

Case Report

Angiomyxoma of thigh in a young lady: rare presentation

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ABSTRACT

Angiomyxoma is a rare tumor that typically occurs in the perineum in women of reproductive age. It is very rare to present in the subcutaneous plane at thigh region in a young female. Angiomyxoma belongs to one group of myxomas tumors. They occur at various locations such as heart, bones, skin subcutaneous and aponeurotic tissues, urogenital tract and skeletal muscles. These tumors are characterized by having abundant mucoid and myxoid extracellular matrix. Angio myxoma is a rare benign mesenchymal tumor in approximately in 95% cases. The incidence is reported as varying from 0.1 to 0.13 per 100,000 population. These tumors are encompassing a broad spectrum of clinical behavior ranging from benign to malignant. Most of these tumors located in intramuscular plane and in our case the tumor located in the subcutaneous plane and presented in a young lady. We have done wide local excision for that tumor and came out as cutaneous angiomyxoma of thigh. We are concluding that due to these tumors having broad spectrum of clinical behavior as well as high rate of recurrence, preoperatively thoroughly investigated to rule out the soft tissue sarcomatous conditions as well as to plan exact mode of treatment.

Keywords: Myxoid myxoma, Angiomyxoma, Extracellular matrix, Neoplasms, Sarcoma

INTRODUCTION

Myxoid tumors are mesenchymal in origin and rare benign type.¹ They are known to originate from primitive mesenchymal cells. These tumors are encompassing a broad spectrum of clinical behavior ranging from benign to malignant. The common sites include the thigh, upper arm, calf, and the buttocks. Based on radiological and histopathological findings, benign myxomas are further divided into intramuscular myxoma, aggressive angiomyxoma, superficial angiomyxoma, myxolipoma, dermal myxoma, and acral fibromyxoma.² Magnetic resonance imaging is the radiological modality of choice in diagnosing these cases.³ The tissue biopsy is the gold standard to diagnose it.⁴ There are more than 60 reactive and neoplastic entities currently classified under its domain. These tumors having varied clinical and histopathologic features continue to pose a diagnostic challenge to clinicians and pathologists. Here we describe a rare case of angiomyxoma of thigh in young female in subcutaneous plane.

CASE REPORT

A 22-year-old female patient presented to us with a swelling of the right thigh. The swelling was painless and had gradually increased in size since its appearance 2 years prior to presentation to us. The patient had observed the swelling grow to its present size which was relatively painless in the past, but painful for the past 3 months, thereby causing difficulty in his activities of daily living. On clinical examination the swelling was in size located at lateral side of thigh. The swelling was firm in consistency mobile well defined and in subcutaneous plane. After history and clinical examination with a diagnosis of soft tissue tumor of thigh she underwent blood and radiological investigations. Routine blood and urine examination had not revealed any abnormality. The patient was investigated with X-ray, ultrasound and MRI. The plain radiograph of the limb showed no bony involvement. Ultrasound of thigh swelling showed hypoechoic lesion with vascularity in with in it. MRI revealed a well circumscribed encapsulated lobulated soft

tissue mass measuring 4.5×4.2×4.8 cm in the lower half of the right thigh in the subcutaneous plane with suggestive of dermatofibrosarcoma protuberance. Fine needle aspiration cytology (FNAC) revealed features of a benign cystic lesion-myxolipoma. Tumor workup such as computed tomography of the chest and ultrasonography of the abdomen was normal. With this pathological diagnosis, surgical excision was planned after valid consent. Under spinal anesthesia wide local excision with 2 cm margin was performed with by an elliptical incision incorporating the biopsy site into it. Post operative course was uneventful. post operative histopathological examination revealed hypocellular area composed of scattered spindle and satellite shaped cells and inflammatory cells and thin-walled blood vessels on myxoid stroma suggestive of superficial angiomyxoma. Atypia is absent. Patient was discharged on post operative day-4 and followed up in the OPD.

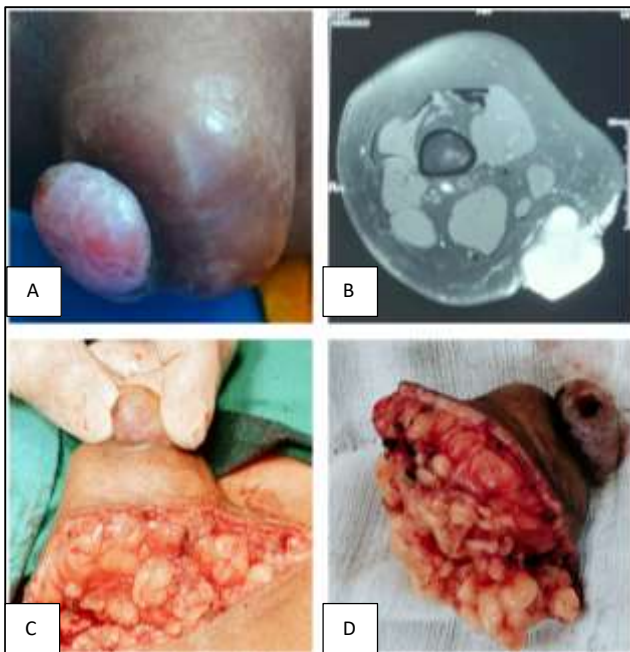


Figure 1 (A-D): Preoperative picture around 5×4 cm of soft tissue mass presenting at right thigh. Preoperative MRI shows a well circumscribed encapsulated lobulated soft tissue mass measuring 4.5×4.2×4.8 cm in the lower half of the right thigh in the subcutaneous plane. Intra operative picture of tumor wide local excision done. Post operative excised specimen with 2 cm margin.

DISCUSSION

Angiomyxoma is a rare benign mesenchymal tumor. They occur in approximately 95% of cases, occurs in females of reproductive age. Angiomyxoma patients are usually asymptomatic due to the slow and insidious growth pattern of the tumor.⁵ As previously reported the female to male ratio is approximately 6.6:1. Steeper and Rosai reported the locally aggressive behavior of the tumor and its tendency to recur locally.⁶ These benign

mesenchymal tumors occurring almost exclusively in the pelvis and perineum of pre-menopausal age group. These benign tumors appears to be hormonally regulated.

Patients typically present with a slowly growing, painless mass, which is usually very large and extensive by the time of diagnosis. In our index she presented with painless mass in right thigh with gradual progressive in size. In women, these rare lesions are often misdiagnosed clinically as a vulvar abscess, lipoma, Gartner's duct cyst, Bartholin's cyst or vaginal prolapse. Angiomyxoma may be discovered incidentally and occasionally discovered during pelvic examination or imaging.⁷

The imaging appearances of aggressive angiomyxoma have been well described, but are limited to case reports or small case series. On cross-sectional imaging, these neoplasms typically appear well circumscribed. They appear hypoechoic on ultrasound, and are usually hypodense on CT.⁸ In our case the MRI suggestive of loose myxoid matrix due to high water content. The tumors often enhance following intravenous contrast administration, reflecting their high vascularity. Mildly heterogenous high signal is generally demonstrated on fluid-sensitive MR sequences.⁹ A distinctive but inconstant finding is the presence of swirling strands of lower attenuation and low signal coursing through the tumour.¹⁰

After postoperative surgical excision histological examination shows hypocellular area composed of scattered spindle and satellite shaped cells and inflammatory cells and thin-walled blood vessels on myxoid stroma suggestive of superficial angiomyxoma. Atypia is absent.

Histological analysis typically shows small, spindle shaped mesenchymal cells dispersed in a loose myxoid matrix, with characteristic dilated blood vessels.¹¹ Mitoses and cellular atypia are generally absent. The usual immunoprofile of this neoplasm is vimentin, desmin and CD34 positive, and S-100 protein and muscle-specific actin negative.¹² The majority of lesions in women stain strongly positive for oestrogen and progesterone receptors, but those in men are usually negative.¹³ Ultrastructural studies suggest an origin from a pluripotential perivascular primitive mesenchymal cell.

We have sent for required immunohistochemical staining of this tumor, particularly the reactivity for desmin, α -smooth muscle actin, muscle-specific actin, vimentin, CD-34, S-100 protein, estrogen and progesterin receptors.

In a review of the literature by Jingping et al the characteristic appearance using B-ultrasound imaging for the diagnosis of angiomyxoma was described as a hypoechoic, well-demarcated mass with multiple thin, echogenic internal septa.¹⁴ The sonographic findings in our index case were similar to those described by Jingping et al. On color doppler mode it shows internal

blood flow. Angiomyxoma is derived from myofibroblasts as a phenotypic variant of the basic fibroblast.

Surgery is the most effective method for the treatment of angiomyxoma.¹⁵ The excision with wide tumor-free margins should be performed for these types of tumors to avoid recurrence. We have done wide local excision with 2 cm margin in our index case. In some cases it may require adjuvant therapy also. In case of bigger size angiomyxomas angiographic embolization is required to shrink the tumor size preoperatively.¹⁶ Accurate diagnosis is required prior to surgery because specific treatment planned for angiomyxoma.

CONCLUSION

In conclusion we reported an uncommon case of cutaneous angiomyxoma of the thigh in a young female. On imaging it has sharply demarcated borders and a heterogeneous echotexture and weak blood flow. The mainstay of treatment for angiomyxoma of the thigh is surgical excision. Early definitive diagnosis is crucial to determining appropriate treatment strategies as well as to avoid recurrence.

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