

Aneurysmal Fibrous Histiocytoma: A rare variant

Sheethal Shanibi T M^{1*}, Vineetha. K.V¹, A.M Sahabudheen² and Vidyadhar Rao¹

¹Dept of Pathology, Kannur Medical College, Anjarakandy, Kannur, Kerala, India

²Tely Hospital, Thalassery, Kannur, Kerala, India

ABSTRACT

Aneurysmal fibrous histiocytoma (AFH) is a rare variant of benign fibrous histiocytoma. This tumor accounts for less than 2% of all fibrous histiocytomas, 1.7% of all cutaneous fibrous histiocytoma and it is typically known to occur in the extremities of middle-aged patients.

A 23 year old male presented with slow growing, pigmented, nodular lesion on the left leg for the past 2 years. The swelling was initially non-tender, but there was a sudden increase in size associated with pain for the past 2 months. With a clinical suspicion of malignant melanoma an excision biopsy was performed and further evaluated by histopathology. The skin covered pigment lesion measuring 1.5x0.8x0.5cm showed fibrohistiocytic cellular proliferations containing blood filled spaces lacking endothelium on microscopy. Ancillary testing with bleaching, perls prussion blue reaction demonstrated the pigment as hemosiderin. Subsequently the lesion was negative for CD34 and positive for Factor XIIIa. Based on the histomorphology, ancillary testing and IHC, the diagnosis of Aneurysmal fibrous histiocytoma was made.

Aneurysmal fibrous histiocytoma can be a challenging diagnostic entity due to its close clinical resemblance to melanoma and hemangioma. Although the pathogenesis is unclear, it is believed to be caused by extravasation of erythrocytes from capillaries leading to formation of the cystic blood spaces.

Keywords: Benign fibrous histiocytoma, Dermatofibroma, Cutaneous Histiocytoma, Aneurysmal Variant

Introduction

Aneurysmal fibrous histiocytoma (AFH) is a rare variant of benign fibrous histiocytoma (FH), posing diagnostic difficulty due to morphological resemblance to malignant melanoma, angiomatoid fibrous histiocytoma, kaposi sarcoma and angiosarcoma. Since AFH has a benign course but higher recurrence rate than FH⁽¹⁾ it is essential to differentiate it from the other close differentials. This tumor accounts for less than 2% of all fibrous histiocytomas, and it is typically known to occur in the extremities of middle-aged patients⁽²⁾⁽³⁾. On histopathology, in addition to the typical features of a dermatofibroma, it contains large cleft-like or cavernous blood-filled spaces with numerous hemosiderin pigments. Atrophic dermatofibroma is also a rare variant of dermatofibroma, and the combination of aneurysmal and atrophic features is a much more rare⁽⁴⁾. We report a case of aneurysmal benign fibrous histiocytoma with atrophic features in a 23-year-old male who presented with a pigmented papillomatous lesion in the leg.

Case Report

A 23 year old male presented with slow growing pigmented papillomatous lesion on the leg for the past 2 years. The swelling was non tender, but there was a sudden increase in size for the past 2 months associated with pain. There

was no history of trauma, previous surgery or injection to the involved site. An excision biopsy was done, and we received a skin covered tissue whole ms 1.5x0.8x0.5cm. Skin shows a symmetrical pigmented lesion ms. 0.8cm in diameter. (Fig.1.) C/S shows grey brown areas in the superficial dermis with grey white area surrounding it.

On microscopy, section studied showed skin with epidermis, dermis and subcutaneous tissue. The epidermis was atrophic. Upper dermis showed a well circumscribed lesion exhibiting focal storiform pattern (Fig.4) and composed of proliferating spindle shaped cells and macrophages containing hemosiderin pigments (Fig.2). Touton giant cells with hemosiderin pigment were also seen. (Fig.3). Centre of lesion showed large blood filled cavernous spaces that lacks endothelial lining (Fig. 2).Areas of collagen wrapping seen on the sides of lesion (Fig.5). Deeper dermis and subcutaneous tissue appears normal.

Bleaching was done, that retained the pigment and ruled out the possibility of melanocytic lesion (Fig.6).Perls stain confirmed the brown pigment as hemosiderin (Fig.7)

Immunohistochemistry for CD34 confirmed the absence of an endothelial lining around the small blood filled spaces (Fig.8) and ruled out the possibility of dermato

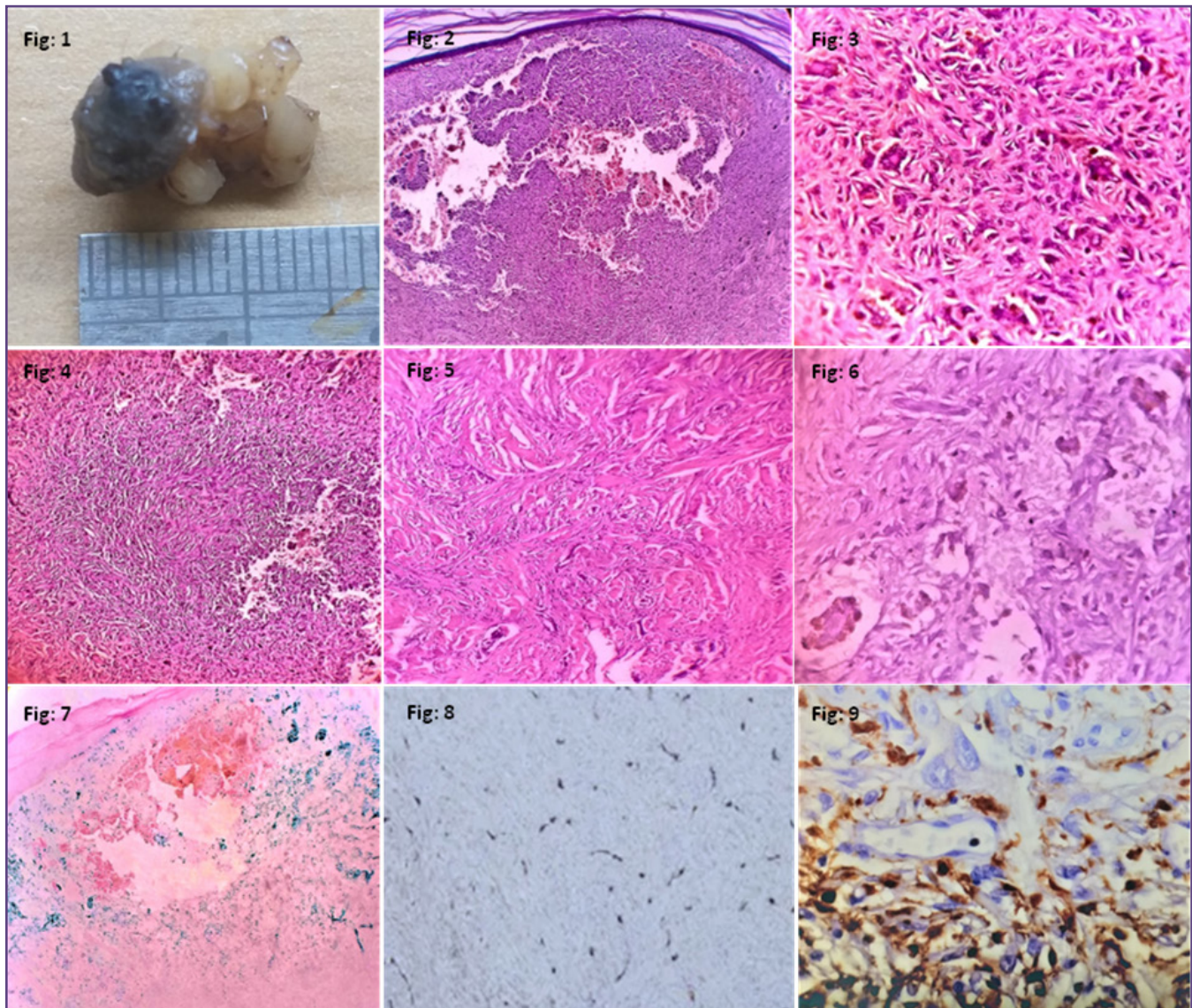


Fig. 1: Macroscopic appearance of the lesion.**Fig.2:** Dermal lesion showing cavernous space with hemosiderin.**Fig.3:** Touton giant cells.**Fig.4:** Storiform pattern (10x). **Fig. 5:** Collagen wrapping at periphery of the lesion (10x). **Fig.6:** Retained hemosiderin after bleaching. **Fig.7:** Perls stain showing hemosiderin pigments.**Fig.8:** IHC showing CD34 negativity. **Fig. 9:** IHC factor XIIIa positivity.

fibrosarcoma protuberans. Hence a final diagnosis of Dermatofibroma – Aneurysmal Variant was given.

Discussion

BFH usually presents as single or multiple firm reddish brown nodules located on the lower extremities of young adults. Histopathological findings show a tumor mass in the dermis that is composed of fibroblast-like spindle cells and histiocytes.⁽⁴⁾ AFH is one of the rarest variants of cutaneous fibrous histiocytoma with reported incidence of 1.7% of all cutaneous fibrous histiocytoma⁽⁵⁾. It was first described by Santa Cruz and Kyriakos in 1981⁽⁴⁾⁽⁶⁾. AFH

is characterized by fibrohistiocytic cellular proliferations containing blood filled spaces lacking endothelium. Although the pathogenesis is unknown, it may be caused by extravasation of erythrocytes from capillaries leading to the cystic blood spaces⁽⁷⁾⁽⁸⁾. Clinically, this may be confused with malignant melanoma or haemangioma. Histologically, there is frequent confusion with other vascular tumors⁽⁷⁾ Among the differential diagnosis of aneurysmal fibrous histiocytoma, malignant melanoma, Kaposi's sarcoma, spindle cell hemangioma, angiosarcoma, and angiomatoid fibrous histiocytoma should be considered⁽³⁾ ⁽⁹⁾. Kaposi Sarcoma is clinically found to occur in elderly

men and affects the lower extremities. It presents as flat, pink patches which latter acquires blue-violet papular appearance. Microscopically, the vascular spaces are lined by endothelial cells positive for CD34, a major point that differentiates Kaposi Sarcoma from AFH⁽⁶⁾. Cutaneous angiosarcoma usually occurs on the face or scalp of elderly men and contains atypical endothelial cells that separate collagen bundles⁽⁷⁾. The present case showed CD34 negativity of cells lining the vascular spaces ruling out the possibility of Kaposi's sarcoma and cutaneous angiosarcoma. Bleaching done this retains the pigment and ruled out malignant melanoma.

Conclusion

It is important to distinguish this benign lesion from similar appearing malignant skin lesions. As a rare case, aneurysmal fibrous histiocytomas have the potential to interfere with other malignant mesenchymal neoplasms in the stage of diagnosis and are skin lesions with high recurrence rates, which should be kept in mind during reporting. The confirmed diagnosis for AFH should depend on histopathology and immunohistochemistry. Surgical resection is the best treatment for AFH.

Acknowledgements

Dr. Dimple ET (Resident in Pathology), Greeshma K (Histopathology Technician)

Funding

Kannur Medical college

Competing Interests

The authors declare that they have no competing interests

Reference

1. Gao M, Lin X. Clinic Pathological Study of Aneurysmal Fibrous Histiocytoma. *J Biosci Med.* 2019;07(05):1–5.
2. Chaithanya K CUD. Utility of fine needle aspiration cytology in diagnosing soft tissue tumors- experience in our institution. *J Diagnostic Pathol Oncol.* 2017;2(4):76–81.
3. Akpınar M, Yigit O, Durma YM, Sekercan O, Havare SB. Aneurysmal Fibrous Histiocytoma in Nasal Vestibule Mucosa. *Istanbul Med J.* 2015;16(2):80–2.
4. Shin JW, Park HS, Kim BK, Kim YA, Kim MG, Won CH, et al. Aneurysmal benign fibrous histiocytoma with atrophic features. *Ann Dermatol.* 2009;21(1):42–5.
5. Antony A, Kiran CM, Phansalkar M, Jothi C, Jayakar J. Aneurysmal variant of fibrous histiocytoma- A rare entity known for recurrence. *J Clin Diagnostic Res.* 2017;11(6):ED08–9.
6. Santa Cruz DJ KMA (“angiomatoid”), 1981;47:2053-fibrous histiocytoma of the skin. C, 2061. No Title. 1981;
7. Sheehan KM, Leader MB, Sexton S, Cunningham F, Leen E. Recurrent aneurysmal fibrous histiocytoma. *J Clin Pathol.* 2004;57(3):312–3.
8. McKenna DB, Kavanagh GM, McLaren KM et al. A fibrous, Acad histiocytoma: an unusual variant of cutaneous fibrous histiocytoma. *JE,* 1999;12:238–40. DV. No Title.
9. Calonje E FC. No Title. Aneurysmal benign fibrous histiocytoma Clin Anal 40 cases a tumour Freq misdiagnosed as a Vasc neoplasm. 1995;26:323-31.

*Corresponding author:

Dr. Sheethal Shanibi T M, Associate professor, Dept of Pathology, Kannur Medical College, Anjarakandy-670612, Kannur, Kerala, India

Phone: +91 7012509480

Email: centlab.kmch@gmail.com

Financial or other Competing Interests: None.