Abstract

Superficial angiomyxoma (SA) is one of the superficially located myxoid soft tissue tumor. Because of the rarity of this tumor, there is paucity of fine needle aspiration cytology (FNAC) findings. Here, we present a case of SA in a 14-year-old girl who presented with a left leg swelling. The cytology of the lesion along with the histopathologic correlation is presented.

Keywords: Carney’s complex, myxoid tumor, superficial angiomyxoma

Introduction

Superficial angiomyxoma (SA), also known previously as cutaneous myxoma, is a very rare neoplasm commonly associated with the Carney’s complex. Allen et al. had first described it when they reported myxoid tumors of the dermis and subcutaneous region. Fine needle aspiration cytology (FNAC) of this rare tumor is thus limited. Here, we report a case of lower limb angiomyxoma in a young girl, its cytology, as well as the subsequent histopathological correlation.

Case Report

A 14-year-old girl presented to the surgical outpatient department with a swelling on the left leg upper medial aspect for the past 3 years. It was gradually progressing from the size of a peanut to 5 × 5 cm. There was no history of pain, redness, trauma, fracture, movement compromise, anorexia, and weight loss. On local examination, it presented as a 5 × 5 cm swelling over the left leg medial aspect; it was nontender, nonerythematous, freely mobile, and not attached to the overlying skin or underlying muscle. On radiological examination, X-ray showed a well-defined radio-opaque shadow in the posteromedial aspect of the upper left leg and not involving the bone. On magnetic resonance imaging (MRI), a capsulated smooth margined lesion measuring 5 × 2 × 0.3 cm involving the skin and subcutaneous planes at the site described above with no infiltration to the adjacent muscles or neurovascular bundles. The underlying muscles and bone were free.

The patient was referred for FNAC, which was conducted with a 22 Gauze needle under aseptic condition. The aspirate smears yielded abundant myxoid/fibrillary fragments. Entrapped in these fragments are plump spindle cells, with bland nucleoli and occasional prominent nucleoli in some of them. There was only mild pleomorphism and atypical mitosis or areas of necrosis were absent. Following possibilities were considered: (1) adnexal tumor with myxoid differentiation and (2) mesenchymal neoplasm with myxoid differentiation. Histopathological correlation was advised.

Subsequently, an excision specimen of the mass with the surrounding soft tissue was received. The specimen measured 7 × 6 × 2 cm grossly. Macroscopically, a myxoid area was identified measuring 4 × 2.5 × 2 cm. Microscopic examination revealed a sparse proliferation of spindle, stellate, and oval cells within a fibromyxoid stroma, with numerous arborizing blood vessels with focal lymphoplasmacytic as well as eosinophilic infiltrate with occasional neutrophils predominantly in the perivascular region. Only

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mild pleomorphism was noted and mitosis was rare; and was reaching the deeper dermis. The overlying skin, papillary dermis, and all the surgical margins were free of tumor.

On immunohistocytochemistry (IHC), the tumor cells were found to be positive for vimentin and CD34. Based on the immunohistomorphological findings, a final diagnosis of SA was rendered.

Retrospectively, cytology slides were reviewed with the final diagnosis in mind and cyto-histo correlation was established.

**DISCUSSION**

Allen *et al.* in 1988 described SA as a lesion commonly presenting in the head, neck, trunk, and genital region. In 1995, Francisco *et al.* first described the cytology of SA in a 46-year-old woman who presented with an areolar swelling. Since then, further cytological findings of SA is lacking. Most of the cases reports have been on histopathology with hemorrhagic/inconclusive FNAC.

SA should be differentiated from all the superficially located myxoid neoplasms, both malignant and benign lesions, which includes myxoid neurofibroma, dermal nerve sheath myxoma, aggressive angiomyxoma (AA), and low-grade myxofibrosarcoma. Myxoid neurofibroma is well-demarcated, can be solitary/multiple and can be variably sized. The cells are benign with wavy nuclei and intraleisonal neuritis with myxoid stroma. Dermal nerve sheath myxoma is well-demarcated and lobulated with fibrous tissue septation and has a myxoid stroma. The cells are benign, spindle-shaped arranged in fascicles and whorls with mild atypia and mitoses. The latter two entities have atypia, mitosis, and necrosis that differentiate them from the other benign lesion. AA is common among adult females, are exclusively present in genital or pelvic regions, are usually larger, solitary, and aggressive locally. AA microscopically is poorly delineated and usually infiltrative. It has a paucicellular myxoid stroma with many thin and thick-walled blood vessels with occasional hyalinization and foci of hemorrhage. Clusters and whorls of smooth muscle cells are seen surrounding the vessels. Mast cells are frequently present.

SA can be sporadic or be associated with Carney’s complex. The solitary cutaneous SA is of minor clinical importance. However, multiple lesions and comparable solitary lesions located in the breast or external ear may be associated with cardiac myxomas, pigmented skin lesions, and endocrine abnormalities as part of the Carney complex, which are inherited in an autosomal dominant manner.

Solitary SA have been reported without the association with carney’s complex, as Rosado *et al.* reported in the parotid, Kozo *et al.* in the inguinal region, and Mahendra *et al.* in the toe. Our case also had no other swellings besides the one with which the patient presented with; moreover, the swelling was not accompanied by noncutaneous myxomas, pigmentation of the skin or mucous membrane, or endocrine disorders.

SA is benign but local recurrence following surgical excision is common if the resection is marginal or incomplete. The recurrence rate is 30–40% and is due to inadequate resection, according to Allen *et al.* and Calonje *et al.* The patient has no known recurrence and is doing well as per the last follow up.

**Conclusions**

The presence of abundant myxoid stroma in FNAC smear with bland spindle out cells in the background with mild atypia and no mitosis and necrosis should prompt a diagnosis of SA. A wide local excision is the mainstay in the treatment of this disease because incomplete excision can lead to a recurrence. A cytologist should, thus, keep a differential of SA in patients...
presenting with solitary lesion with a predominant myxoid stroma and spindled out bland cells.

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**Conflicts of interest**
There are no conflicts of interest.

**REFERENCES**
1. Carney JA, Headington JT, Su WPD. Cutaneous myxomas: A major component of myxomas, spotty pigmentation & endocrine overactivity. Arch Dermatol 1986;122:790-8.
2. Allen PW, Dymock RB, MacCormac LB. Superficial angiomyxomas with and without epithelial components. Report of 30 tumors in 28 patients. Am J Surg Pathol 1988;12:519-30.
3. Izquierdo FM, Martin L, Burgos F, Lacruz C. Fine-needle aspiration cytology of superficial angiomyxoma (myxoid perifollicular fibroma): Report of a case. Diagn Cytopathol 1995;13:247-51.
4. Khadilkar UN, Khadilkar NP, Rao PS, Chakravorty S, Goel G. Superficial angiomyxoma of the external ear not associated with Carney’s complex: A case report. Kathmandu Univ Med J 2007;5:546-9.
5. Graadt van Roggen JF, Hogendoorn PCW, Fletche CDM. Myxoid tumours of soft tissue. Histopathology 1999;35:291-312.
6. Carney JA, Hruska LS, Beauchamp GD, Gordon H. Dominant inheritance of the complex of myxomas, spotty pigmentation, and endocrine overactivity. Mayo Clinic Proc 1986;61:165-72.
7. Carney JA, Toorkey BC. Myxoid fibroadenoma and allied conditions (myxomatosis) of the breast. A heritable disorder with special associations including cardiac and cutaneous myxomas. Am J Surg Pathol 1991;15:713-21.
8. Ferreiro JA, Carney JA. Myxomas of the external ear and their significance. Am J Surg Pathol 1994;18:274-80.
9. Rosado Rodríguez P, de Vicente JC, de Villalain L, Blanco V. Superficial angiomyxoma of the parotid region and review of literature. Acta Otorrinolaringol Esp 2012;63:147-9.
10. Yamamoto K, Kondo A, Iwashita K, Umezawa Y, Ohta Y, Matsuyama T, et al. A case of Superficial Angiomyxoma. Tokai J Exp Clin Med 2006;31:43-5.
11. Kura MM, Jindal SR. Solitary superficial acral angiomyxoma: An infrequently reported soft tissue tumor. Indian J Dermatol 2014;59:529.
12. Calonje E, Guerin D, MacCormac D, Fletcher CDM. Superficial Angiomyxoma. Am J Surg Pathol 1999;23:910-7.