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Case Report

Benign Fibrous Histiocytoma - Aneurysmal Variant as a Differential Diagnosis for Soft-Tissue Sarcoma, in a Physically Challenged and Mentally Retarded Child: Case Report

Avinassh Tippani[1], Bachu Brahmani[2]

[1]M.S (Gen), DNB-Surgical Oncology-NIMS, PDCC Surgical Gastroenterology-SVIMS, FIAGES, EFIAGES, FMAS, FAIS, Ex-Assistant Professor, Department of General Surgery, Surgical Oncologist – Gastro Surgeon, [2]Ex-Assistant Professor, Department of Pharm. D, Balaji Institute of Pharmaceutical Sciences, Sai Shree Hospitals Cancer and Surgical Gastro Centre, Warangal, Telangana, India

Abstract

Santa Cruz and Kyriakos described a special variant of cutaneous fibrous histiocytoma (FH) which was characterized by large, blood-filled cystic spaces, and diffuse hemosiderin deposits. They proposed the term aneurysmal (angiomatoid) FH of the skin. We report and discuss a case of benign FH - aneurysmal variant, in a female patient aged 17 years who is physically challenged and mentally retarded visited our hospital.

Key words: Aneurysmal variant, Angiomatoid, Benign fibrous histiocytoma, Soft-tissue tumor

INTRODUCTION

Fibrous histiocytoma (FH) is a benign and often superficial subcutaneous proliferation of oval cells resembling histiocytes and spindle shaped cells resembling fibroblasts and myofibroblasts. The clinicopathological variants of FH include cellular FH, aneurysmal FH, atypical FH, epitheliloid FH, and atropic FH.

Aneurysmal FH (AFH) is a rare and distinct variant of benign FH (BFH) with a reported incidence of 1.7% of all BFH. BFH is also known as dermatofibroma which usually grows rapidly and may attain a very large size.[1-3] It shows extensive hemorrhage, with prominent cavernous such as pseudovascular spaces that are not lined by endothelial cells. It affects mostly children and young adults with median age of 14 years. The tumor is rare, accounting for approximately 0.3% of all soft-tissue neoplasms.[4] It arises in sites of normal lymphoid tissue such as antecubital fossa, axilla, inguinal, and supraclavicular regions. AFH has higher risk of local recurrence compared with dermatofibroma.

Clinically, AFH presents as a solitary polypoid, blue to brown cystic lesion most commonly on the extremities of young to middle aged adults. The nodule may be associated with rapid growth due to sudden hemorrhage with in the lesion leasing to suspicion of malignancy and may be associated with pain.[3-5]

The diagnosis of AFH is made based on histopathology and immunohistology. Immunohistological staining can be performed to differentiate AFH from other similar appearing lesions. This can be done with the S-100 stain for sarcoma, neurofibroma, and melanoma. Melan-A, which is less sensitive than S-100, but is more specific can also be used for melanoma.[6,7]

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CASE SUMMARY AND DISCUSSION

A 17-year-old female patient who is bedridden, mentally retarded, and physically challenged visited our hospital with complaint of swelling over left forearm which was ulceroproliferative with bleeding and foul smell discharge.

Corresponding Author: Dr. Avinassh Tippani, Sai Shree Hospitals Cancer and Surgical Gastro Centre, Warangal, Telangana, India.

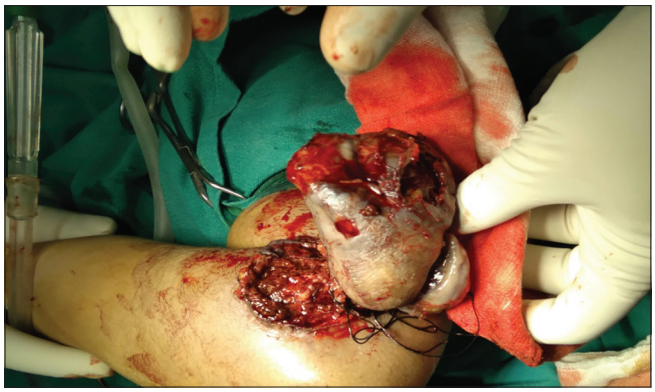

Figure 1: Swelling over left forearm which is ulceroproliferative with bleeding

Figure 2: Before the closure of the wound

Figure 3: Wide local excision of the mass

Figure 1 show in the hematology reports revealed anisocytosis-anemia. Biochemistry reports reveal decrease in sodium, albumin, and globulin proteins. Ultrasound reveals mild splenomegaly. Patient is further evaluated for soft-tissue sarcoma of the left elbow. Incision biopsy of forearm lesion suggested Spindle Cell Neoplasm. Immunohistochemistry findings showed B cell lymphoma (Bcl)2 positive, cluster of differentiation (CD)99 positive, CD34 highlight blood vessels, negative in tumor tissue, S100 negative, smooth muscle antibody-negative, and Ki67-10-15%. In view of

Figure 4: After the closure of wound

Figure 5: Post-operative image after the closure of wound with specimen aside

Bc12, CD99 positivity, and CD34 negativity, possibility of Synovial sarcoma to be considered. FDG PET-CT was done for disease staging, which was normal.

Wide local excision was done under high risk and informed consent was taken from patient attenders as the patient was physically and mentally challenged and bed ridden. Regional block anesthesia was given as the patient was not fit for general anesthesia. Operative and post operative images are shown in Figures 2-5.

Histopathology Findings

On gross examination mass contains skin covered soft tissue altogether measuring 6.5 × 5.3 × 2.4 cms. External examination shows ulceration. Cut surface shows grey white areas with extensive hemorrhagic areas. Microscopic examination shows skin with epidermis lined by squamous epithelium which shows elongated hyper pigmented rete ridges with focal ulceration. The dermis shows a lesion comprised of plump spindled to rounded cells dispersed among collagen bundles. The nuclei of these cells are rounded to elongated and vesicular. At some places, storiform pattern is seen. At the periphery, collagen is typically trapped, forming

collagen balls. Large blood filled spaces are seen. The lesion is extending into subcutaneous tissue with focal necrosis. Skin margin and posterior margin are involved by tumor. All these features suggest BFH and aneurysmal variant.

CONCLUSION

BFH should be a differential diagnosis while evaluating soft-tissue swellings where a major surgical amputation could be avoided in selected cases with poor performance status and at areas of critical importance such as joints. In our patient, incision biopsy was suggestive of spindle cell neoplasm but final diagnosis after wide local excision showed BFH – aneurysmal variant. In our patient with poor performance status, we have avoided major surgery where patient was unfit for general anesthesia.

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