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CASE REPORT

Angiomatoid Fibrous Histiocytoma Mimicking Lymphadenitis: A Unique Case Study

Brijesh Thakur, Saqib Ahmed

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ABSTRACT

Abstract: Angiomatoid fibrous histiocytoma (AFH) is basically mesenchymal neoplasm with an incidence of 0.3%, only indicating its rarity amongst the soft tissue neoplasms. It commonly occurs first twenty years of life, though its occurrence has been documented across all age groups with no gender predilection. The tumor usually arises in the extremities, in the upper layer soft tissues in the head, neck and sometimes trunk regions. The present study deals with a unique case of a 58-year-old female who presented with a steadily increasing and uncomfortable lump in the posterior triangle of the neck. The initial clinical impression suggested cervical lymphadenitis. The unusual age and anatomical location make this case noteworthy. AFH should be considered in the differential diagnosis of persistent soft tissue lesions in all age groups, even though it is uncommon in older people.

KEYWORDS: *Angiomatoid Fibrous Histiocytoma, CD34, Ki67, Lymphadenitis.*

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INTRODUCTION

Angiomatoid Fibrous Histiocytoma (AFH), a member of soft and connective tissue lesion family is quite rare with just 0.3% occurrence rate among all soft tissue tumors.[1] As there has been no definite molecular or immunohistochemical evidence to confirm its classification. Due to this, World Health Organization (WHO) has categorized it as an intermediate soft tissue tumor of uncertain differentiation.[2] AFH typically is known to occur in the extremities, in the superficial regions with rarer occurrence in the head, neck and sometimes upeer trunk regions. Young adults below 20 years of age is a common age for its occurrence, though rarely it can be seen in elderly also.[3] Due to its woundring clinical, pathological and radiological characteristics, its exact and early diagnosis is made difficult. This article represents a case report of 58 years old female involving AFH at an atypical site and an older patient, who was mistakenly diagnosed with cervical lymphadenitis.

CASE REPORT

The case study belongs to a 58—year—old female and have a gradually enlarging swelling on the left side of her neck, persisting for approximately two years. The paint indicates that she has intermittent pain and local discomfort in this swelling area. Her past medical history was notable for pulmonary tuberculosis, for which she had completed a six-month course of antitubercular therapy. Clinically, the swelling was presumed to be cervical lymphadenitis, potentially tuberculous in etiology, given the absence of systemic symptoms and her prior history.

Department of Pathology, Shri Guru Ram Rai Institute of medical and health sciences, Dehradun.

Corresponding Author: Saqib Ahmed

Email: saqibahmed.mail@gmail.com

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An excisional biopsy was performed, and the resected tissue specimen was sent to pathology for histopathological examination to know the features of tissue. In histopathogy, the specimen measured approximately 2.2 × 1.2 × 1 cm in dimension and consisted of soft to firm, gray-brown tissue. On cut section, the lesion appeared predominantly solid with focal cystic areas; no definitive lymph node architecture was identified. Microscopic examination revealed an unencapsulated neoplastic proliferation composed of variably sized nodules containing predominantly spindle-shaped cells. These cells demonstrated oval to elongated nuclei, fine chromatin, and poorly defined pale eosinophilic cytoplasm. Spindle cells were occasionally arranged in fascicles and bundles, interspersed with lymphoplasmacytic inflammatory infiltrates and fibrous strands. Several blood vessels were seen within the nodular areas, and the intervening stroma displayed lymphoid aggregates and collagen bundles. Notably, increased mitotic activity was observed (Figures 1.1 to 1.4). No residual lymph node tissue was identified in the specimen. At the end of tissue histopathological features, a provisional diagnosis of angiomatoid fibrous

histiocytoma was made. To validate our results further, immunohistochemistry was also subsequently performed and to confirm the diagnosis and rule out all possibilities and causes including amelanotic melanoma, inflammatory myofibroblastic tumor, dermatofibrosarcoma protuberans (DFSP), and neural tumors. The tumor cells exhibited strong cytoplasmic positivity for desmin, vimentin, and S100 (Figure 2.1 to 2.3). Focal membranous staining for epithelial membrane antigen (EMA) was observed (Figure 2.4). The Ki-67 proliferation index was low (<2%) (Figure 2.5). The result Cleary shows that tumor cells were negative for all three main area smooth muscle actin protein which is associated with fibroblast (SMA), CD34, and Melan A, a final opinion of angiomatoid fibrous histiocytoma was validated.

Figure 1.1: Unencapsulated lesion composed of variably sized nodules of predominantly spindle-shaped (H and E, X4)

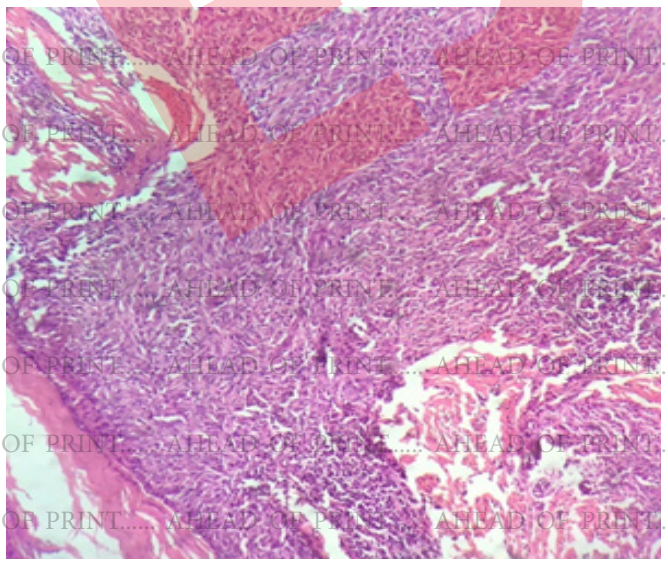

Figure 1.2: Variably sized nodules of spindle-shaped cells having oval to elongated nuclei, fine chromatin and moderate ill-defined pale eosinophilic cytoplasm (H and E, X10)

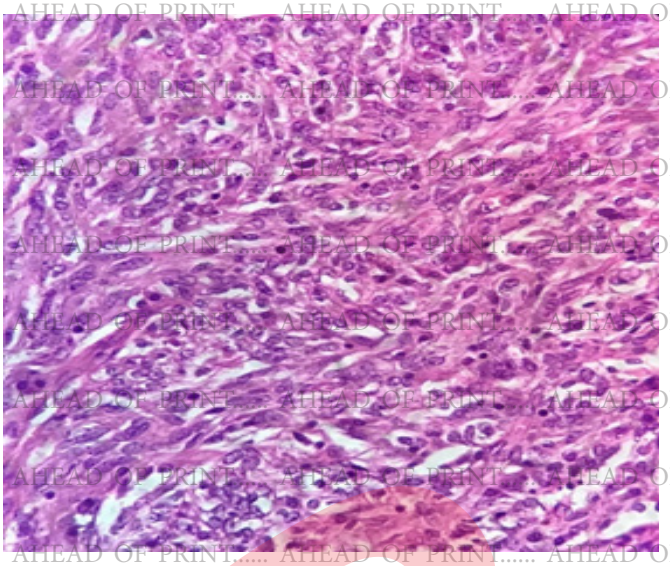

Figure 1.3: Spindle cells arranged in fascicles and bundles with intervening lymphoplasmacytic inflammatory infiltrates (H and E, X40)

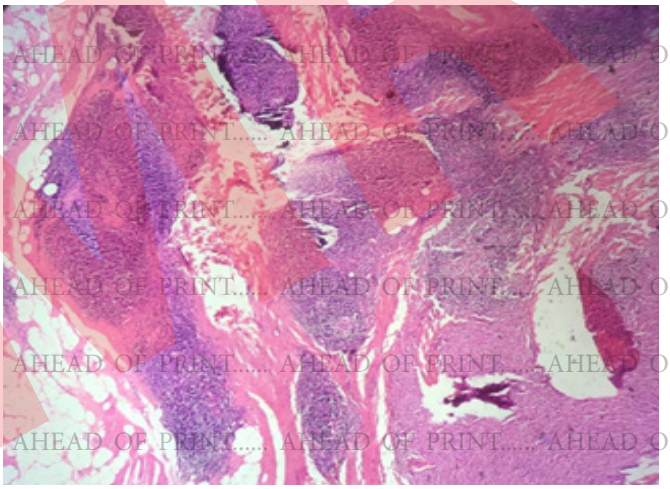

Figure 1.4: Internodular areas showing lymphoid aggregates and collagen bundles (H and E, X10)

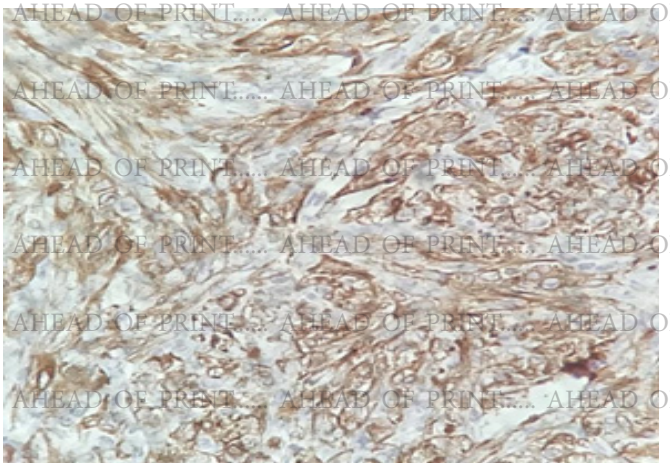

Figure 2.1: Immunocytochemistry with Desmin shows positive cytoplasmic staining in tumour cells (X40)

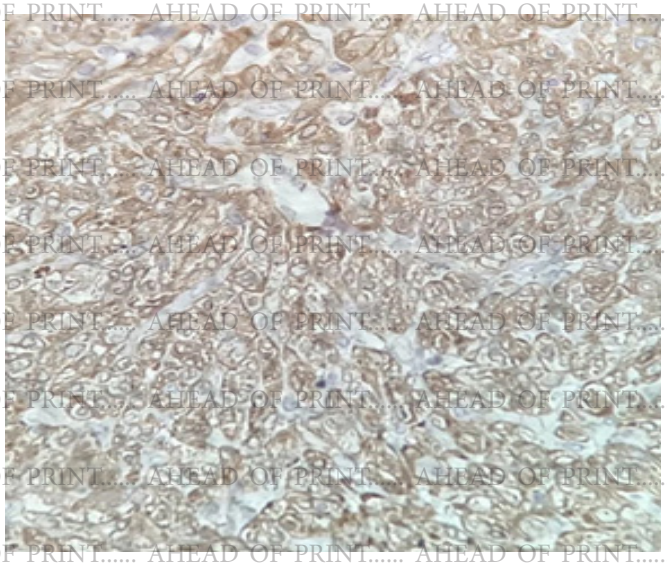

Figure 2.2: Immunocytochemistry with S100 shows both positive cytoplasmic and nuclear staining in tumour cells (X40)

Figure 2.3: Immunocytochemistry with Vimentin shows positive cytoplasmic staining in tumour cells (X40)

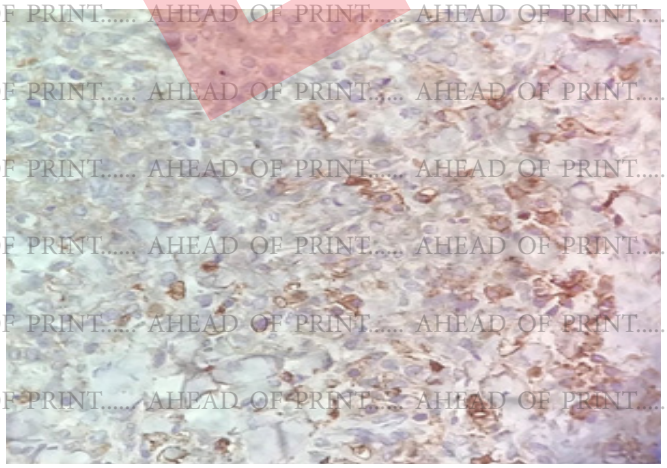

Figure 2.4: Immunocytochemistry with Epithelial Membrane Antigen (EMA) shows focal positive membranous staining in tumour cells (X40).

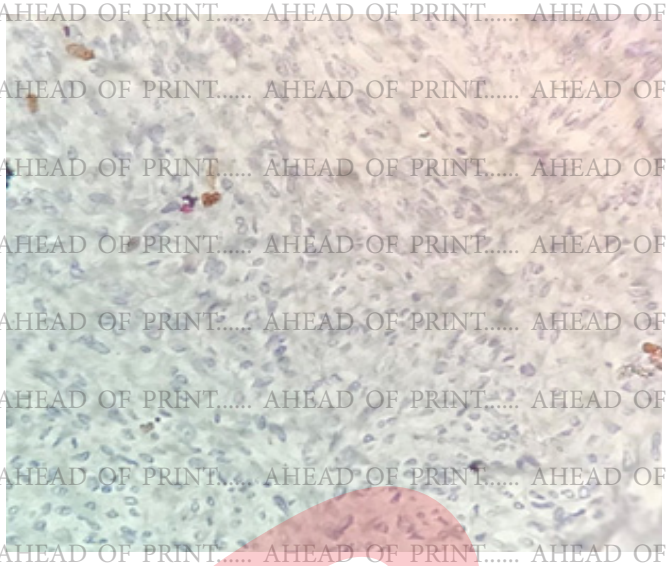

Figure 2.5: Immunocytochemistry with Ki67 showed proliferation index <2% in tumour cells (X40)

DISCUSSION

Angiomatoid fibrous histiocytoma (AFH) is the rarest soft tissue neoplasm and have uncertain origin.[4] It is basically presented as a superficial palpable mass which is painless but can also arise from the trunk, head and neck.[5] In this case, the patient reported pain, which is less common. AFH often arises in various areas in which the axillary, inguinal, and supraclavicular regions are common but has also been reported in some unusual body sites of people such as the brain, sometimes lungs and bone, and in some cases in breast area it was also found.[5] Although AFH has more predilection for young age but elderly age group is less common as in this case.[3] Sometimes it proceeds with a history of trauma, though lesser studies are present to establish the same. Associated symptoms, though less common include fever, fatigue, and signs like anemia, likely due to cytokine release by the tumor. Rare associations with paraneoplastic syndromes, including platelet function disorders, have also been noted.[6] As a result, the diagnosis of AFH becomes a more challenging and difficult task to understand its nonspecific clinical and radiological features. The differential diagnosis includes benign and malignant conditions such as hematoma, hemangioma, soft and connective tissue sarcoma, and metastatic lymph nodes.[7] Histopathologically, AFH has a pseudocapsular architecture, which is usually fibrous dilated and congested vascular channels are seen along with inflammatory infiltrate of some lymphocytes and plasma cells. Tumor cells show a syncytial pattern, which are usually uniform, with moderate anisokaryiosis and are spindled to epithelioid. Other minor histological features include stromal fibrosis and sclerosis, myxoid changes, microcystic changes, small blue round cell appearance, nuclear grooving, nuclear clearing and sometimes a hypocellular and hypercullar pattern like

schwannoma.[8,9] Genetically, most common rearrangement seen is involving the FET gene family, most commonly between EWSR1 and less often involving FUS. Fluorescence in situ hybridization (FISH) for EWSR1 rearrangement is a useful tool in confirming the diagnosis, particularly when histology is inconclusive.[10]

CONCLUSION

AFH is a moderate soft tissue intermediate-grade malignancy generally found in younger individuals. Its rarity, as well as the lack of distinguishing clinical and radiological characteristics, might lead to misinterpretation and confusion with other diseases. Accurate and early identification, good prognosis and diagnosis is very crucial. Consequently, AFH should be included in the differential diagnosis of soft and connective tissue swellings, and a comprehensive excision along with prolonged medical and radiological monitoring of the patient and sufferer is immensely recommended time to time.

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Orcid ID:
Brijesh Thakur - https://orcid.org/
Saqib Ahmed - https://orcid.org/