Aneurysmal fibrous histiocytoma

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ABSTRACT

Aneurysmal fibrous histiocytoma is rare clinicopathological variant of Cutaneous Fibrous Histiocytoma. The clinical manifestation of Aneurysmal Fibrous Histiocytoma is often confusing to distinguish from other skin lesions. Most of the cases showed rapid increase in size or a history of recurrence, however histologically all are almost similar. Rarity of aneurysmal fibrous histiocytoma and high numbers of recurrence rate poses a big diagnostic challenge. Late treatment will result in a decrease until loss of function of the affected region. In this article, author reported the case of aneurysmal fibrous histiocytoma of the hand in 7 years old girl with restricted at 2nd-3rd metacarpophalangeal joints. The patient had undergone a series of investigations until finally a wide excision was carried out. Excision tissue was performed CD 68 and CD 34 immunocytochemical smear to establish the diagnosis. It was not simple to make diagnosis aneurysmal fibrous histiocytoma. While it is benign, the lesion can appear malignant, and one should consider an excisional biopsy to rule out malignant conditions. The diagnosis had to be confirmed by histopathological and performed immunocytochemical smear. It was often necessary to take aggressive actions with wide excision and reconstruction.

Keywords: Aneurysmal, Fibrous histiocytoma, Immunocytochemical, Metacarpophalangeal joints, Skin lesions

INTRODUCTION

Aneurysmal fibrous histiocytoma is a rare variant of cutaneous fibrous histiocytoma that results from blood vessel proliferation and haemorrhage into a fibrous histiocytoma.1,2 This is an uncommon lesion. Aneurysmal fibrous histiocytoma (AFH) is reported to have 1.7% incidence of all known fibrous histiocytoma. AFH is a neoplasm that most commonly affects children and young adults. The tumor is rare, accounting for approximately 0.3% of all soft tissue neoplasms, however, the incidence may be underestimated due to overlapping histopathological findings.3-5 The majority of cases occur in the extremities, although cases have been reported in the head and neck region (10%) and trunk.2,5

Clinically, a cutaneous fibrous histiocytoma is described as a typically dense, dome-shaped papule that ranges from a few millimeters to 5 cm in size. Clinicians may describe the lesion as adhered to subcutaneous tissue and pinching a fibrous histiocytoma can produce a downward movement of the papule, called “dimple sign”. Numerous non-classical variants exist besides aneurysmal, namely, atypical, cellular, clear cell, myxoid and palisading. These varying presentations can make the diagnosis of cutaneous fibrous histiocytoma difficult.6,7
CASE REPORT

A 7-year-old girl patient presented us with complaints of pain and swelling over the palmar aspect of the left thumb since 8 months. Swelling was insidious in onset, initial size was like peanut which gradually progressed, movements are completely restricted at 2nd-3rd metacarpophalangeal joints. The gross examination of the excised lesion revealed a skin covered tissue measuring 5 x 4 x 1 cm with a central raised nodular polypoid lesion measuring 3 x 2 x 3.5 cm (Figure 1).

Figure 1: Clinical appearance presents a solitary polypoid, blue to brown cystic lesion.

It slowly grew for the first two months and then rapidly enlarged in size over the next six months. Tissue excised was dark brown with focal areas of haemorrhage (Figure 2).

Figure 2: Cut surface dark brown with size 3 cm x 2 cm x 3.5 cm.

Imaging showed swelling of cystic, multiloculated, fluid level, hipointens without bony involvement. Histopathological detailing was done with Fine Needle Aspiration Biopsy (FNAB) which reported features suggestive of giant cell tumor which is consisted of round-oval nucleated histiocytes cells (Figure 3).

Excision tissue shows vascularized proliferative cells at the edge of the capillary with oval nucleus, without anaplastic cells. Immunocytochemical smear CD 68 revealed histiocytes with scattered giants' cells (Figure 4).

Figure 3: Hematoxylin and Eosin stain showed small vascular space surrounded by tumor cells with round-oval nucleated histiocytes cells.

Figure 4: Immunocytochemical stain CD 68 revealed histiocytes with scattered giants cells, numerous extravasated erythrocytes, and large blood filled cavernous spaces.

Immunocytochemical for CD34 showed negative staining for the cells surrounding the vascular spaces ruling out the possibility of endothelial cells (Figure 5).

Figure 5: Immunocytochemical stain CD 34 showed negative staining for the cells surrounding the
vascular spaces ruling out the possibility of endothelial cells.

Patient has been performed wide excision and flap (Figure 6,7). Observation for 1.5 months shows good clinical. The flap is viable and movement of metacarpophalangeal joints going better.

Microscopically, AFH lesions are located entirely within the dermis. They are highly cellular lesions composed of fibroblastic and histiocytic cells arranged in a storiform pattern. All AFH lesions contained blood filled spaces that vary from narrow slit-like cracks to large cavernous channels. These spaces are not lined by endothelial cells but are lined and surrounded by fibroblasts and histiocytes. Multinucleated giant cells, foamy cells and haemosiderinladen macrophages are often seen at the margin of the lesion. Any atypia noted in AFH, can raise the doubt of malignancy. All these features were noted in the present case.2,3,6

Aneurysmal fibrous histiocytoma has a good prognosis, but its recurrence rate is up to 19%. This recurrence rate is significantly higher than with common fibrous histiocytoma, which recurs in less than 2% of cases. Most likely this is due to an incomplete removal of the tumor, given its large size, and not a biological component. Regular reevaluations are thus recommended to ensure that the aneurysmal fibrous histiocytoma does not recur.6,7

CONCLUSION

Aneurysmal fibrous histiocytoma is a rare variant of fibrous histiocytoma. While it is benign, the lesion can appear malignant, and one should consider an excisional biopsy to rule out malignant conditions. There are lots of obscurities about this case, so more prospective well-designed studies including large number of cases are necessary to determine the prognostic factors, recurrence rate and proper treatment protocol for each individual patient. Appropriate and prompt action is required in such cases.

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