An unusually large superficial angiomyxoma with delayed presentation due to COVID-19: a case report

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ABSTRACT

Background: Superficial angiomyxoma is a rare, benign, soft-tissue tumor. It has a non-specific presentation and there is no clear diagnostic investigation. Therefore, it is usually not suspected pre-operatively and managed incorrectly with a biopsy or narrow margin excision, which increases the recurrence rate.

Case Presentation: A case of an unusually large superficial angiomyxoma is described with a delayed presentation to a plastic surgery clinic due to the COVID-19 pandemic. The patient was a 53-year-old male with an atraumatic 8 × 4 cm subcutaneous, mobile swelling to the left thigh. Excision was performed with no evidence of recurrence at 6 months. The mass was an uniloculated cyst with gelatinous myxoid stroma. Immunohistochemistry was positive for Alcian blue and CD34.

Conclusion: This case report raises awareness of this rare lesion and provides a review of diagnostic aids as well as clear intra-operative photography of what a surgeon might expect when this tumor is encountered.

Keywords: superficial angiomyxoma, angiomyxoma, cutaneous tumor, carney complex, case report.

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Background

Superficial angiomyxoma is a rare, benign, cutaneous tumor. It was first reported in 1985 in association with the Carney complex although it may also arise sporadically [1,2]. Also known as cutaneous angiomyxoma, superficial angiomyxoma stands in contrast to "aggressive angiomyxoma" which is found in deeper tissues, particularly in the female genital region and pelvis.

On clinical examination, superficial angiomyxoma may present as a cutaneous nodule or papule, or as a lobulated subcutaneous mass [3]. It can, therefore, easily be mistaken for many other soft tissue growths including cysts, lipomas, amelanotic melanoma, cutaneous focal mucinosis, dermatofibroma, neurofibromatosis, and other myxoid tumors. The diagnosis is often not suspected until after excision and final histology and immunohistochemistry results.

Herein, an unusual case of a very large superficial angiomyxoma is presented with delayed diagnosis and management due to the COVID-19 pandemic. Written informed consent was obtained from the patient to publish this case report and the accompanying images.

Case Presentation

A 53-year-old male teacher presented to our clinic with a painless mass in the left thigh. He was otherwise fit and well, a non-smoker, and took no medication. The non-tender lump did cause some discomfort when sitting down. He could not recall any trauma to the affected area. There was no history of the lesion causing discharge or becoming infected.

The patient's general practitioner arranged an ultrasound scan which showed a well-defined, ovoid, non-compressible, hypoechoic, inhomogeneous lesion within the subcutaneous tissue, measuring 3.7 × 2.0 × 2.2 cm. No vascularity was demonstrated and there was no evidence of a punctum to the skin surface. The general practitioner planned an excision under local anesthetic, but this procedure was delayed and re-scheduled several times due to the COVID-19 pandemic. During this waiting period, the lesion grew and became no longer amenable to removal under local anesthesia. The change in size also prompted a repeat ultrasound and a 2-week wait referral to exclude more sinister pathology. Our patient was then seen in the orthopedic clinic and an MRI was arranged urgently

to exclude a sarcoma. The MRI showed a subcutaneous cystic lesion measuring 7.3 × 4.4 × 4.5 cm. There was a thin cyst wall with no complexity of the cyst content and no intramuscular component.

With sarcoma excluded, the patient was then referred to our plastic surgery team for removal of this suspected cyst. By the time of assessment in our clinic the patient had been troubled with the mass for over 6 years. Physical examination revealed a cystic lesion to the postero-lateral left thigh around 8.0 × 4.0 cm in size. The mass was subcutaneous, mobile, and transilluminable. There were no overlying skin changes and no regional lymphadenopathy.

The lesion was excised under general anesthesia with the patient in the right lateral position (Figure 1). A single dose of intravenous co-amoxiclav was administered on induction as it was suspected that the thin wall of the cyst might easily rupture. A longitudinal skin incision was made over the mass and dissection was performed to find the lesion lying superficially in the subcutaneous fat. The lesion was macroscopically cystic and appeared to contain gelatinous material. It was well-defined and easily freed in its entirety from the surrounding tissue without rupturing the lesion's thin wall. A feeding vessel was found at the base and cauterized, and the lesion was sent for histology (Figure 2). The wound was closed in layers and reviewed after 1 week at the outpatient clinic.

Histopathological examination described an encapsulated cyst-like structure of 8.5 × 5.0 × 3.9 cm. The cyst was uniloculated and on sectioning contained translucent gelatinous material. Microscopically there was prominent myxoid stroma and atypical mitoses were not present. The lesion tested positive for Alcian blue and CD34 but negative for CD31 and S100. Based on these findings a diagnosis of superficial angiomyxoma was made.

The wound healed well and there was no clinical evidence of recurrence at 6 months. The patient was assessed for manifestations of the Carney complex but there were no other clinical features to suggest this diagnosis. At the skin multidisciplinary team discussion, the recommendation was made to review the patient at 9 months with a repeat ultrasound to look for disease not evident to palpation.

Discussion

Superficial angiomyxoma was first described in 1985 in association with the Carney complex. This is a rare autosomal dominant disorder presenting with cardiac and endocrine tumors as well as cutaneous growths, including superficial angiomyxomas [1]. Following Carney's work, a case series described patients with isolated superficial angiomyxomas [2]. All patients with superficial angiomyxoma should be evaluated for features of the Carney complex, especially if multiple angiomyxomas are present.

The tumor usually presents as a slow-growing painless mass in the skin or superficial subcutaneous fat. It can be found anywhere in the body but is most often found in the trunk, head and neck, and lower extremities. Rare presentations in the upper limb have also been reported [3]. Superficial angiomyxoma can occur at any age but has a peak incidence in the fourth decade of life. It has an incidence of 0.008%-3% and is usually only a few millimeters in diameter but may grow to up to 5 cm in size [4]. Excision with clear margins is both the diagnostic and therapeutic modality of choice; however, local recurrence is seen in up to 38% of cases [5]. There is no metastatic potential and there have been no reports of malignant transformation to date.

Although the tumor is usually asymptomatic it may be tender to palpation. Cutaneous manifestations of the disease include a pink papule or nodule, or "red planet sign;" however, our patient presented with a non-specific subcutaneous swelling [6].

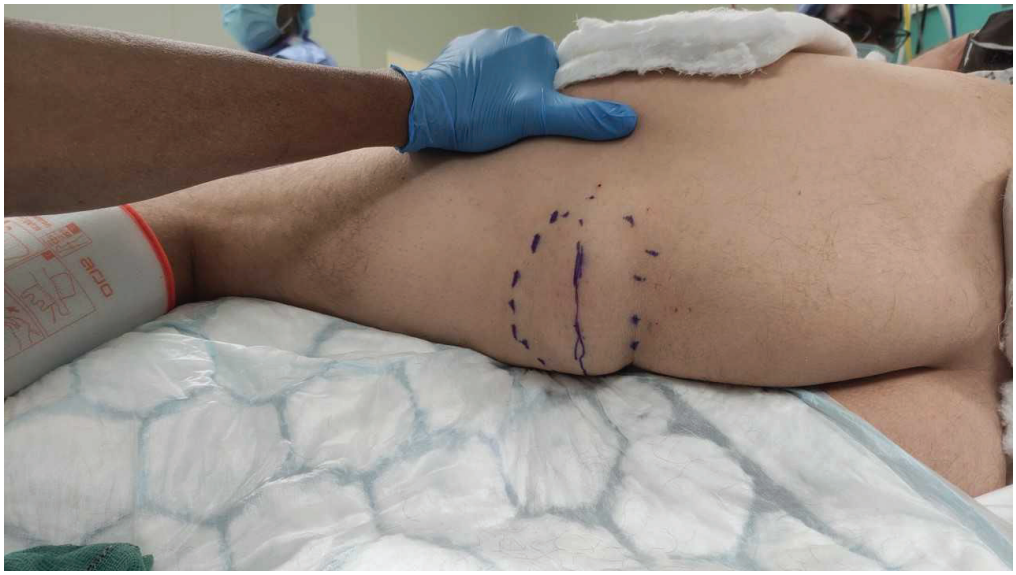

Figure 1. *Surface anatomy clinical appearance of lesion.*

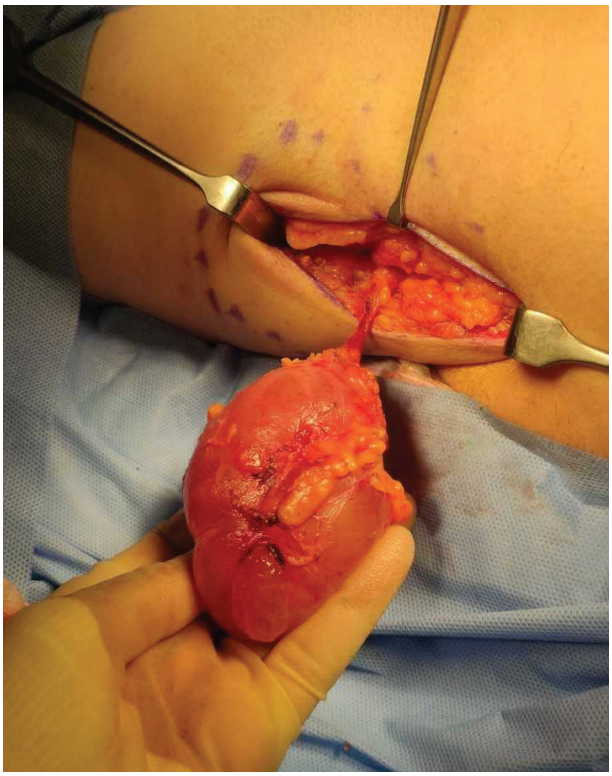

Figure 2. *Macroscopic appearance of lesion after dissection.*

Due to its rarity and non-specific presentation, the diagnosis of superficial angiomyxoma is often challenging and the pre-operative differential diagnosis is wide depending on the appearance of the lesion. As with the patient in our case report, patients may wait for several years before receiving a diagnosis. Imaging studies may delineate the mass relative to surrounding structures, rule out some more aggressive tumors, and aid with pre-operative planning but no scan is diagnostic for superficial angiomyxoma.

A definitive diagnosis can only be provided with histology and immunohistochemistry of tissue. The histological appearance of superficial angiomyxoma is of myxoid stroma with mucin pools, many thin-walled small blood vessels, and scattered bland stellate and spindled cells. Pleomorphism and mitotic figures are rare [7]. On immunohistochemistry, superficial angiomyxomas nearly always test positive for CD34. Smooth muscle actin, muscle-specific actin, factor XIIIa, and S-100 protein may be present to varying degrees. Desmin, keratin, estrogen receptor (ER), and progesterone receptor (PR) are usually negative [8].

Superficial angiomyxoma must also be differentiated from other myxoid tumors. Loss of PRKAR1A is a useful adjunct in differentiating superficial angiomyxoma from other histological mimics [9]. Cutaneous focal mucinosis is typically smaller and lacks the lobular structure and vascular infiltration characteristic of superficial angiomyxoma. Myxoid neurofibroma can be distinguished by the presence of Schwann and perineural cells and is nearly always positive for S100 on immunohistochemistry. Myxoid dermatofibrosarcoma protuberans and dermal nerve sheath myxoma both have multiple mitotic figures, high cellularity, and atypia [8]. Aggressive angiomyxoma is typically larger, more poorly defined, and locally infiltrative. Even if the tumor macroscopically appears relatively well defined, on microscopy it demonstrates infiltration into the surrounding adipose tissue. The vascular network of aggressive angiomyxoma is of a larger caliber. It is found in deeper tissues than superficial angiomyxoma, usually in the genital and pelvic regions, and tests positive for desmin, ER, and PR [10].

The treatment of superficial angiomyxoma is radical surgical excision, ideally with a wide margin of normal tissue to reduce the risk of recurrence; however, this may not always be achieved as the diagnosis is often not known until after surgical excision and histology results. Excision should be performed carefully to avoid rupturing the lesion and exposing surrounding tissues to the tumor cells. Radiotherapy may be used if the angiomyxoma is located in a cosmetically sensitive area or is too large for complete excision, or as an adjuvant therapy in cases of incomplete excision or recurrence. There are no systemic therapies for superficial angiomyxoma, and chemotherapy is not recommended. Due to the rarity of this tumor and the limited number of cases reported in the literature, the long-term prognosis is not well established.

Conclusion

Superficial angiomyxoma is a rare cutaneous tumor that is often pre-operatively mistaken for a common benign soft tissue tumor and managed with either a biopsy or narrow margin excision. This frequently leads to spillage of tumor cells, close margins, and incomplete excision which all increase the recurrence rate of the disease. This case report raises awareness of this rare lesion and describes an unusually large lesion but with a typical history and examination for superficial angiomyxoma and clear intra-operative photography of what a surgeon might expect this tumor to look like if encountered. We recommend wide surgical excision to minimize the risk of local recurrence.

What is new?

Superficial angiomyxoma is a rare cutaneous tumor that is often pre-operatively mistaken for a common benign soft tissue tumor and managed with either a biopsy or narrow margin excision. This frequently leads to spillage of tumor cells, close margins, and incomplete excision which all increase the recurrence rate of the disease. This case report raises awareness of this rare lesion and describes an unusually large lesion but with a typical history and examination for superficial angiomyxoma and clear intra-operative photography of what a surgeon might expect this tumor to look like if encountered.

List of Abbreviations

MRI Magnetic resonance imaging

Conflicts of interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

Funding

None.

Consent for publication

Written consent was obtained from the patient.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	**Patient (gender, age)**	53 years old male
2	**Final diagnosis**	Superficial angiomyxoma
3	**Symptoms**	Painless, growing mass
4	**Medications**	N/A
5	**Clinical procedure**	Surgical excision under general anesthetic with histology and immunohistochemistry testing
6	**Specialty**	Plastic surgery
7	**Objective**	To raise awareness of superficial angiomyxoma. This relatively rare tumor is usually not suspected pre-operatively and therefore is often managed incorrectly with a biopsy or narrow margin excision, which increases the recurrence rate.
8	**Background**	Superficial angiomyxoma is a rare, benign, soft-tissue tumor. It has a non-specific presentation and there is no clear diagnostic investigation.
9	**Case report**	An unusually large superficial angiomyxoma with a delayed presentation due to COVID-19
10	**Conclusions**	This case report raises awareness of this rare lesion and provides a review of diagnostic aids as well as clear intra-operative photography of what a surgeon might expect when this tumor is encountered. We recommend wide surgical excision to minimize the risk of local recurrence.
11	**MeSH keywords**	superficial angiomyxoma; angiomyxoma; cutaneous tumour; Carney complex; case report