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Pathology

ATYPICAL ANGIOMATOID FIBROUS HISTIOCYTOMA CLINICAL MIMICKER OF NODULAR MALIGNANT MELANOMA

Dr. Nechal Kaur Post graduate student Department of Pathology, Maharishi Markandeshwar Medical College and hospital, Kumarhatti, Solan, Himachal Pradesh.

Dr. Neelam Gupta* Professor and Head Department of Pathology, Maharishi Markandeshwar Medical College and hospital, Kumarhatti, Solan, Himachal Pradesh. *Corresponding Author

Dr. Vikas Dubey Assistant Professor Department of Pathology, Maharishi Markandeshwar Medical College and hospital, Kumarhatti, Solan, Himachal Pradesh.

ABSTRACT Angiomatoid fibrous histiocytoma (AFH) is a rare soft tissue tumor of low malignant potential. Because of its rarity, nonspecific radiological and diverse pathological findings, it is often clinically misdiagnosed. However only few clinical reports have described this tumor. We are presenting a case where lesion is present on the nipple areolar complex.

KEYWORDS : Angiomatoid fibrous histiocytoma, Breast, Atypical.

INTRODUCTION

Angiomatoid fibrous histiocytoma (AFH) is a rare soft tissue tumor that is most commonly reported to arise in the subcutaneous tissues of the upper extremities in adolescents and young adults.[1] The main presentation is painless soft tissue mass, accompanied by systemic symptoms, like anemia, weight loss, and fever, although atypical presentations have been reported. [2] Clinically and radiographically the lesion is easily confused with a hematoma, soft tissue hemangioma, or malignant fibrous histiocytoma.[3] While the lesion is rare, due to the potential for local recurrence and metastasis. The entity was first described by Enzinger in 1979 and was termed Angiomatoid "Malignant" Fibrous Histiocytoma. Due to low reported recurrence rates (2-10% locally) and even lower reported risk of metastases (< 1% of cases), its name was changed to AFH, and it is now considered a malignancy with intermediate risk.[1] Angiomatoid fibrous histiocytoma is associated with 3 characteristic gene fusions, EWSR1-CREB1 and EWSR1-ATF1 and rarely FUS-ATF1.[4]

Case Report

A 31 year old female presented to Surgery OPD with chief complaint of breast lesion on left breast since 6 years, bloody discharge since 2-3 years which is on and off. On left breast examination black papillomatous lesion measuring 2cm above nipple aerolar complex was noted. Mild tenderness present. No lymphadenopathy was noted. Excision of breast lesion with 1 cm margins was done and specimen was sent for histopathological examination.

Grossly received well oriented skin covered tissue bit measuring 2.5x2x0.8cm with a black coloured nodular lesion measuring 0.5x0.5cm in the centre of the skin. Cut surface revealed multiple, black coloured tumors (maximum depth 0.8cm), reaching upto the base. Grossly, all margins appear free. On microscopic examination sections shows keratinized stratified squamous hyperplastic epithelium. Dermis shows an infiltrative tumour comprising of plump spindle shaped cells arranged in tight storiform pattern showing mild nuclear atypia with small inconspicuous nucleoli. Also seen are large blood filled cystic spaces, extravasation of RBCs and slit like spaces lined by spindle cells having RBCs in lumen. Branched staghorn type blood vessels seen. In between the spindle cells large aggregates of hemosiderin laden macrophages, abundant lymphoplasmacytic infiltrate and occasional lymphoid follicles formation in the periphery. Dermal appendages are entrapped by the tumour cells. Tumour is abutting overlying epithelium and reaching upto the base. Tumour shows honeycoombing of subcutaneous adipose tissue. Mitoses 3-4 per 10 HPF including occasional atypical mitoses. All the margins (superior, inferior, medial, lateral) are free. Base is involved by the tumour. No lymphovascular/perineural invasion noted. Perl's stain was done which was positive for hemosiderin. Histopathological features are consistent with Atypical angiomatoid fibrous histiocytoma - Nipple areolar complex. By Immunohistochemistry, the tumor show positivity for Vimentin, CD 68, MIC-2, SMA, while negative for CK, CD 34, S-100 protein, SOX-10, p53, ER, C-KIT and Calponin which was conducted in higher centre.

DISCUSSION

AFH was first described by Enzinger and termed as angiomatoid malignant fibrous histiocytoma. Because of their relatively rarity of metastasis and the overall excellent clinical course, it was classified as an intermediate tumour in WHO classification of tumours of soft tissue and bone. AFH usually occurs in the soft tissue forming a well circumscribed subcutaneous nodule on the extremities, head and neck and trunk. Recently, AFH has been reported in unusual sites including the lung, mediastinum, vulva, retroperitoneum, ovary, pulmonary artery, kidney, and intracranial.[5] AFH is a neoplasm that most commonly affects children and young adults, with a median age of 14 years. The tumor is rare, accounting for approximately 0.3% of all soft tissue neoplasms, incidence may be underestimated due to overlapping histopathological findings. Presentation usually involves a painless, slow growing mass within the deep dermis and subcutis. It most commonly arises in sites of normal lymphoid tissue such as the antecubital fossa, axilla, inguinal and supraclavicular regions.[3] The characteristic histological features of AFH have been well described. These features include the following: (i) multinodular growth of myoid spindled or histiocytoid cells with a distinctive syncytial appearance, (ii) pseudoangiomatous spaces filled with blood and surrounded by tumor cells, (iii) a thick fibrous pseudocapsule with prominent hemosiderin deposition, and (iv) peritumoral lymphoplasmacytic cuffing with occasional germinal center formation.[6] Furthermore, AFH sometimes has variant histological patterns. Unusual morphologic features have included sclerosis, nuclear grooving, clear cell change, rhabdomyoblast-like cells, groups of small cells with scanty cytoplasm reminiscent of Ewing sarcomas, a perineurioma-like pattern, a pulmonary edema-like pattern, a schwannoma-like pattern with nuclear palisading and hyalinized vessels, and reticular patterns of cells in myxoid stroma.[7] Angiomatoid fibrous histiocytoma can occur in diverse anatomical locations, and additional new sites of involvement will almost certainly be recognized in future. We speculate that angiomatoid fibrous histiocytoma may be an under recognized tumor outside somatic soft tissues, and bona fide cases have probably been subsumed among cases diagnosed as inflammatory myofibroblastic tumor, follicular dendritic cell sarcoma, poorly differentiated carcinoma or meningioma. Increased awareness of the morphological characteristics of angiomatoid fibrous histiocytoma (in particular, the peritumoral lymphoid cuff) and its potential occurrence in unusual sites will aid in its recognition.[8] Furthermore, extended resection of AFH is recommended because the tumor is considered intermediate.[9]

CONCLUSION

Classical morphologic features of AFH were present in this case and are the most important criteria to correctly diagnose AFH. Immunohistochemical studies were also done to confirm the diagnosis.

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Conflict Of Interest

The authors declare that they have no conflict of interest.

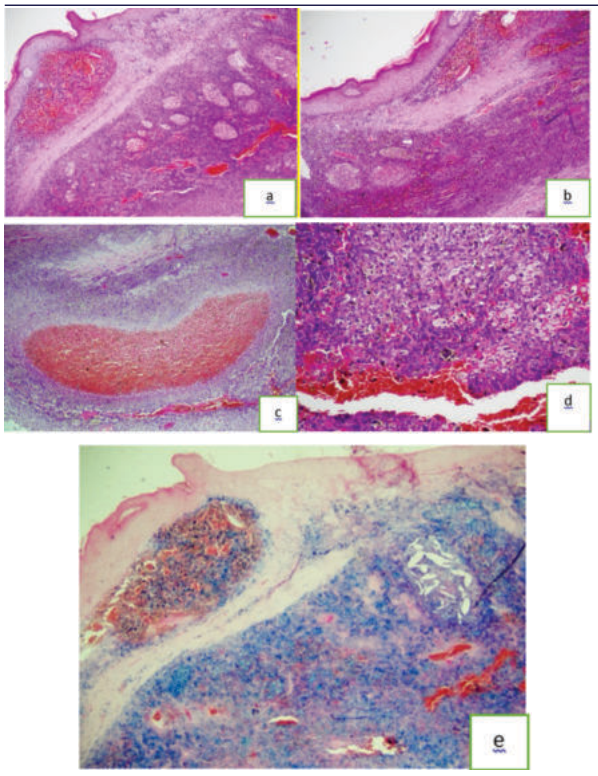

Figure 1- a), Low power view showing keratinized stratified squamous hyperplastic epithelium and dermis shows an infiltrative tumour. b),c) large blood filled cystic spaces, and extravasation of RBCs. d) large aggregates of hemosiderin laden macrophages. e) Perl's stain positive for hemosiderin.

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