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Case Report

Benign cutaneous ancient schwannoma masquerading as well differentiated nerve sheath tumor: a case report

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ABSTRACT

Ancient schwannomas are a rare entity of benign cutaneous nerve sheath tumor in patients without von Recklinghausen disease. Head and neck, mediastinal, retroperitoneal ancient schwannomas have been reported and has high degree for malignant transformation. Ancient schwannoma of the peripheries have been reported in only few cases and in hand only two cases have been reported. We present this case for its rarity and the diagnostic dilemma occurring with FNAC. Thus these tumors if present in periphery and in non-neurofibromatosis patient can masquerade as malignant tumors but still can safely are treated with excision biopsy alone and with almost nil recurrence.

Keywords: Ancient schwannoma, Nerve sheath tumor, Malignant transformation

INTRODUCTION

Nerve sheath tumors arise from the nerve sheath cells or the schwann cells, most of them are benign. Peripheral nerve sheath tumors occur most frequently in setting of neurofibromatosis, most often in face and chest. Peripheral nerve sheath tumors of extremities are rare. Here we present an interesting case of benign shwannoma masquerading as well differentiated malignant tumor.

CASE REPORT

A 66 year old male, farmer by occupation presented to us with a swelling in the left hand along the medial edge medial to hypothenar eminence for the past 2 years. The swelling initially started as a painless swelling and slowly progressed to attain the present size. The swelling was not associated with pain, distal numbness. No history of trauma preceeding to the onset. On examination the swelling was oblong ovoid shaped non tender on the medial aspect of the left hand along the medial edge, at the junction of palmar and dorsal aspect, medial to hypothenar eminence, fifth metacarpal and extending distally up to the base of the proximal phalanx of the little finger on its medial aspect. Swelling attached to skin firm, mobile with the skin, not fixed to deeper structures, no distal paresthesia or tinnel sign was noted. Surface had a 0.5×0.5 shaped ulcer with minimal slough and no evidence of foul smelling discharge or bloody discharge noted. A clinical diagnosis of skin adnexal tumor was made, with advanced age, pain less swelling, superficial ulceration malignant skin adnexal tumor was to be primarily ruled out.

Ultrasonogram of the local area showed a well-defined solid tumor with decreased echogenicity in the central area that extended up to the superficial ulcer, vascularity was diffuse heterogeneous except the central hypoechoic area. A magnetic resonance imaging with contrast was done which showed a T1 isointense / T2 / STIR hyper intense well defined lesion in the subcutaneous plane with irregularity in the surface suggestive of ulceration

was noted, features suggestive of dermoid, and was advised to correlate with FNAC.

Fine needle aspiration cytology was done which showed plenty of spindle cells embedded in a fibrillary matrix with moderate nuclear atypia and pleomorphisms, suggestive of well differentiated spindle cell tumor, malignant nerve sheath tumor or vascular tumor to be considered and was suggested biopsy for further confirmation.

With diagnostic dilemma, patient was taken up for excision biopsy with plan to revise for wide local excision if biopsy proves to be malignant. Under wrist block an elliptical incision was made including the ulcerated skin and tumor was excised in Toto along with ulcerated skin after ligation of the vascular pedicle. Primary closure of the defect was achieved. The excised specimen was firm grey tan on cut section with necrosis in the center. Patient was followed up and histopathology was obtained which showed benign neoplasm in the deep dermis covered with stratified squamous epithelium composed of spindle shaped cells arranged sheets and bundles and fascicles in hypo and hyper cellular zones with marked nuclear pleomorphisms with atypia without any mitosis, with verocay bodies thick walled hyalinised blood vessels, areas of hyalinization cyst formation large areas of hemorrhage suggestive of Ancient Schwannoma. Immunohistochemistry was done which showed S100 positivity confirming schwannoma. Being benign no further treatment was advised to the patient.

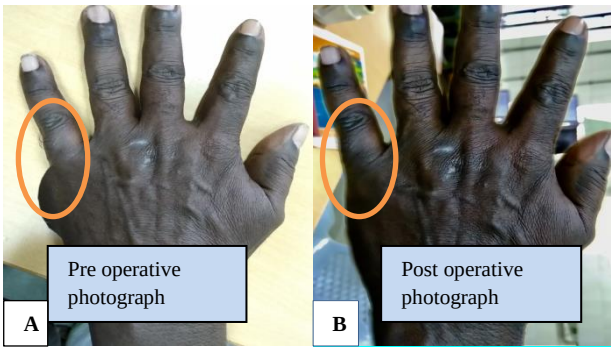

Figure 1: (A) Pre and (B) post operative images of the tumor.

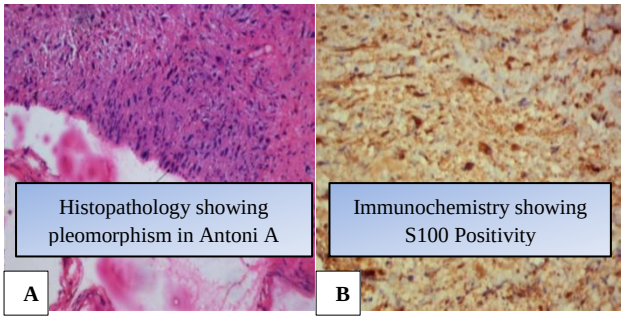

Figure 2: (A) Histopathology and (B) immune histochemistry images of the tumor.

Figure 3: Represents follow up age.

DISCUSSION

Schwannoma is a slow-growing benign tumor and may be a large tumor with degeneration, especially when the tumor is situated in the deep regions such as the mediastinum and retroperitoneum.[1] Schwannoma with pronounced degenerative changes is known as ancient schwannoma, which is a rare variant of schwannoma and is usually a deeply situated large mass of long duration [1,2] representing 0.8% of all soft-tissue tumors.[3] The term ‘‘ancient neurilemmoma’’ was first suggested by Ackerman and Taylor. In their first report of ancient schwannoma in 1951, 10 of 48 neurogenous tumors of the thorax reviewed could be distinguished from typical neurilemmoma by the pronounced histological degenerative changes they had undergone. As many studies have reported, ancient schwannoma tends to develop in areas deep inside the body where detection is relatively difficult (e.g., in the head and neck or thoracic region, retroperitoneum, or pelvic cavity).[4,5] In such cases, patients tend not to be aware of the schwannoma, resulting in delayed diagnosis and the discovery of ancient schwannoma after many years. Schwannoma, also termed neurilemmoma or neurinoma, is a benign nerve sheath tumor consisting of two components. Antoni A areas are more organized and are hyper cellular, and are composed of spindle cells arranged in short bundles or interlacing fascicles. Antoni B regions are hypo cellular, less organized and contain more myxoid, loosely arranged tissue, with high water content. These components are intermixed within schwannomas and occur in varying amounts.[6]

Histopathologically, the tumor shows a mixture of Antoni type A and B patterns and may contain prominent blood vessels with hyalinised walls or calcification. These degenerative changes are thought to be caused by insufficient blood flow due to the continuously enlarging tumor.[7] Our literature search for ancient schwannoma of the hand or finger revealed report of ancient schwannoma one arising in the middle finger and another in the thumb palmar aspect of the distal phalanx.[8] White reported schwannomas of the hand and wrist presenting with pain or tenderness and paresthesia in 89% and 59% of cases, respectively, but our patient experienced no pain and was not troubled by the schwannoma, leaving it untreated until it eventually changed into ancient schwannoma and

ulcerated.[9] Humber et al performed preoperative CT imaging of ancient schwannoma in the upper lip and reported that the CT findings were extremely useful for accurate determination of the tumor margin and cystic degenerative changes.[10] Total excision with no need for margins is reported to be the only treatment for ancient schwannoma and post resection recurrence is rare.[11] Indeed, no recurrence was observed at 6 months postoperative in the present case following total excision.[12] Although malignant transformation of ancient schwannoma is extremely rare, caution must be paid when examining patients with ancient schwannoma because it is sometimes difficult to differentiate it from malignant tumors due to the presence of large cystic myxomatous areas with variable bizarre spindle cells and occasional mitoses in FNAC.[10,13] Dahl et al About 8% of lesion reported as pseudosarcomatous lesions in a six year follow up was found to be ancient schwannoma.[14]

CONCLUSION

Ancient schwannomas are a rare entity of benign cutaneous nerve sheath tumor in patients without von Recklinghausen disease. Head and neck, mediastinal, retroperitoneal ancient schwannomas have been reported and has high degree for malignant transformation. Ancient schwannoma of the peripheries have been reported in only few cases and in hand only two cases have been reported. Authors present this case for its rarity and the diagnostic dilemma occurring with FNAC. Thus these tumors if present in periphery and in non-neurofibromatosis patient can masquerade as malignant tumors but still can safely be treated with excision biopsy alone and with almost nil recurrence.

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