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Case Report

Pilomatricoma Mimicking Tubercular Lymphadenopathy

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ABSTRACT

Pilomatricoma, also known as calcifying epithelioma of Malherbe, is a rare benign skin tumor arising from hair follicle matrix cells. It mostly involves head and neck region but can affect other body parts also. Usually, it presents clinically as a single swelling but may present at times as multiple swellings. The affected hair follicle matrix cells undergo calcification and form a hard lump beneath the skin. We present a case of 7-year-old male child who presented with a painless, progressively enlarging swelling on the right side of the neck. The child was otherwise healthy, with no associated symptoms or significant medical history. Multiple unilateral anterior cervical lymph nodes at cervical level 2, 3, 4 which were firm, matted, mobile and non- tender, as identified on clinical examination. Ultrasonography showed prominent lymph nodes and fine-needle aspiration cytology (FNAC) showed clusters and sheets of atypical cells. Excisional biopsy showed sheets of basaloid cells, ghost cell, giant cell reaction and calcification with no malignant cells which confirmed pilomatricoma without malignancy. Follow-ups at 1 and 3 months showed no recurrence. The child is still monitored continuously. Pilomatricoma should always be considered in such cases of cervical lymphadenopathy for accurate diagnosis and management.

Keywords: Pediatric skin tumor, Calcifying epithelioma, Hair follicle tumor, Cervical lymphadenopathy

INTRODUCTION

Pilomatricoma is also referred to as calcifying epitheliomas of Malherbe. Pilomatricoma are uncommon benign tumors associated with follicular skin appendages[1]. Approximately 60% of casesmanifest in first 20 years of life. Children and young adults are predominantly affected with a slight female predilection[2,3]. The head and neck region is the most frequently reported location for these tumors[4]. These lesions typically appear as either solitary or infrequently multiple masses, with a tendency to be asymptomatic. They present as firm, subcutaneous nodules with varying degrees of calcification, commonly attached to the skin but exhibit mobility over the underlying tissues. As the tumor progresses towards the surface, there may be a noticeable bluish hue or ulceration of the overlying epidermis.[5] Due to such presentation they are usually misdiagnosed with other etiologies like tuberculosis, lymphoma and head & neck malignancy. This paper aims to present a case involving a 7-year-old male with a cervical pilomatricoma.

Case Presentation

A 7-year-old male child presented to OPD with right sided painless but progressively enlarging neck swelling noted for 3 months. The lesion was noticed by the parents 3 months ago which was small (1cm*1cm) initially, and gradually increased to present size (4cm*3cm). There were no associated symptoms such as pain, tenderness, or skin changes. There was no history of fever or weight loss. Prior to presentation there was no associated family history. There was no previous history of any chronic illness or medication. The child was immunized for age and his scholastic performance was good.

On admission, he was conscious, alert with stable vitals (heart rate of 84 per minute, respiratory rate of 24 per minute, blood pressure of 102/66mm Hg, and temperature of 98.6°F). The oxygen saturation was 98% at room air. Anthropometry was suggestive of poor nutrition with weight of 16.4 kg (-2SD to -3SD), length 112.5 cm (-1SD to -2SD), BMI 13(-2SD to -3SD). Multiple unilateral anterior cervical lymph nodes were palpable on the right side of the neck with largest measuring 4cm x 4.5 cm. The lymphnodes were firm, matted, mobile and non-tender. Overlying skin was normal without signs of inflammation or discolouration. There was no discharge or visible punctum. There were no palpable lymph nodes at any other site. There was no clubbing, cyanosis, icterus, or pallor on general examination. On abdominal examination there was no organomegaly. On respiratory examination chest was clear with no added sounds. Cardiovascular system was unremarkable. Child was conscious, alert and there was no focal neurological deficit.

Based on above clinical presentation, differential diagnosis of Infective lymphadenopathy (HIV, Tuberculosis); inflammatory lymphadenopathy and malignancy (lymphoma) was made and investigations were planned accordingly.

Management and Follow-Up

Investigations revealed a normal peripheral blood picture with no abnormal cells (Table 1). Peripheral smear showed normocytic normochromic picture with presence of no atypical cells. Erythrocyte sedimentation rate, C - reactive protein, liver function and Kidney function tests were within normal limits (Table 1).

Table 1: Laboratory investigations

Investigations	Results
Hemoglobin	13.4 gm/dl
Total Leukocyte Counts	6720 cells/cumm
- Lymphocytes	45%
- Neutrophils	38%
- Eosinophils	04%
- Monocytes	13%
- Basophils	0%
Platelets	2.66 lakh/cumm
Peripheral Smear	Normocytic normochromic picture
Erythrocyte Sedimentation Rate	8 mm in 1st hour
C-Reactive Protein	8 gm/L
Blood urea	19.8 mg/dl
Creatinine	0.23 mg/dl
Uric acid	3.6 mg/dl
Protein	7 g/dl
Phosphorus	5.4 mg/dl
Calcium	9.9 mg/dl
Sodium	136 mmol/L
Potassium	4.5 mmol/L
Chloride	104 mEq/L
Total bilirubin	0.8 mg/dl
SGOT	31.9 IU/L
SGPT	18.7 IU/L
ALP	164 IU/L
Protein	7 g/dl
Albumin	4.5 g/dl
Globulin	2.47 g/dl
A/G ratio	1.8 g/dl

Work up for tuberculosis included Mantoux test, gastric aspirate for CBNAAT, chest X-Ray and were within normal limits. HIV was non-reactive. USG abdomen was done which showed no significant abnormality and/or organomegaly. USG neck showed multiple bilateral variable sized discrete to conglomerate prominent lymph nodes with few of them showing loss of fatty hilum, largest measuring approximately 2.3 x 8.5 mm at right cervical level 2. FNAC of

largest mass in right side of neck showed clusters and sheets of atypical cells. Atypical cells have hyperchromatic round to oval nucleus with scanty to moderate amount of cytoplasm. Few multinucleated giant cells and scattered lymphoid cells are also seen in the background (Figure 1).

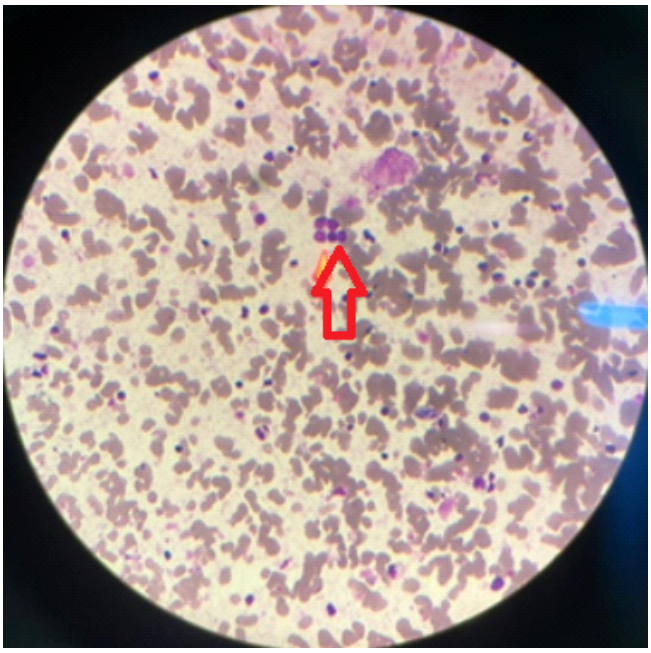

Figure 1: FNAC from cervical lymphnode showing cluster of basaloid cells with deeply staining nuclei and scant cytoplasm (Giemsa stain, X40), showed by arrow (in red) in the figure below

Due to presence of atypical cells in the FNAC, excisional biopsy of lymph node was done which showed sheets of basaloid cells, ghost cells, keratinisation, foreign body giant cell reaction and calcification which are suggestive of pilomatricoma. No evidence of any malignant cell was found (Figure 2).

Regular dressing was done on alternate day for 1 week, and child was discharged thereafter. Follow-up examinations at 1 month and 3 months following excision were not suggestive of recurrence, and the wound had healed without complications. Multiple other small lymph nodes which were earlier palpable were also reduced. The child remains under regular follow-up for monitoring.

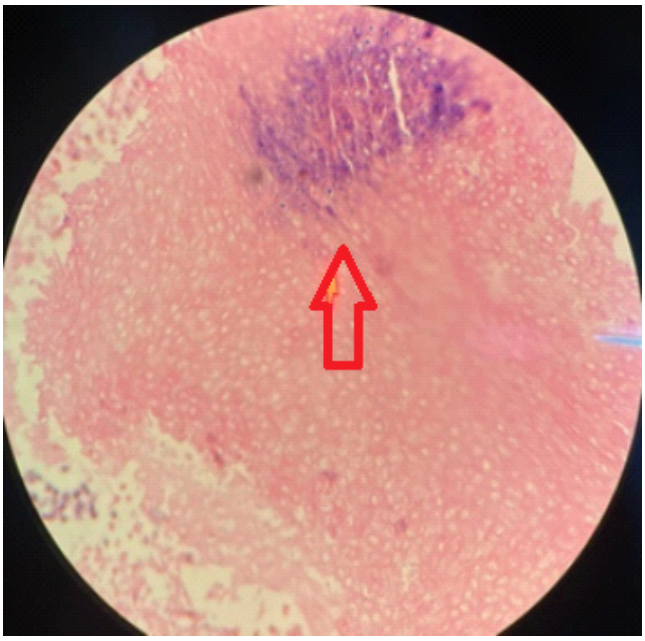

Figure 2: Excision biopsy showing a nucleated ghost cell (shadow cells) (H & E, X40), showed by arrow (in red) in the figure below

DISCUSSION

Pilomatricoma is a rare, benign, dermatological tumor that is superficially located and most commonly occurs in the head and neck region. Most cases occur in the first and second decades of life. Females are more often affected[3]. Although malignant transformation has been described, it is extremely rare. Diagnosis is usually suspected based on palpation of a superficial, rock-hard nodule because of underlying calcification. Tumours were often mistaken for other lesions like epidermal cyst; calcific lymph node. Preoperative investigations were not helpful and the diagnosis is confirmed by histopathological examination. Since this neoplasm doesn't spontaneously regress, surgical excision is both diagnostic and curative as well. Recurrence is rare[2].

In a similar case report by Mohr and Tschen, a 4-year-old boy presented with cervical lymph node swelling on the left side of his neck with a draining sinus. FNAC showed atypical and ghost cells. Lesions exhibiting ghost cells include pilomatricomas, craniopharyngiomas, calcifying cystic odontogenic

tumours, and odontomas.[1] However, the tumour location in this case was characteristic of a pilomatricoma or pilomatrix carcinoma. Also the ghost cells and draining sinus narrowed the diagnosis to perforating pilomatricoma. To further confirm the diagnosis, authors had planned an excisional biopsy.

In another case series by Bansal *et al.*[5] 53 cases of 83700 histopathology samples were reported as Pilomatricoma over a period of 13 years. The study showed female preponderance, with head and neck being the commonest site. Out of 14 biopsy proven cases of pilomatrixoma, cytology findings revealed pilomatricoma in 7 cases based on ghost cells, groups of basaloid cells, squamous cells in combination with multinucleated giant cells and calcium deposits in a background of debris. The main reasons for erroneous diagnosis were predominance of one component over the others and non-representative aspirated material. In our case, FNAC showed atypical cells with ghost cells which were similar to above case study.

Although the recurrence rate of pilomatricoma is rare but Hasan *et al.*[6] in their study of 802 patients showed that the most common location was head and neck (58%), followed by upper limbs (23%), trunk (14%), and lower limbs (5%) with mean lesion diameter of 14.0 ± 7.4 mm.

Several differential diagnoses can be proposed for pilomatricoma lesions, including trichilemmal cyst, basal cell carcinoma, keratoacanthoma, squamous cell carcinoma, hemangioma, metastasis, and other benign tumours of skin appendages such as cylindroma and spiroadenoma[7,8].

Suggested treatments for pilomatricoma include excision of the lesion, punch incision, and curettage. In the case of malignant pilomatricoma lesions, it is recommended to remove the lesion with a margin of 0.5 to 1 cm.

CONCLUSION

Although Pilomatricoma is a rare tumor of head and neck region, but it should be considered in the differential diagnosis of superficial head and neck masses in pediatric patients. Recognizing it early, achieving an accurate diagnosis, and implementing suitable management through surgical excision, are crucial for achieving optimal outcomes.

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