Imaging Characteristics of Angiomatoid Fibrous Histiocytoma: A Case Report and the Diagnostic Challenges in Differentiating Fluid–Fluid Level Soft Tissue Lesions

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AUTHORS' CONTRIBUTIONS
Equal contributions by all the authors have been made.

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DISCLOSURES
This clinical case is presented in two separate publications. The present manuscript concentrates on the radiological diagnostic features, whereas the other article authored by our colleagues addresses the clinical aspects and pathological findings. Together, these publications provide a comprehensive and multidisciplinary perspective on the case.

CONSENT
Yes

HUMAN AND ANIMAL RIGHTS
None

ABSTRACT

Angiomatoid fibrous histiocytoma is a rare mesenchymal tumor. Due to its non-specific clinical and radiologic features, angiomatoid fibrous histiocytoma is often misdiagnosed as other soft tissue lesions. We report the case of a 10-year-old boy who presented with a painless mass in the gluteal region. Initial ultrasound and Magnetic Resonance Imaging suggested a hemorrhagic lesion with fluid–fluid levels. Despite two inconclusive percutaneous biopsies, the patient underwent wide local excision, and histopathological examination confirmed the diagnosis of angiomatoid fibrous histiocytoma. This case highlights the diagnostic challenges of angiomatoid fibrous histiocytoma and underscores the importance of recognizing its imaging characteristics—particularly in distinguishing it from other soft tissue lesions that also exhibit fluid–fluid levels on Magnetic Resonance Imaging—in the appropriate clinical context.

CASE REPORT

BACKGROUND

Angiomatoid fibrous histiocytoma is a rare mesenchymal tumor that typically affects children and young adults. Although it is generally considered to have a low-grade clinical course, Angiomatoid fibrous histiocytoma poses considerable diagnostic challenges due to its nonspecific clinical presentation and overlapping radiologic features with other soft tissue lesions. One notable imaging characteristic is the presence of fluid–fluid levels on Magnetic Resonance Imaging, which is a finding shared by various cystic or hemorrhagic soft tissue tumors, both benign and malignant. Accurate diagnosis often depends on a combination of imaging findings, histopathological examination, and immunohistochemical analysis.

In this case report, we present a 10-year-old boy with a painless gluteal mass initially diagnosed by imaging as a hemorrhagic lesion based on ultrasound and a soft tissue aneurysmal bone cyst on Magnetic Resonance Imaging. Despite two ultrasound-guided biopsies yielding inconclusive results, the patient underwent wide local excision, and histopathological examination confirmed the diagnosis of

angiomatoid fibrous histiocytoma. This case highlights the diagnostic challenges associated with angiomatoid fibrous histiocytoma and underscores the importance of recognizing its imaging characteristics to differentiate it from other soft tissue tumors with similar appearances.

CASE REPORT

A 10-year-old-boy presented with a painless mass in the gluteal region of one month's duration. Constitutional symptoms such as fever or weight loss were absent. Physical examination revealed a large, immobile mass measuring approximately 15x15cm in the left gluteal region, non-tender to palpation and without spontaneous pain. The overlying skin was normal, without erythema (Figure 1). The patient was able to walk freely but reported occasional discomfort. Routine laboratory investigations, including complete blood count, alkaline phosphatase, lactate dehydrogenase, C-reactive protein and erythrocyte sedimentation rate were within normal limits.

The pelvic X-ray revealed a soft-tissue density mass in the left gluteal region, measuring 12x8cm, without bone erosion or visible calcifications (Figure 2).

Ultrasound of the left gluteal soft tissue revealed a large, multiloculated mass containing heterogeneous hyperechoic fluid-debris levels consistent with hemorrhagic content, measuring 10x6 cm. The mass was firm, without a change in shape upon probe compression. Peripheral vascular proliferation of the lesion was observed on color Doppler imaging. The lesion was initially diagnosed as a hematoma on ultrasound (Figure 3).

A contrast-enhanced Magnetic Resonance Imaging (MRI) study revealed a well-defined, multiloculated soft-tissue mass located in the left gluteal region, within the adipose tissue between the gluteus maximus and the gluteus medius/minimus muscles, measuring 10x6x10cm. The lesion exhibited heterogeneous signal characteristics across the imaging sequences, with locules containing fluid–fluid levels showing heterogeneous hyperintensity on T2WI and T2-STIR sequences, suggestive of hemorrhage. In addition, a nodule within a large cystic locule demonstrated hyperintensity on T1WI and hypointensity on both T2WI and T2-STIR sequences, compatible with a blood clot. The peripheral region of the mass was surrounded by a pseudocapsule that appeared markedly hypointense on both T1- and T2-weighted images, demonstrating enhancement after intravenous gadolinium administration. Minimal linear hyperintensity tracking along the adjacent peritumoral fascial planes was evident on fat-suppressed T2-weighted images, consistent with edema, without involvement of the adjacent muscular structures. The MRI was initially interpreted as consistent with a giant cell tumor (GCT). However, upon retrospective review, the imaging findings were also suggestive of an extraosseous aneurysmal bone cyst (ABC) (Figure 4).

Several other lesions that also demonstrate fluid–fluid levels or hemorrhagic components on imaging should be considered in the differential diagnosis, including soft tissue hematoma, soft tissue giant cell tumor, osteosarcoma, hemangioma, and fat necrosis. This overlap in imaging features makes accurate diagnosis challenging without histopathological confirmation.

Ultrasound-guided percutaneous biopsy was performed twice but did not yield conclusive results due to insufficient samples (Figure 5).

After wide surgical resection (Figure 6), histological examination of the surgical specimen confirmed the diagnosis of Angiomatoid fibrous histiocytoma (AFH) (Figure 7).

DISCUSSION

AFH is a rare mesenchymal soft-tissue neoplasm, accounting for approximately 0.3% of all soft tissue tumors, with uncertain histogenesis. First identified by Enzinger in 1979, AFH was initially considered one of five variants of malignant fibrous histiocytoma, and was referred to as angiomatoid malignant fibrous histiocytoma. Nonetheless, according to the WHO Classification of Soft Tissue Tumors 2020, AFH is categorized as an "intermediate tumor of uncertain differentiation", because of its slow-growing, benign microscopic appearance and favorable prognosis, with a local recurrence rate of 15–20% and a distant metastasis rate of 1–3% (Table 1) [1–3].

AFH predominantly affects pediatric and young adult populations, with a median age of 13 years. It most commonly arises in the extremities, though cases have also been documented in the trunk and head and neck regions. More recently, AFH has been identified in atypical anatomical locations, such as the lungs, mediastinum, vulva, retroperitoneum, ovaries, pulmonary artery, kidneys, and intracranial compartments. While AFH usually presents as a superficial mass involving the subcutaneous tissue or deep dermis, approximately 18% of cases demonstrate infiltration into deeper structures, including skeletal muscle and the periosteum [4–6].

The clinical presentation of AFH is often vague and nonspecific, most commonly appearing as a slow-growing, painless mass. In rare cases, patients may exhibit systemic symptoms resembling paraneoplastic syndromes, such as low-grade fever, anemia, or general fatigue, which further obscure clinical suspicion. Pain and tenderness are uncommon, contributing to frequent misdiagnoses as hematomas or vascular malformations like hemangiomas [7,8].

The imaging characteristics of AFH remain insufficiently defined and are often nonspecific. In most reported cases, AFH appears as a lesion containing both cystic and solid components, where the cystic areas frequently consist of blood products, reflecting the tumor's angiomatoid features. This overlapping appearance makes it difficult to distinguish AFH from other lesions with fluid–fluid levels or even organized hematomas [9,10].

Since this tumor originates from soft tissue, conventional imaging modalities such as plain radiography and computed tomography have limited diagnostic utility. In contrast, ultrasound and MRI are more informative and are therefore the preferred approaches. Notably, ultrasonographic features have been described in only a small number of reported cases [11,12].

In one case series involving four pediatric patients who underwent ultrasound, three presented with predominantly solid-appearing lesions, two of which showed small cystic components, while the fourth displayed a multicystic mass with fluid-fluid levels and heterogeneous echotexture.

Peripheral and septal vascular flow was detected on color Doppler imaging [11].

In our case, ultrasonography revealed a multicystic lesion containing fluid-fluid levels, internal heterogeneity, and Doppler signal along the periphery and within septations. Based on these findings, the lesion was initially interpreted as a hematoma [10].

Although certain MRI features of AFH have been previously described, none are considered pathognomonic. The most suggestive finding is the presence of intralesional cystic spaces demonstrating fluid-fluid levels [13].

Additional imaging characteristics that may support the diagnosis include the presence of a pseudocapsule, internal hemorrhagic components such as hemosiderin, and perilesional edema in some cases [10].

A specific MRI finding that has been proposed is the "double rim sign," characterized by a high signal intensity outer rim and a low signal inner rim, visible on both T2-weighted and post-contrast sequences. While this sign may be incomplete or subtle, in our case, only a peripheral low signal rim was identified, consistent with a fibrous pseudocapsule [13].

The broad spectrum of imaging appearances outlined above can be observed in various soft tissue lesions. Moreover, percutaneous core biopsy may sometimes fail to establish a definitive diagnosis. Consequently, in our case, the differential diagnosis based on MRI included soft tissue aneurysmal bone cyst, giant cell tumor of soft tissue, hematoma, hemangioma, soft tissue sarcoma, and fat necrosis (Table 2). Correlating imaging findings with clinical context and patient age is essential to formulate the most accurate preoperative diagnosis, pending histopathological confirmation following surgical excision [14,15].

The standard treatment for AFH involves wide local excision, followed by imaging-based surveillance to detect any recurrence. In cases where the tumor is unresectable or has metastasized, adjuvant therapies such as chemotherapy and/or radiotherapy may be considered. Generally, AFH carries a favorable prognosis and clinical outcome, particularly when complete surgical resection is achieved [16,17].

TEACHING POINT

Angiomatoid fibrous histiocytoma (AFH) is a rare soft tissue tumor with non-specific imaging features that should be differentiated from other soft tissue lesions presenting with fluid–fluid levels on Magnetic Resonance Imaging. Recognizing suggestive findings such as intralesional cystic spaces with fluid–fluid levels, a surrounding pseudocapsule, hemorrhagic components (e.g., hemosiderin), and perilesional edema—when correlated with appropriate clinical context—is essential for raising suspicion of AFH and guiding appropriate management, as preoperative histopathological diagnosis via percutaneous biopsy may be challenging.

QUESTIONS

Question 1: Which of the following answer choices is false?

a. Angiomatoid fibrous histiocytoma is classified as an intermediate tumor of uncertain differentiation.

b. Angiomatoid fibrous histiocytoma was first described by Enzinger in 1979.

c. Angiomatoid fibrous histiocytoma is a common soft tissue tumor. (applies)

d. Angiomatoid fibrous histiocytoma has a local recurrence rate of 15–20%.

e. Angiomatoid fibrous histiocytoma has a distant metastasis rate of 1–3%.

Explanation: (c) Angiomatoid fibrous histiocytoma is rare, accounting for only about 0.3% of all soft tissue tumors. ["Angiomatoid fibrous histiocytoma is a rare mesenchymal soft-tissue neoplasm, accounting for approximately 0.3% of all soft tissue tumors..."]

Question 2: Which of the following answer choices is false?

a. Angiomatoid fibrous histiocytoma typically affects pediatric and young adult patients.

b. The median age at diagnosis is 13 years.

c. Angiomatoid fibrous histiocytoma is most frequently found in the brain. (applies)

d. Angiomatoid fibrous histiocytoma can involve unusual sites such as the lungs or vulva.

e. Angiomatoid fibrous histiocytoma may infiltrate skeletal muscle and periosteum.

Explanation: (c) Angiomatoid fibrous histiocytoma most commonly arises in the extremities, not the brain. ["It most commonly arises in the extremities..."]

Question 3: What is the most common anatomical site of Angiomatoid fibrous histiocytoma?

a. Retroperitoneum

b. Head and neck

c. Extremities (applies)

d. Kidneys

e. Mediastinum

Explanation: (c) Although Angiomatoid fibrous histiocytoma can appear in atypical sites such as retroperitoneum or kidneys, the **extremities** are the most common location. ["It most commonly arises in the extremities..."]

Question 4: Which of the following is the most characteristic imaging feature of Angiomatoid fibrous histiocytoma on Magnetic resonance imaging?

a. Central calcification
b. Intralesional cystic spaces with fluid-fluid levels (applies)
c. Uniform high T1 signal
d. Extensive fatty infiltration
e. Large necrotic core without peripheral rim

Explanation: (b) **Fluid-fluid levels within cystic spaces** are the most suggestive, though not pathognomonic, imaging feature of Angiomatoid fibrous histiocytoma on Magnetic Resonance Imaging. ["The most suggestive finding is the presence of intralesional cystic spaces demonstrating fluid-fluid levels."]

Question 5: What is the standard first-line treatment for Angiomatoid fibrous histiocytoma?

1. Chemotherapy
2. Radiotherapy
3. Wide local excision (applies)
4. Observation only
5. Embolization

Explanation: (c) The standard approach to treating Angiomatoid fibrous histiocytoma is **wide local excision**, with additional surveillance for recurrence. ["The standard treatment for angiomatoid fibrous histiocytoma involves wide local excision, followed by imaging-based surveillance..."]

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FIGURES

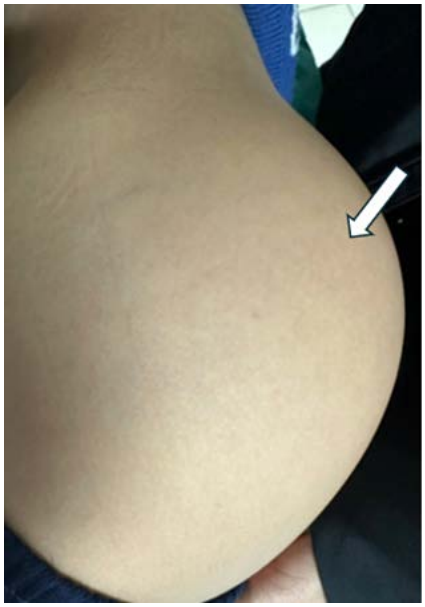

Figure 1: Clinical image of a 10-year-old male patient showing a skin-colored mass measuring approximately 15×15 cm in the left gluteal region (*white arrows)*.

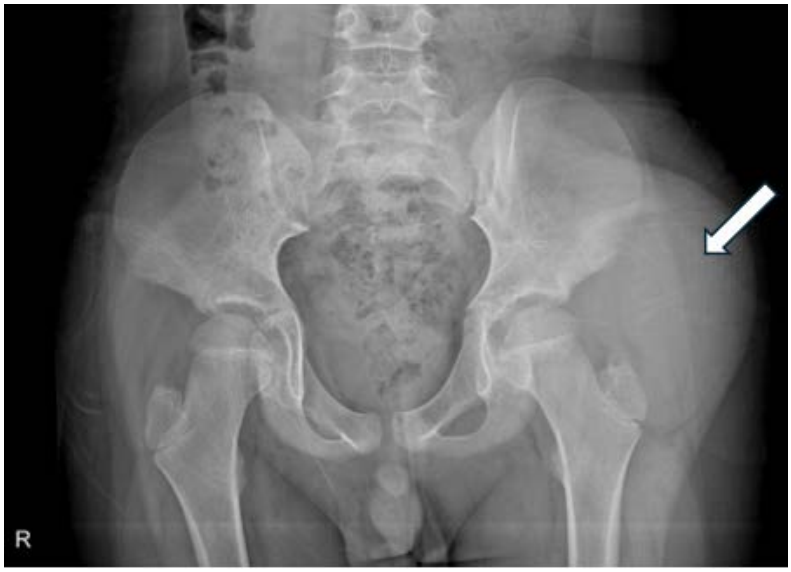

Figure 2: Pelvic X-ray of a 10-year-old male patient reveals a soft tissue density mass in the left gluteal region, measuring 12x8 cm, with no evidence of bone erosion or calcification.

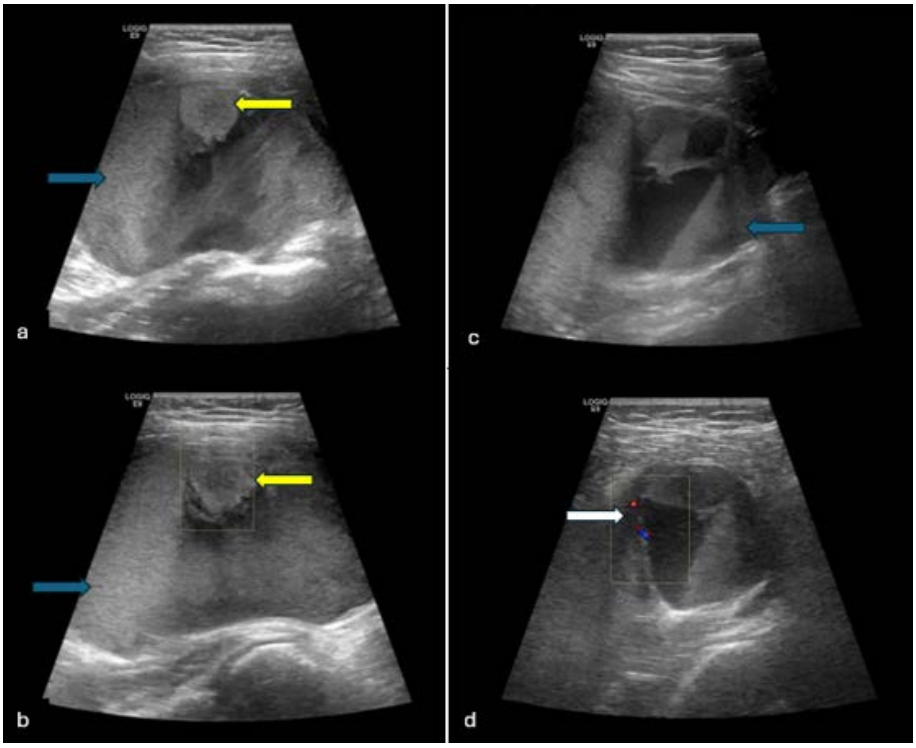

Figure 3: Ultrasound of the left gluteal soft tissue of a 10-year-old male patient revealed a large multiloculated soft tissue mass containing heterogeneous hyperechoic fluid-debris levels consistent with hemorrhagic fluid (blue arrow), measuring 10x6cm. Additionally, a nodule on the cyst wall showed no vascular signal on color Doppler, suggestive of a blood clot (yellow arrow). The mass was firm, without a change in shape upon probe compression. Peripheral vascular proliferation of the lesion was observed on color Doppler imaging (white arrow). The lesion was initially diagnosed as a hematoma on ultrasound.

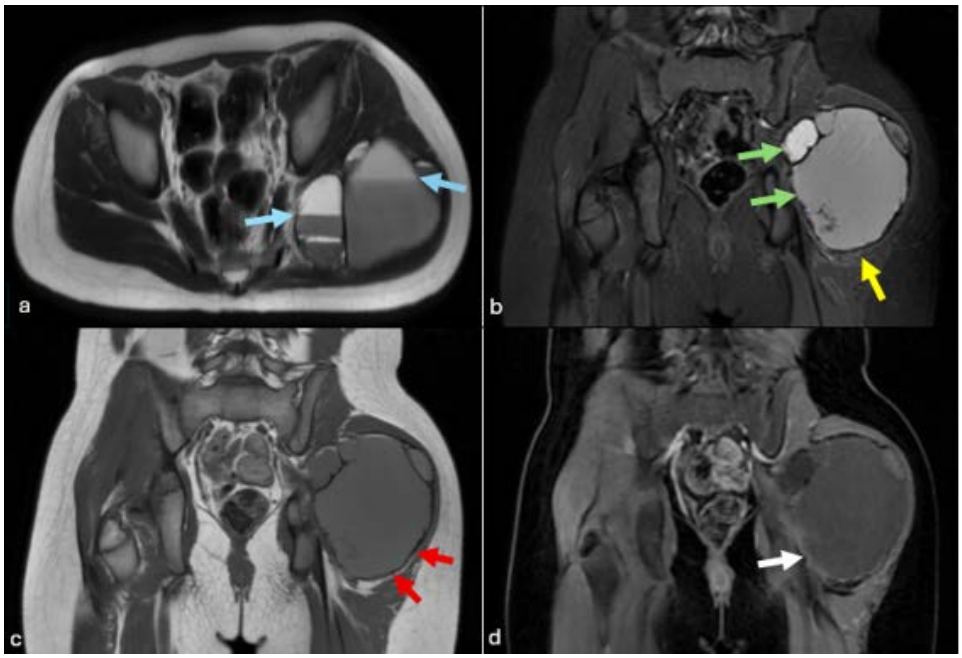

Figure 4: MRI sequences of the left gluteal region in a 10-year-old male patient: (a) T2-weighted image (T2W), (b) T2-STIR image, (c) T1-weighted image (T1W), and (d) post-contrast T1-weighted image.
A soft tissue mass measuring 10x6x10cm is seen in the left gluteal region. The lesion demonstrates heterogeneous signal intensity on T1W, T2W, and STIR sequences, with cystic components (green arrow) containing fluid–fluid levels (blue arrow), a pseudocapsule (red arrow), and perilesional edema (yellow arrow). Post-contrast images show partial gadolinium enhancement (white arrow).

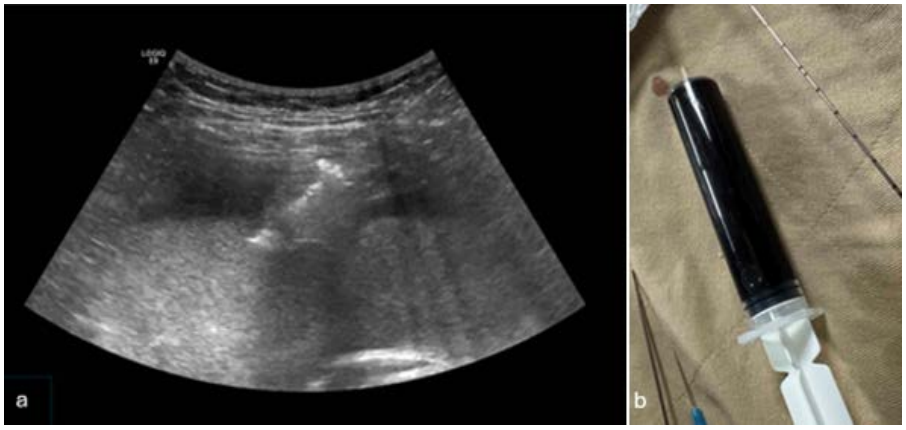

Figure 5: Image of percutaneous biopsy of the soft tissue mass in the left gluteal region of a 10-year-old male patient, along with the aspirated fluid from the mass, which appeared red and blood-like.

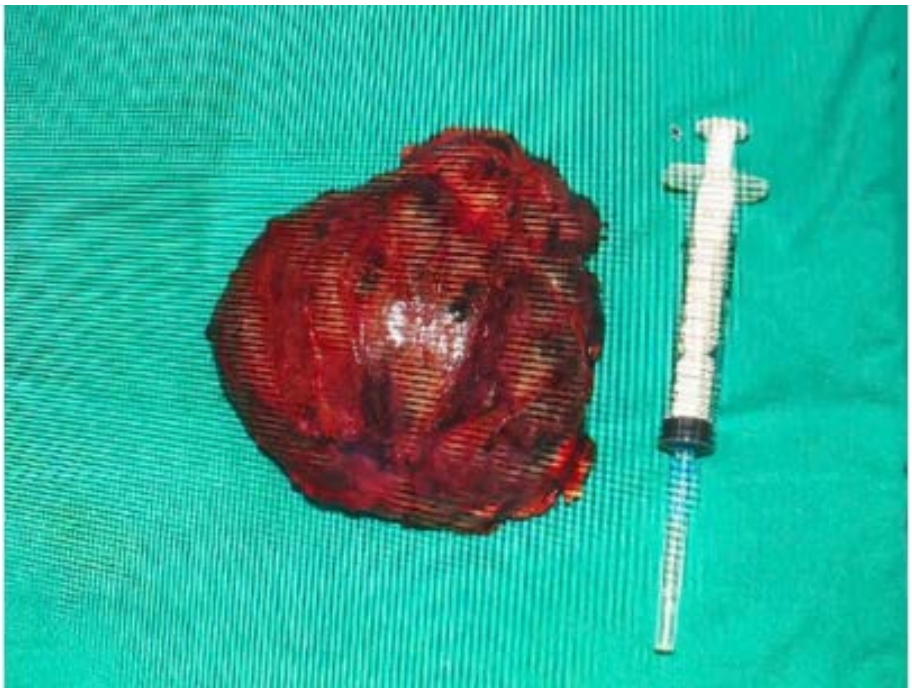

Figure 6: Intraoperative findings in a 10-year-old male patient revealed a well-encapsulated soft tissue tumor with a smooth surface, located between the gluteus maximus, and gluteus medius, gluteus minimus muscles in the left gluteal region. The tumor measured approximately 15 × 15 cm in size.

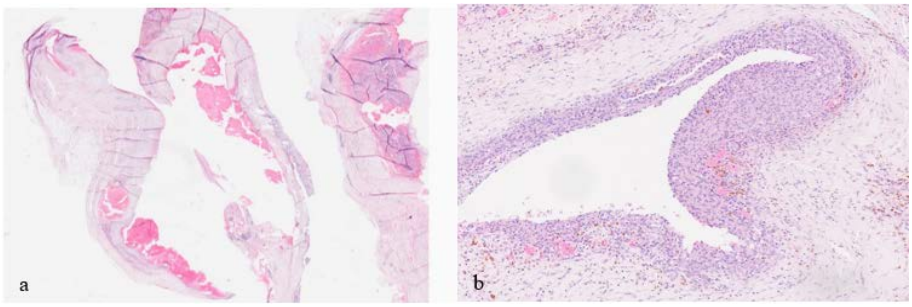

Figure 7: A 10-year-old male patient with angiomatoid fibrous histiocytoma located in the soft tissue of the left gluteal region. Histopathological examination and diagnosis were performed using both Hematoxylin and Eosin (H&E) and Periodic Acid-Schiff (PAS) staining. The tumor tissue was well demarcated and composed of spindle-shaped or round nuclei cells, with prominent proliferation of numerous small, thin-walled blood vessels containing abundant red blood cells. No necrosis or mitotic figures were observed. Multiple areas showed markedly dilated vascular spaces. Immunohistochemical staining results (not shown): Tumor cells were positive for CD68 and CD99, focally positive for EMA, with a Ki-67 index of 3%. They were negative for Desmin, SMA, S100, and CD34.

Table 1: Summary table of Angiomatoid Fibrous Histiocytoma (AFH)

Feature	Details
Etiology	Unknown; no definitive causative factors identified
Incidence	Accounts for approximately 0.3% of all soft tissue tumors
Gender Distribution	Slight male predominance
Age Predilection	Most commonly affects children and young adults (<30 years old)
Risk Factors	No established risk factors
Treatment	Wide local surgical excision
Prognosis	Favorable; distant metastases reported in approximately 1–3% of cases

Table 2: Differential diagnosis of angiomatoid fibrous histiocytoma (AFH) from soft tissue tumors exhibiting fluid-fluid levels on magnetic resonance imaging (MRI).

Entity	Clinical Features	X-ray	Ultrasound	Magnetic resonance imaging (MRI)
AFH (Angiomatoid Fibrous Histiocytoma)	- Age <30 - Slowly growing subcutaneous mass - Painless or mildly painful - May present with systemic symptoms (fever, anemia)	- Soft tissue shadow - No calcification - No bone involvement	- Well-defined hypoechoic mass - Possible fluid-fluid levels - Peripheral vascularity	- Fluid-fluid levels (due to hemorrhage) - Pseudocapsule - High T1 (methemoglobin) and T2 signal - Mild to moderate enhancement of solid areas
Soft Tissue ABC (Aneurysmal Bone Cyst)	- Teenagers and young adults - Rapidly growing mass, may be painful - No systemic symptoms	- Soft tissue mass - No bone involvement - No calcifications	- Multiloculated cystic mass - Clear fluid-fluid levels - No significant vascularity	- Multiple fluid-fluid levels - No solid enhancing components - No invasion - Minimal or septal enhancement
Soft Tissue GCT (Giant Cell Tumor)	- Middle-aged adults - Periarticular location - Sometimes painful - No systemic symptoms	- May show soft tissue mass - Possible fine calcifications	- Mixed solid-cystic lesion - Prominent vascularity	- Fluid-fluid levels (due to degeneration) - Enhancing solid areas - High T2 signal - Usually non-invasive
Hematoma	- History of trauma or procedure - Soft, changing mass over time - Often painful in acute phase	- Nonspecific soft tissue density - May show increased density if acute	- Fluid-fluid levels - No internal vascularity - No solid components	- Signal varies with age of blood - T1 hyperintense if subacute - No enhancement unless infected
Hemangioma	- Present since childhood - Soft mass - Changes with position - May cause pain or bleeding	- May show phleboliths (round calcifications)	- Mixed echogenicity - Low-flow vessels - May show fluid-fluid levels if bleeding	- Fluid-fluid levels possible - Very high T2 signal - Septal enhancement - May show phleboliths (low signal on all sequences)
Soft Tissue Sarcoma	- Any age (typically >40) - Rapidly growing painful mass - No systemic symptoms - Possible neurovascular involvement	- Large soft tissue mass - May cause bone erosion - Calcification uncommon	- Heterogeneous mass - Prominent neovascularization - Ill-defined margins	- May show fluid-fluid levels (necrosis/hemorrhage) - Strong enhancement of solid areas - Infiltrative, irregular margins
Fat Necrosis	- History of trauma, injection, or surgery - Painless soft mass - Slowly changing or stable	- Soft tissue opacity - May show fat density or calcification - May contain gas	- Heterogeneous echotexture - May show fluid-fluid levels	- Mixed signal intensity - Fat signal (drops on fat-sat T1) - No significant enhancement - No invasive features

Key Features Suggesting AFH:

Finding
- Young age (<30), superficial soft tissue location
- Fluid-fluid levels due to hemorrhage
- Pseudocapsule appearance on MRI
- Mild enhancement of solid portion
- No invasion of surrounding tissues
- Possible systemic symptoms (fever, anemia)

KEYWORDS

Angiomatoid fibrous histiocytoma; MRI; X-ray; Ultrasound; soft-tissue.

ABBREVIATIONS

AFH = Angiomatoid Fibrous Histiocytoma
MRI = Magnetic Resonance Imaging