PILOMATRIXOMA OF THE ARM: A RARE CASE WITH IMAGING FEATURES

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ABSTRACT

Pilomatrixoma, a benign skin appendageal tumor, is seen commonly in head and neck. Occurrence of pilomatrixoma in the upper extremities is not common and has been reported infrequently in the available literature. We present the case of a 13-year girl with a pilomatrixoma of the arm, its radiological evaluation and the relevant review of literature.

Key words: Appendageal tumor, Pilomatrixoma, benign skin tumor, Radiology

Introduction

Pilomatrixoma is a rare benign tumor arising from hair root follicles of skin. It is also known as Malharbe's calcified epithelioma.[1-3] It is most often seen in children and young adults as a slowly growing nodule in the skin of the head and neck. Occurrence in the upper extremities is relatively rare.[3]

The baseline method used in the assessment of bone and soft tissue tumors is plain radiography. Ultrasonography (USG), computerized tomography (CT) and magnetic resonance (MR) imaging can be used for further charactrerization.[4] We present a case of pilomatrixoma of upper arm with radiological features on multiple modalities. Such a case has not been reported from Pakistan earlier. It highlights the importance of considering pilomatrixoma in the clinical and radiological differential diagnosis of dermal or subcutaneous nodule even in locations other than head and neck region.

Case Report

A 13-year old girl was referred to the radiology department with biopsy proven pilomatrixoma of right arm for proper localization of extent. She gave history of gradually increasing swelling in right arm since last two years after an intramuscular injection. Then she developed pain and discharge from the swelling. Local examination showed a firm mobile swelling on the lateral aspect of right arm. Then the plain radiography, ultrasonography and MRI of the arm were performed. The X-Ray showed only soft tissue mass without any calcification (Fig. 1). High frequency ultrasonography showed a heterogeneous predominantly solid mass in the subcutaneous tissues with significant vascularity on color Doppler. On MRI, it appeared as a well-defined subcutaneous mass measuring 4.1 x 3.8 cm in right arm along the posterolateral aspect of right arm with preserved fat planes with underlying fascia and muscles. The mass lesion appears isointense on TIWI, and heterogeneously hyperintense on T2WI and STIR sequences, showing heterogeneous thick peripheral post contrast enhancement (Fig. 2 a-c).

The patient underwent complete excision of the mass with uneventful recovery. There was no recurrence on a 10 months follow up.

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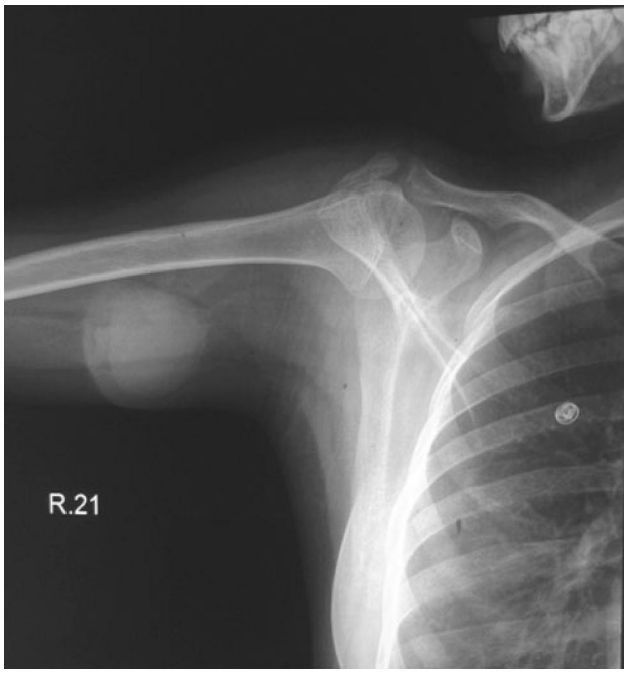
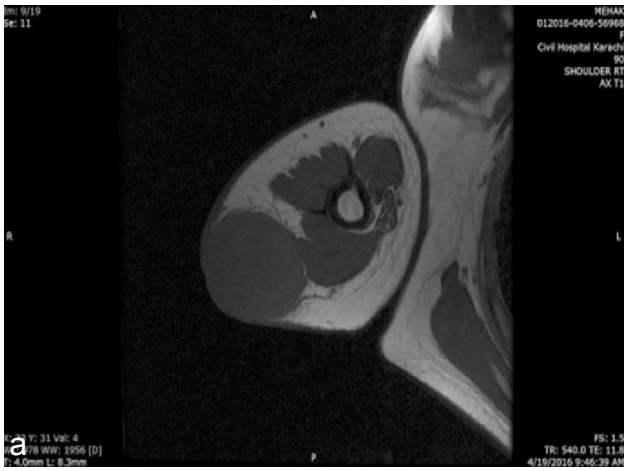
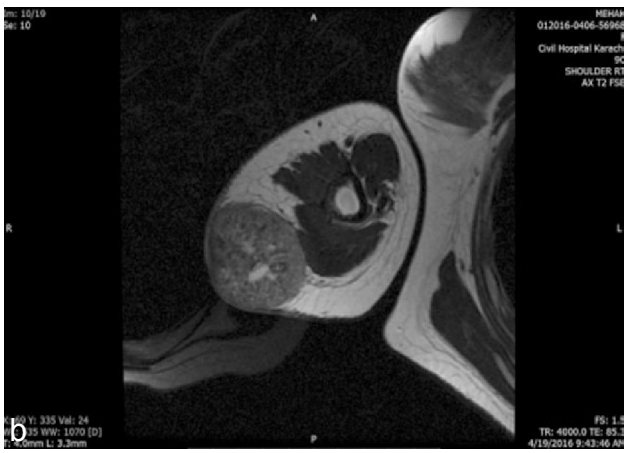


Figure 1: X-Ray of right arm showing a well circumscribed soft tissue mass.





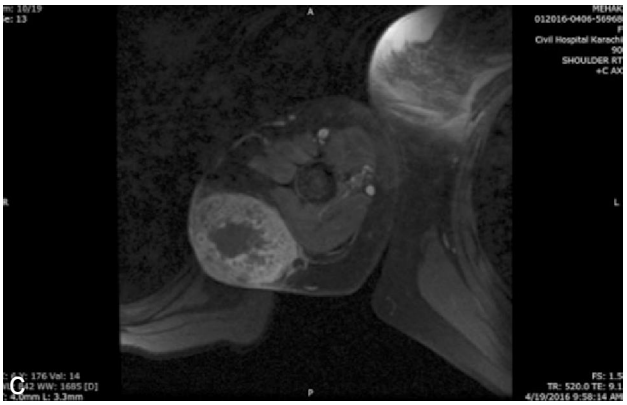


Figure 2 (a-c): Axial T1, T2 and post gadolinium T1 WI showing a well defined subcutaneous mass along the posterolateral aspect of right arm.

Discussion

Pilomatrixoma is a benign skin appendageal tumor with differentiation towards hair follicle matrix cells. This lesion occurs over a wide age range with two peaks: less than 20 years and over 50 years.[5] They are more commonly noted in females.[3,5] (Our patient was a teenage girl).These lesions are typically found in the head and neck region.[3,6] Only a few isolated locations at exceptional locations as in this case have been reported in the literature.[3,6] The largest case series in the literature includes 346 pilomatrixomas of which 15.3% were observed in the upper extremities.[3] Even though trauma and infection were thought as possible triggers of tumor, but its exact etiology remains unknown.[7] There was no any history of infection and trauma excepthistory of intramuscular injection in our case.

Among skin tumors, pilomatrixomais observed at a rate of 0.1%.4 Clinically, it appears as a single, painless, firm, red-blue mobile mass in the subcutaneous tissue.[3,4] The size of the lesion generally ranges between 0.5 and 3 cm,[3] however, few cases of giant pilomatrixoma sized more than 5 cm in diameter were also reported.[8]

Reported discussion of this lesion is essentially limited to the literature of otolaryngology, pathology, and dermatology and radiological descriptions are not available in detail. Diagnostic imaging is generally not employed in the evaluation of pilomatrixomas as they are usually superficial, small, and well-circumscribed. But for surgical management they should be evaluated

radiologically for proper extent to avoid recurrence. Conventional radiography may demonstrate soft tissue mass with foci of calcification.[6,8] Ultrasonography demonstrates a well-defined mass with inner echogenic foci and a peripheral hypoechoic rim or a completely echogenic mass with strong posterior acoustic shadowing in the subcutaneous layer.[8,9] CT demonstrates a sharply demarcated, subcutaneous lesion of soft tissue density, with or without calcification.[8,10] MR imaging may reveal a rim-enhancing lesion with small areas of signal dropout which may be consistent with calcifications.[8,10] In the presently reportedcase the plain radiography showed a nonspecific soft tissue mass without any calcification or bone involvement. Ultrasound showed a heterogeneous predominantly solid mass in the subcutaneous tissues with significant vascularity on color Doppler. Such findings raise the suspicion of an aggressive lesion if biopsy report is not available beforehand. USG of extremity soft tissuesis technically also challenging because of the mobility of lesion and difficulty in maintaining arm position while the scanning is conducted. On MR imaging it appeared as isointense mass on TIWI, heterogeneously hyperintense on T2WI and STIR sequence, showing heterogeneous thick peripheral post contrast enhancement. It excluded deeper invasion and accurately determined the size and extent of the mass. The authors recommend the use of multisequential contrast enhanced MRI for soft tissue lesions that appear suspicious on ultrasound.
Recommended management is surgical excision. Recurrence is uncommon after adequate excision hence the importance of the determination of extent for adequacy of surgical excision. The clinical course is generally benign although malignant transformation has been reported.[3]

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