A Rare Case Of An Ulcerated Pilomatrixoma Of The Right Axilla

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Citation

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Abstract

Pilomatrixomas are benign tumors of hair matrix cells characterized by basaloid and eosinophilic ghost cells. Other names for pilomatrixoma include pilomatricoma and calcifying epithelioma of Malherbe. Most cases of pilomatrixoma occur in children under the age of 10, and the condition is twice as common in females as males.Pilomatrixomas are most common in the head and neck region and may occur at eyebrow, lids and medial canthus. Occurrence of pilomatrixomas at the axilla is rare. We would like to report such a rare case of an ulcerated pilomatrixoma in the right axillary region.

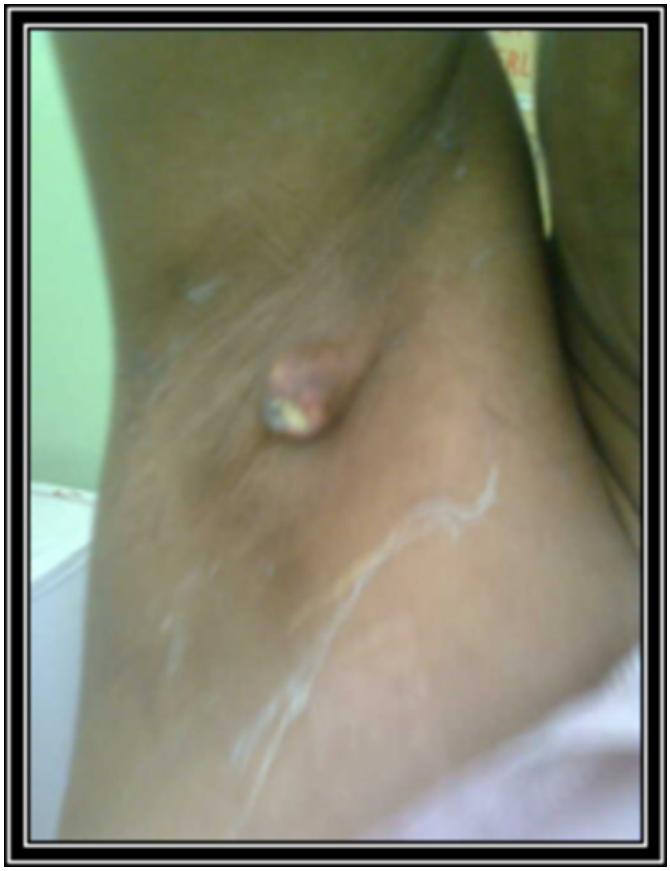
A 45-year-old female, housewife by occupation, came with a lump in the right axillary region for the past 3 years. She had noticed the lump 3 years ago, but had not paid much attention to it. It grew from the original size of about 0.4cm to the present size of about 1cm in diameter. One week before presentation, the lump had burst open giving rise to some purulent discharge and ulceration. She did not have any fever with chills. She did not have any trauma at that site. She did not have symptoms suggestive of tuberculosis, i.e. no fever with evening rise of temperature or cough. She did not have any ulcer on the right upper limb, nor did she have any breast lump or discharge from the nipple. She did not have any loss of weight or appetite.

She had attained her menopause one year ago. She was a known hypertensive for the past 10 years and was taking regular antihypertensive medications and her blood pressure was within control at the time of admission.

On examination, there was a single, globular swelling of 1 x 1 x 0.5cm in the right axillary region at the level of the anterior axillary line. The overlying skin was ulcerated with undermined edges and indurated margins. The lump was firm on palpation, attached to the overlying skin and free from the underlying tissues. Axillary lymph nodes were not enlarged.

Figure 1

Fig. 1: Pilomatrixoma of the right axillary region

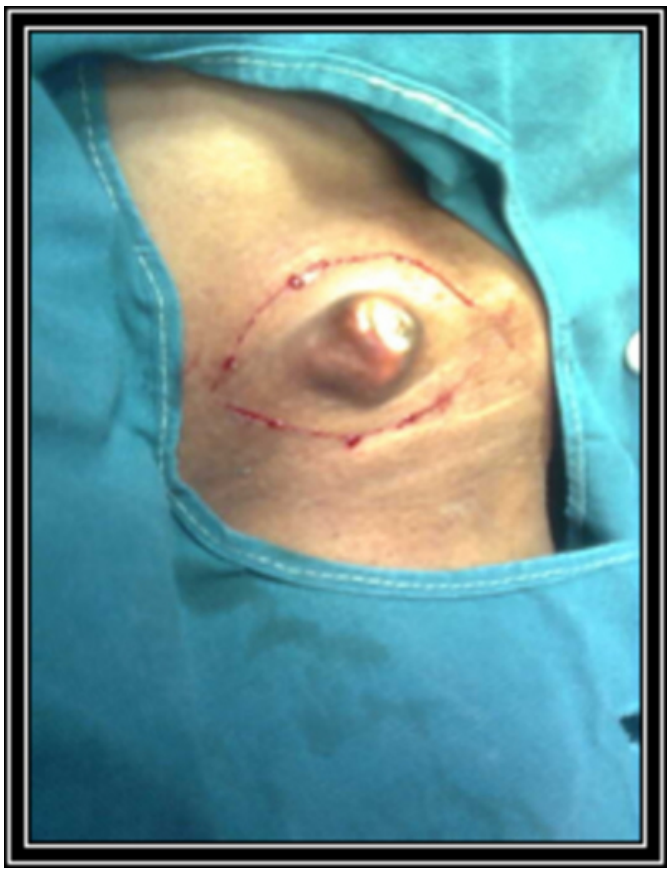


All her blood investigations were within normal limits, except for mild leukocytosis. FNAC report indicated the presence of a pilomatrixoma. She was subjected to a wide excision of the lump with 1cm margins all around and

primary closure of the wound.

Figure 2

Fig. 2: Incision for wide local excision

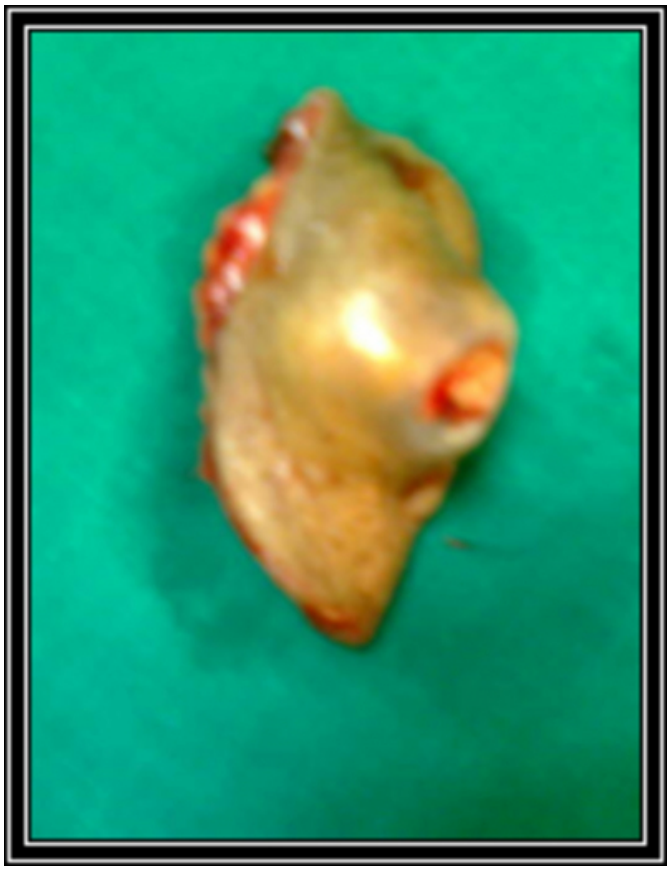


The histopathology report confirmed the presence of pilomatrixoma.

The sutures were removed after a week of surgery. The patient is doing well and is advised to follow up regularly.

Figure 3

Fig. 3: Specimen of excised pilomatrixoma



DISCUSSION

Pilomatrixoma, is a benign, calcifying, cutaneous tumor of children and young adults originating from pluripotential precursors of hair matrix cells. It has a bimodal peak presentation in the first and sixth decade, although it can appear at any age.

Mutations of the beta-catenin gene were detected in 75% of the pilomatrixomas but the exact role of such mutations remains to be elucidated [1]. In one study of 10 pilomatrixoma lesions, all immunostaining results were strongly positive for BCL2. This is a proto-oncogene that helps suppress apoptosis in benign and malignant tumors; these data suggest that faulty suppression of apoptosis contributes to the pathogenesis of these tumors.

More recently, investigators have demonstrated that the proliferating cells of human pilomatrixomas show prominent staining with antibodies directed against LEF-1 (a marker for hair matrix cells). Evidence also indicates that S100 proteins can be used as biochemical markers in characterization of pilomatrixomas. These data provide biochemical support of morphological evidence that these tumors are derived from

hair matrix cells. Furthermore, investigators have shown that at least 75% of persons with pilomatrixomas who have been examined have mutations in the gene CTNNB1; these data directly implicate beta-catenin/LEF misregulation as the major cause of hair matrix cell tumorigenesis in humans.

It presents as a firm to hard, well-defined mass adherent to skin but not fixed to underlying tissue. Reddish-brown hue of overlying skin suggests the diagnosis of pilomatrixoma [2]. Pilomatrixoma are usually solitary but can be multiple, are most common in the head and neck region and may occur at eyebrow, lids and medial canthus. Multiple or familial pilomatrixomas are associated with myotonic dystrophy, Gardner syndrome, Rubinstein Taybi syndrome, Turner syndrome and Trisomy 9.

On ultrasonography, pilomatrixomas are heterogeneously hyper-echoic with internal echogenic foci and a peripheral hyper-echoic rim or completely echogenic with strong posterior acoustic shadowing in the subcutaneous layer.

On magnetic resonance imaging (T1W1), pilomatrixoma gives a homogenous intermediate intensity signal and does not enhance on contrast.

Viable basaloid cells in the periphery, shadow cells in the central part and foci of calcification are characteristic histopathological features of pilomatrixoma. Shadow cells are pathognomonic of pilomatrixoma [3].

Since it does not regress spontaneously, excision is the treatment of choice. Surgical excision including clear margins and its overlying skin prevent recurrence in most cases. Although it is exceedingly rare, rapid growth, pain, itching, ulceration and bleeding may suggest malignant transformation of the pilomatrixoma. The malignant variant of pilomatrixoma, pilomatrix carcinoma, is a low-grade malignant lesion with a tendency to recur and carries a risk of distant metastases. Infiltration of surrounding tissue, necrosis, high number of atypical mitotic figures and peri-neural or vascular invasion are the typical histological features but no feature is specific to confirm whether a malignant pilomatrixoma has arisen de novo or it is a malignant transformation of a preexisting benign pilomatrixoma [4].

It is frequently misdiagnosed as epidermoid cyst, sebaceous cyst, dermoid cyst, foreign body reaction, calcification in lymph node, fat necrosis, pyogenic granuloma, chalazion and keratoacanthoma. Perforating pilomatrixoma may present as draining crusted nodule or ulcer [5]. Perforating or rapidly growing pilomatrixoma can mimic a neoplastic lesion [6].

In our case, the site of the pilomatrixoma was in the right axilla, which is a rare site for the occurance of a pilomatrixoma. The lesion was ulcerated. We did a wide local excision for the lesion and the patient is following up with us regularly.

References

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