from conventional cutaneous fibrous histiocytoma. This entity was initially synonymously called ‘angiomatoid’ fibrous histiocytoma [9]. However, evolution of cases led angiomatoid fibrous histiocytoma to be recognized as a completely different entity with a more variable histology and different biological behavior [10].

It is characterized by blood-filled pseudo vascular spaces, extensive hemosiderin deposition, and cystic degeneration, which often result in both clinical and histopathologic misdiagnosis as vascular or melanocytic lesions. In the present study, we analyzed 18 cases of AFH, representing one of the more recent single-institution series in the literature. Unlike the female predominance reported in the original series by Santa Cruz *et al.*, our cohort showed a slight male predominance. Earlier studies by Santa Cruz *et al.*, reported 14 female and 3 male patients ranging from 13 to 67 years of age (mean 46 years), with a predominance in the extremities [9]. Across published series, AFH has been reported over a wide age range from adolescence to advanced age (12–92 years), most commonly affecting young to middle-aged adults [2].

The back/scapular region was the most common anatomical site (n = 6), followed by the lower limb (n = 4). The upper limb and the head and neck region (scalp/ear) each accounted for three cases. This is consistent with both classical and contemporary studies, which demonstrate a predilection for the extremities and trunk, whereas head and neck involvement remains relatively uncommon [13]. Although uncommon, unusual locations have been described in isolated case reports, including the anal region, earlobe, and even primary intracranial involvement, highlighting the broad anatomic spectrum of this lesion [11, 12].

Histologically, all cases demonstrated characteristic pseudo vascular spaces lacking endothelial lining, filled with blood and associated with dense hemosiderin deposition. These findings are considered the defining feature of AFH and are critical in distinguishing it from true vascular neoplasms such as angiosarcoma. Within the spectrum of vascular variants of dermatofibroma, hemosiderotic and aneurysmal types are closely related. Zaballos *et al.*, described hemosiderotic dermatofibroma as containing numerous small vessels with extravasated erythrocytes and abundant hemosiderin deposition, whereas aneurysmal dermatofibroma exhibits larger blood-filled pseudovascular spaces without endothelial lining. Some authors have proposed that these entities may represent different stages along a common pathologic continuum.

The predominant storiform architecture observed in our series further supports AFH as part of the broader dermatofibroma spectrum. This concept is widely accepted in modern dermatopathology literature, where AFH is considered a morphological variant of benign fibrous histiocytoma rather than a distinct neoplasm [13, 14]. Additional features such as collagen trapping, Touton giant cells, and variable inflammatory infiltrate were also observed, consistent with previously reported histologic variability in fibrous histiocytoma variants [13].

A major diagnostic challenge in AFH is its close resemblance to vascular tumors, particularly angiosarcoma and epithelioid hemangioendothelioma, as well as melanocytic lesions due to heavy hemosiderin pigmentation [15]. Misdiagnosis is especially common in limited biopsy specimens where hemorrhagic pseudocystic spaces may be mistaken for true vascular channels. The absence of endothelial lining remains the most important histologic feature in excluding true vascular neoplasms [15].

Recent literature continues to emphasize the role of immunohistochemistry in difficult cases. AFH typically shows positivity for factor XIIIa and CD68, while being negative for endothelial markers (CD31, CD34), melanocytic markers (S100, Melan-A), and muscle markers, supporting fibrohistiocytic

differentiation [16]. A 2024 large retrospective study of dermatofibroma variants also highlighted that aneurysmal lesions often show increased cellularity and hemorrhagic change but retain benign immunoprofiles [13].

Although AFH is benign, recurrence has been reported more frequently than in conventional dermatofibroma, with rates up to approximately 20% in earlier series [4-15]. In our study, recurrence was observed in 2 of 12 cases with available follow-up (16.7%). However, this likely reflects incomplete excision and limited follow-up rather than aggressive biological behavior. A recent review also supports the concept that recurrence is primarily surgery-related rather than tumor-driven [16].

Rare cases of aggressive behavior and even metastasizing fibrous histiocytoma have been reported, but these are considered exceptional and not representative of typical AFH biology [16]. Overall, AFH remains a benign lesion with locally recurrent potential rather than malignant transformation risk.

Limitations

This study is limited by its retrospective single-institution design and small cohort size of 18 cases, which restricts generalizability and statistical power. A substantial proportion of patients (33.3%) were lost to follow-up, limiting accurate estimation of recurrence rates. Molecular or cytogenetic profiling was not performed, which would have strengthened diagnostic certainty in morphologically ambiguous cases.

CONCLUSION

AFH is a rare but diagnostically challenging variant of benign fibrous histiocytoma that closely mimics vascular and melanocytic lesions. In this series, lesions predominantly affected young- to middle-aged adults, with a predilection for the trunk and extremities. Recognition of its defining histopathologic features, pseudovascular spaces lacking endothelial lining, dense hemosiderin deposition, and storiform fibrohistiocytic proliferation, combined with a targeted immunohistochemical panel, is essential for accurate diagnosis and avoidance of overtreatment. Although benign, AFH demonstrates a propensity for local recurrence, emphasizing the importance of complete excision and appropriate clinical follow-up.

Ethical Approval: This study was approved by the Institutional Ethical Review Committee (ERC # 2021-5980-16764). As the research was conducted through retrospective review of archival pathology reports and previously prepared microscopy slides, a waiver of informed consent was granted. No identifiable patient information was collected or disclosed.

Consent to Participate: Not applicable because this article does not directly involve human participants and/or animals.

Author's Contributions: NU participated in the design of the study, retrieved data, supervised, and edited the manuscript. HF, SS, AS, and SM compiled data, did a literature search, and drafted the manuscript. All authors read and approved the final manuscript.

Finding: The authors received no financial support for the research, authorship, and/or publication of this article.

Declaration of Conflicting Interest: The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Triad Registration: Not applicable.

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