

Solitary superficial angiomyxoma: an infrequent but distinct soft tissue tumor

Superficial angiomyxoma (SA) is a distinct soft tissue tumor characterized by a circumscribed collection of spindled and stellate fibroblasts that are admixed with thin-walled blood vessels and embedded in a mucinous stroma. Because of its relative infrequent occurrence, the purpose of this article was to present a classical example of an isolated superficial angiomyxoma and discuss the differential diagnosis.

Satter EK. Solitary superficial angiomyxoma: an infrequent but distinct soft tissue tumor.

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Superficial angiomyxoma (SA) is a distinct type of soft tissue myxoma characterized by increased numbers of stellate fibroblasts and thin-walled blood vessels admixed with a variable inflammatory infiltrate. Approximately 30% of lesions additionally contain an entrapped epithelial component consisting of keratin cysts or thin strands of squamous epithelium. Despite the fact that 20–30% of lesions recur, metastases have yet to be described.^{1–3} Because of its relative infrequent occurrence, the purpose of this article was to present a classical example of an isolated superficial angiomyxoma and discuss the differential diagnosis.

Case report

A 35-year-old African American man presented with a slowly enlarging asymptomatic nodule on his midback treated 9 months previously with liquid nitrogen and again 1 month later. The patient had no history of endocrine abnormalities, lentigines, blue nevi or other neoplasms. On examination, a soft 1-cm well-demarcated pedunculated nodule was noted (Fig. 1).

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Histological evaluation of a shave biopsy revealed a dome-shaped lesion containing stellate and multinucleated fibroblasts admixed with numerous small thin-walled blood vessels surrounded by pools of mucin that formed cleft-like spaces (Fig. 2). Lymphocytes were scattered throughout the lesion, and centrally, an abortive follicular structure was appreciated. There was no evidence of epidermal effacement, dermal fibrosis or neovascularization suggestive of the prior treatment. Immunohistochemical stains showed strong labeling of the stellate cells with CD68 and factor XIIIa, but CD34 only labeled the blood vessels (Fig. 3A–C). To date, there is no evidence of recurrence after complete excision.

Discussion

Allan depicted SA as a specific entity in 1988; however, similar tumors had been previously described, albeit by different names, at least 30 years earlier.³ Originally proposed to be synonymous with cutaneous focal mucinosis, myxoid perifollicular fibroma, fibrofolliculomas, trichofolliculomas and trichodiscomas, Calonje et al.³ disagreed with this hypothesis concluding that SA was a distinct entity.²

SA has a slight male predilection, typically presenting between the ages of 20–40 years. Although they preferentially occur on the trunk, extremities and head and neck regions, they can arise in a variety of



Fig. 1. One centimeter pedunculated nodule on back.

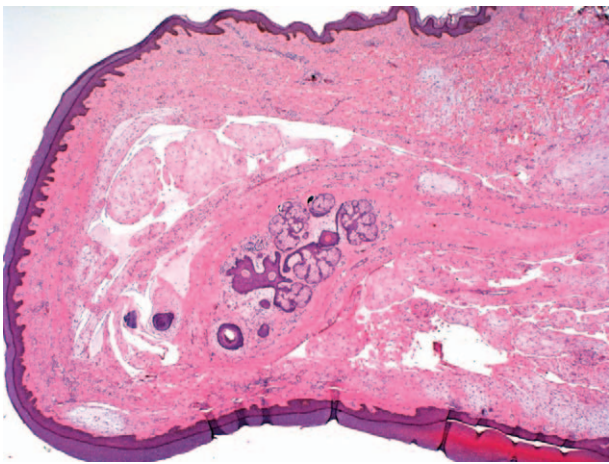


Fig. 2. Dome-shaped lesion composed of increased numbers of spindle cells and multinucleated giant cells associated with increased numbers of thin-walled vessels and copious amounts of mucin (×2).

anatomic locations.¹⁻⁵ The majority of lesions occur in isolation; however, multiple SA, especially those located on the external ear, are considered pathognomonic for Carney complex, an autosomal dominant

disorder associated with multiple myxomas (cardiac, cutaneous and mammary), blue nevi, lentigines, psammomatous melanotic schwannoma and endocrine overactivity.¹⁻⁴

Although the exact incidence is unknown, in the index article, only 27 solitary tumors were identified among 4500 consults seen over 23 years.² To evaluate the incidence at our institution, a Systematized Nomenclature of Medicine (SNOMED) word search engine using the descriptors myxoma, angiomyxoma and superficial cutaneous angiomyxoma was performed within the computerized archives of the

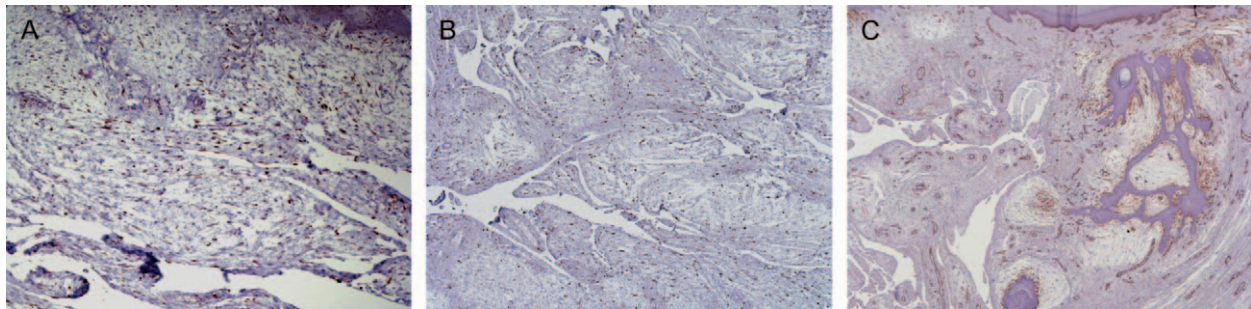


Fig. 3. A–C) Intense labeling of spindle and stellate cells with CD68 (A) and factor XIIIa (B), staining of vasculature with CD34 (C).

Table 1. Clinical features of various soft tissue myxomas*

	Age (years)	Gender	Most common location	Size	Associated conditions
Superficial angiomyxoma	20–40	64% males	37% trunk, 33% lower extremities	Most < 5 cm	Carney's complex
Intramuscular myxoma	20–89	66% females	50% thigh	2–20 cm	Mazabraud and McCune-Albright's syndromes
Juxta-articular myxoma	16–83	72% males	88% knee	0.6–12 cm	Degenerative joint disease
Aggressive angiomyxoma	20–50	95% females	Pelvic and perineal regions	Most > 10 cm	None
Angiomyofibroblastoma	25–60	99% females	Pelvic and perineal regions	Most < 10 cm	None
Superficial acral fibromyxoma	14–72	68% males	Nail bed of the fingers and toes	0.6–5 cm	None
Nerve sheath myxoma	8–72	55% males	86% extremities	0.5–2.5 cm	None

*Mazabraud syndrome: multiple intramuscular myxomas and fibrous dysplasia of bone. McCune-Albright's syndrome: multiple intramuscular myxomas, polyostotic fibrous dysplasia, café au lait macules, precocious puberty and other endocrinopathies. Carney's complex: multiple myxomas (cardiac, cutaneous and mammary), blue nevi, lentigines, psammomatous melanotic schwannoma and endocrine over activity.

Table 2. Histological characteristics of various soft tissue myxomas

	Margin	Vascular pattern	Cellularity	Stroma	Staining characteristics	Recurrence rate
Superficial angiomyxoma	Poor to moderately circumscribed, multilobular	Scattered thin-walled vessels	Moderately cellular, bland spindled and stellate cells, variable inflammatory cell infiltrate	Abundant mucin with clefts. Up to 30% have an associated epithelial component	Vimentin; variable staining with CD34, factor XIIIa, SMA, MSA and S-100	20–30%
Intramuscular myxoma	Poorly circumscribed merges with surrounding muscle	Hypovascular variant; hypervascular variant	Hypocellular variant; hypercellular variant; bland spindle cells	Abundant mucin with cystic spaces. Hypercellular variant has strands of collagen	Vimentin; variable staining with actin, desmin, CD34	None
Juxta-articular myxoma	Poorly circumscribed infiltrates surrounding tissue	Focally vascular	Focally hypercellular, peripheral spindle cells with occasional atypical cells and mitoses	Abundant mucin, 89% of cases contain cystic spaces lined by fibrin or collagen	Vimentin; variable staining with actin, desmin, CD34	34%
Aggressive angiomyxoma	Infiltrative	Uniformly distributed medium-sized blood vessels often with prominent hyalinization	Low to moderately cellular, evenly distributed round, spindled or stellate cells	Loose myxoid to focally collagenous	Vimentin, desmin, SMA, MSA, estrogen and progesterone receptor	36–72%
Angiomyofibroblastoma	Well circumscribed	Abundant thin-walled blood vessels	Alternating hypercellular and hypocellular areas, perivascular condensations of spindled to epithelioid stromal cells	Collagenous to edematous with minimal mucin	Vimentin, desmin, CD34, estrogen and progesterone receptor	No recurrences reported, but rare cases of sarcomatous degeneration
Superficial acral fibromyxoma	Pushing to infiltrative	Mild to moderately accentuated vasculature	Moderately cellular, spindle and stellate cells with a storiform to fascicular pattern, variable mast cells	Myxoid to collagenous	CD34, EMA, CD99	Recurrence rare and primarily for incompletely excised lesions
Nerve sheath myxoma	Well circumscribed, multilobular	Hypovascular	Moderately cellular, spindled cells in fascicles and whorls	Nests of cells separated by collagenous bundles	S-100, EMA	47% if incompletely excised

EMA, epithelial membrane antigen; MSA, muscle-specific actin; SMA, smooth muscle actin.

Department of Pathology, Naval Medical Center San Diego. Over the past 10 years, approximately 180,000 surgical pathology cases were examined and only two cases, other than the one that is the subject of this article, were identified. Because our population is somewhat skewed, several other dermatopathologists at various institutes were queried, and SA represented approximately 0.0008% (14/1,680,000) of all specimens accessioned. Of the two soft tissue pathologists questioned, SA represented less than 0.3% (175/59,500) of all soft tissue neoplasms; therefore, SA is infrequently encountered in routine practice.

The differential diagnosis of SA is extensive; nevertheless, the most common benign neoplasms include intramuscular myxoma, juxta-articular myxoma, aggressive angiomyxoma, angiomyofibroblastoma, superficial acral fibromyxoma and nerve sheath myxoma.¹⁻⁵ While clinical and histological overlap exists among these entities, they are readily differentiated by characteristic clinicopathological findings (Tables 1 and 2).¹⁻¹⁶ Other benign entities with extensive mucinous stroma include ganglion cysts, focal mucinosis, myxoid neurofibroma and angio-myxolipoma.^{3,5} Lastly, a few malignant myxomatous tumors should be included in the differential diagnosis, particularly with large lesions that extend deep, namely myxofibrosarcoma, fibromyxoid sarcoma, myxoid liposarcoma, myxoid dermatofibrosarcoma protuberans and myxoid malignant peripheral nerve sheath tumor.^{1,3,5}

Almost all reported cases that have utilized immunohistochemical stains have been immunoreactive with vimentin and negative with desmin and cytokeratin.^{3,5,10-16} The staining results for CD34, smooth muscle actin, muscle-specific actin, S-100 and factor XIIIa have been more variable. The case presented herein had a slightly different immunophenotype compared with previously reported cases. Although the spindled and multinucleated cells strongly labeled with factor XIIIa, a finding reported in approximately 53% of cases that have utilized this stain,^{5,10,12,13,15} these cells failed to stain with CD34, which has been expressed in approximately 71% of cases.^{5,10-16} Furthermore, the lesion strongly expressed CD68, a finding that has not been previously reported. However, since the use of CD68 has been infrequently reported in the literature,^{12,13} the positive staining of the current case may not be unique and requires further investigation.

In conclusion, the features most useful to differentiate a SA from other myxoid tumors include its superficial location, lack of atypia, stromal inflammatory infiltrate and a frequent association with an entrapped epithelial component. It is essential that

dermatopathologist be aware of this entity to avoid confusion with more aggressive myxoid tumors.

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Note added after online publication: conflicts of interest

None declared.