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Angiofibroma at Unusual Site - A Case Report

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ABSTRACT

Angiofibroma is an uncommon benign mesenchymal neoplasm of soft tissue, first described in 2012 and recognized in the 2020 World Health Organization classification of soft tissue and bone tumors. It has different locations and the diagnosis is suggested by clinical evaluation and imaging findings, and confirmed by histopathological examination of the excised specimen. Microscopically, it appears as a well-circumscribed tumor composed of bland spindle cells within a variably collagenous to myxoid stroma, permeated by a rich network of thin-walled blood vessels. Because of its variable morphology and absence of specific immunohistochemical markers, angiofibroma often poses a broad differential diagnosis. We present the case of a 52-year-old male with an abdominal mass, diagnosed as angiofibroma based on histopathological and immunohistochemical findings..

Keywords: *Angiofibroma (AF), World Health Organization (WHO), Epithelial membrane antigen [EMA], Signal Transducer and Activator of Transcription 6 [STAT6].*

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1. INTRODUCTION

AF is an uncommon benign mesenchymal neoplasm, first reported by Mariño-Enríquez and Fletcher in 2012 [1] and later incorporated into the 5th edition of the WHO classification of soft tissue and bone tumors (2020) [2]. They typically arise in the extremities, most often in the legs, with a predilection for sites adjacent to large joints such as the knee, though unusual locations including the back, abdominal wall, pelvic cavity, and breast have also been reported. The tumor is usually subcutaneous but may also occur intramuscularly. They are often slow growing painless masses [2]. Histologically, they are characterized as circumscribed neoplasm composed of bland spindle cells set within a variably collagenous to myxoid stroma, traversed by a prominent network of thin-walled blood vessels. Immunohistochemically, tumor cells show variable expression of CD34, EMA, and Desmin, while STAT6 is typically negative. The characteristic molecular alteration is a t(5;8)(p15;q13) translocation, leading to the AHRR-NCOA2 fusion gene [3].

We report the case of a 52-year-old male with a 3-year history of abdominal swelling, subsequently diagnosed as angiofibroma on histopathological and immunohistochemical examination.

2. CASE REPORT

A 52-year male presented to surgery OPD with complaints of swelling in the abdomen for 3 years. The patient was apparently normal 3 years back after which he noticed a swelling in the umbilical region which was insidious in onset, gradually increased in size to attain the current size. He also experienced intermittent pain and swelling was reducible on lying down position, increased in size on standing. On examination, a vague mass of size approximately 10 x 8 cm present just below the umbilicus, extending up to the suprapubic region inferiorly and left iliac fossa laterally. The mass was firm in consistency and moves freely in the horizontal axis and not in the vertical axis. Firstly, MRI pelvis, a large well defined lobulated hetero intense lesion measuring~14.4x8.6x12.4 cm noted in mesentery of lower abdomen infra umbilically with extension into pelvic region, lesion appear iso to hypointense on T1 and T2W images with intervening T2 hyper intense areas. The lesion causing mass effect in the form of posterolateral displacement of urinary bladder and supero-lateral displacement of small bowel loops and a small umbilical hernia also noted **[Fig 1]**. The patient underwent USG guided trucut biopsy taken from infra umbilical abdominal lesion. Microscopic examination revealed a lesion consisting of relatively monomorphic benign appearing spindle cells with areas of hyalinized stroma **[Fig 3a].** There were also dilated capillaries in the stroma and no mitosis / necrosis noted. Thus, a diagnosis of benign spindle cell lesion made and requested for initial IHCs - SMA, BETA - CATENIN, Ki-67, VIMENTIN and CD-117 for further categorization. IHCs were deferred at request. Then, the patient underwent laparotomy and proceed with umbilical herniorrhaphy. Intra operatively, the mass

was seen pre peritoneal area and had a feeding artery from internal iliac artery. The feeding vessel is ligated and the mass was excised in toto.

Radiology – MRI

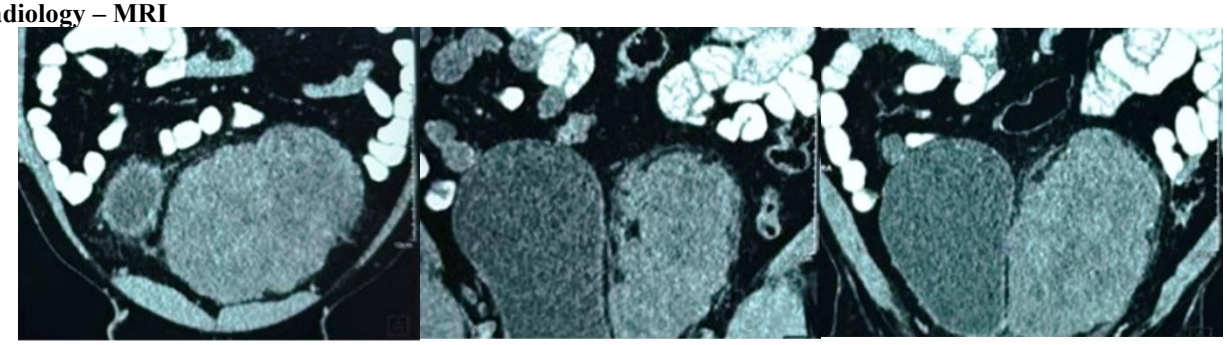

Figure 1: MRI pelvis revealed a large well defined lobulated hetero intense lesion measuring~14.4x8.6x12.4 cm noted in mesentery of lower abdomen infra umbilically with extension into pelvic region

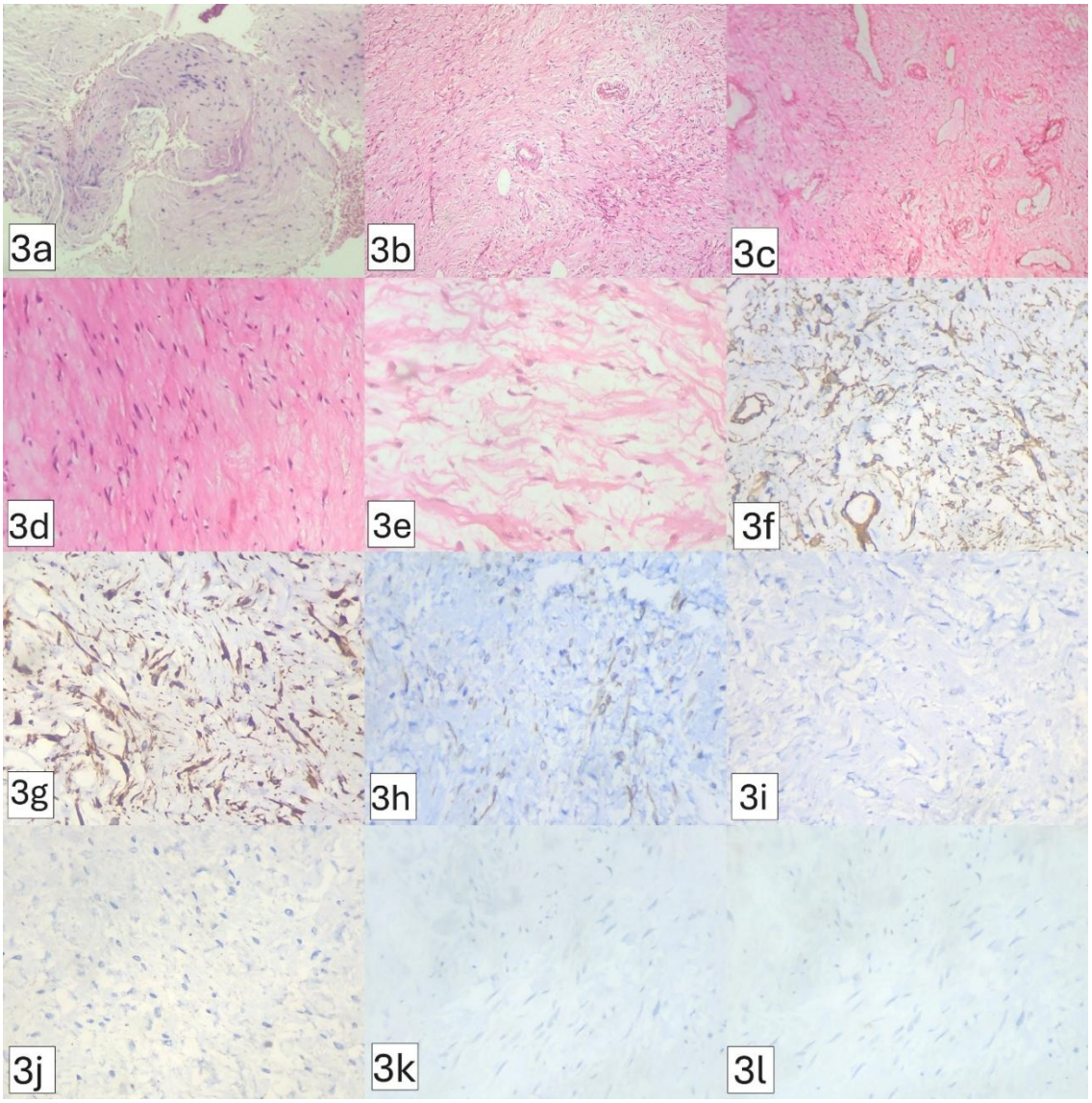

Figure 3: 3a) Microscopy of trucut biopsy shows a lesion consisting of relatively monomorphic benign appearing spindle cells with areas of hyalinized stroma [H&E,100x]. 3b & 3c) Microscopy shows cellular areas of spindle cell tumor along with proliferation of blood vessels [H&E,100x]. 3d) Microscopy shows tumor cells are of bland, spindle-shaped with slender nuclei and syncytial cytoplasm [H&E,400x]. 3e) Microscopy shows hypocellular areas

of the tumor [H&E,400x]. 3f) CD34 shows membranous positivity [IHC,100x]. 3g) Desmin showing cytoplasmic positivity [IHC,100x]. 3h) BCL2 shows variable cytoplasmic positivity [IHC,100x]. 3i) Ki67 index – 2% [IHC,100x]. 3j) Beta Catenin shows negative nuclear staining [IHC,100x]. 3k) STAT6 shows negative nuclear staining [IHC,100x]. 3l) MUC4 shows negative nuclear staining [IHC,100x].

Grossly, we received large globular partially cut open grey white to grey yellow soft tissue mass - 12.5x 11.5 ×8.5 cm. On cut section, shows grey white and grey yellow areas, few areas of haemorrhage and firm in consistency **[Fig 2a,2b].**

GROSS IMAGE

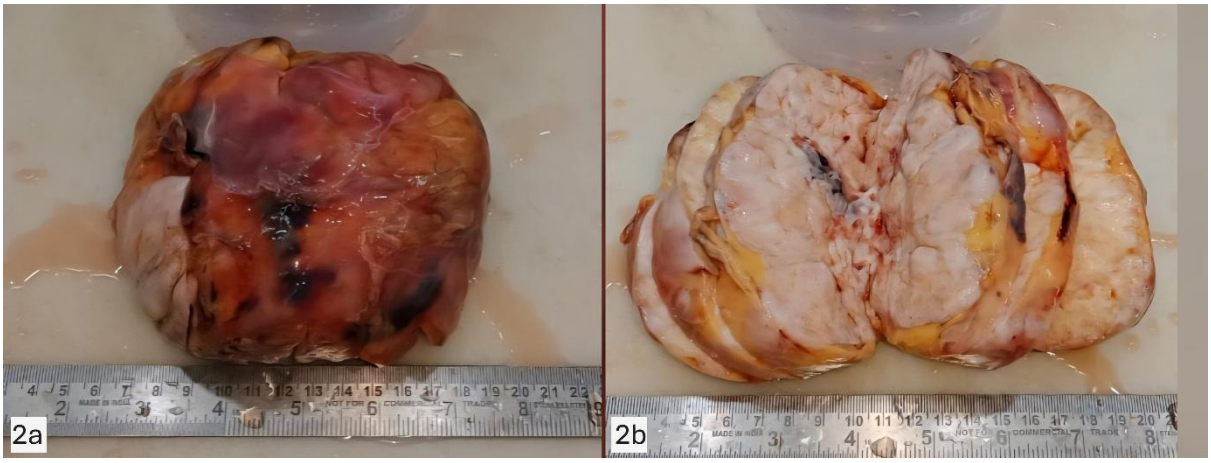

Figure 2: 2a) Gross – a single large globular partially cut open grey white to grey yellow soft tissue mass measuring 12.5x 11.5 ×8.5 cm. 2b) Cut surface shows grey white and grey yellow areas along with few areas of haemorrhage.

Histology image

Microscopically, H&E sections reveal spindle cell proliferation arranged in an irregular sheet-like pattern **[Fig 3b].** The peripheral regions exhibit myxoid changes with fat infiltration and show numerous thin-walled blood vessels, some displaying a staghorn configuration. Cellularity is variable, predominantly hypocellular **[Fig 3e]** with occasional hypercellular foci **[Fig 3c].** The tumor cells are bland, spindle-shaped with slender nuclei and syncytial cytoplasm **[Fig 3d]**. Circumferential margins show multifocal infiltration, and focal areas of haemorrhage are present. No mitotic activity or cytologic atypia is identified. With the above features, we concluded it as spindle cell neoplasm. To categorize the spindle cell neoplasm, we first performed IHC for CD34, BCL2, Beta catenin, and Ki-67. The tumor was CD34 positive **[Fig 3f]** and BCL2 – variable positive **[Fig 3h]**, β-catenin negative **[Fig 3j]**, and showed a low Ki-67 index -2% **[Fig 3i]**. Since β-catenin was negative, we proceeded with additional IHCs - STAT6, Desmin, MUC4. The tumor showed Desmin positivity **[Fig 3g]** with STAT6 and MUC4 negativity **[Fig 3k,3l].**

3. DISCUSSION

AF is an uncommon soft tissue tumor, first characterized by Mariño-Enríquez and Fletcher in 2012 through a case series of 37 patients [4]. AF shows a female predominance, occurs across a wide age range, most commonly involves the lower extremities and rare in other locations such as the back, abdominal wall, pelvic cavity, and breast [2]. **Liu JY et al** [5] reported a case of omental angiofibroma with indirect inguinal hernia leads to a scrotal mass. The exact etiology not known but the pathogenesis include molecular alteration is a t(5;8)(p15;q13) translocation, leading to the AHRR-NCOA2 fusion gene [2]. Very few literatures have shown radiological findings of angiofibroma. **Jun Nishio et al** [6] studied MRI of soft tissue angiofibroma in the left knee shows a mass with iso-signal intensity relative to skeletal muscle on T1-weighted sequences and heterogeneous high signal intensity on T2-weighted sequences. CT and MRI are the better imaging studies for angiofibroma [5]

Grossly, angiofibroma usually appears as a sharply defined, nodular or multilobulated solid lesion of varying dimensions, with a yellowish to whitish, more often glistening cut surface, sometimes containing cystic or haemorrhagic regions [2]. Histologically, nodular or vaguely lobulated architecture composed of spindle cells with alternating hypocellular, edematous or myxoid zones and more cellular, collagen-rich areas. A prominent feature is the intricate vascular network, predominantly composed of numerous small, thin-walled, branching vessels that are evenly distributed throughout the lesion. In addition, medium- to large-sized vessels with variably thickened walls may be present, often exhibiting ectatic lumina and a staghorn-like configuration [1]. The cells are spindled shaped, short tapered nuclei, indistinct nucleoli and inconspicuous pale eosinophilic cytoplasm [2]. Mitotic figures may be observed rarely; however, cytologic atypia and nuclear hyperchromasia are not present [6]. Immunohistochemically, tumor cells show variable expression of CD34, EMA, and Desmin, while STAT6 is typically negative [3]. There are studies that shows angiofibroma is typically positive for

CD163 and ER [1,6]. However, in recent times, WHO recommends to do molecular testing for AHRR-NCOA2 fusion gene mutation [2].

The differential diagnosis of angiofibroma particularly as abdominal mass with the above histological findings are desmoid fibromatosis, solitary fibrous tumor, myofibroblastoma, spindle cell lipoma and low-grade fibromyxoid sarcoma [1,2]. All these tumors can mimic the histological pattern of angiofibroma. Desmoid fibromatosis may mimic cellular angiofibroma histologically due to its spindle cell proliferation in a collagenous stroma. However, unlike angiofibroma, it shows infiltrative borders and nuclear Beta Catenin positivity on IHC [7]

Solitary fibrous tumor (SFT) can resemble angiofibroma histologically with spindle cells and prominent branching ("staghorn") vessels, but SFT typically shows nuclear STAT6 positivity, unlike angiofibroma [1]. Myofibroblastoma may mimic angiofibroma by showing bland spindle cells in a collagenous stroma but with less prominent vasculature; it usually displays strong Desmin positivity with often weaker CD34, distinguishing it from angiofibroma [8]. Spindle cell lipoma may mimic angiofibroma with bland spindle cells in a collagenous background, but it typically shows ropey collagen and is CD34 positive, Desmin negative, helping to distinguish it from angiofibroma [9]. Low-grade fibromyxoid sarcoma may mimic angiofibroma with bland spindle cells, but it characteristically shows alternating fibrous and myxoid areas and is MUC4 positive, Desmin negative, which separates it from angiofibroma [10]. There are very few literatures saying about recurrences of the angiofibroma [4] and being benign in nature no malignant transformation and metastasis reported [4,6].

4. CONCLUSION

Angiofibroma is a relatively uncommon tumor and they are more commonly reported in the pelvic cavity than the abdominal cavity. Histopathologically, they can mimic many benign spindle cell tumors. The rich vasculature, often with thin-walled branching vessels in a collagenous to edematous stroma, helps in distinguishing it from other differentials. Accurate recognition is crucial, as angiofibroma is a benign lesion with excellent prognosis, avoiding unnecessary aggressive treatment that may be considered for its mimics. Given its rarity, especially in the abdominal cavity, awareness of its histopathological spectrum and immunoprofile is essential to prevent misdiagnosis.

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