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Case Report

Pilomatricoma-overshadowed by the Shadow Cells: A Case Report and Review of Literature

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ABSTRACT

Pilomatricoma is relatively an uncommon benign cutaneous neoplasm predominantly in the first two decades of life. It is often misdiagnosed by non-dermatologists due to a lack of knowledge about this tumour. However, it can be easily diagnosed by a pathologist if classical histopathological features are present. We are presenting this case as there were varied microscopical findings that posed a diagnostic challenge to us.

Keywords: Pilomatricoma, Basaloid cells, Shadow cells, Ghost cells

INTRODUCTION

Pilomatricoma or calcifying epithelioma of Malherbe is relatively an uncommon benign cutaneous neoplasm originating from hair follicles. It represents around 0.12% of all skin tumours.[1] It clinically presents frequently as firm to hard subcutaneous nodules predominantly in the first two decades of life in the head and neck region. It is misdiagnosed in 75% of cases by non-dermatologists.[2] Pilomatricoma can be easily diagnosed on histopathology when it has classical features of basaloid cells and shadow (ghost) cells. We are presenting this case in a child, where there was an absence of basaloid cells which posed a diagnostic challenge and to enlighten the pathologists about varied morphological presentations of this entity.

CASE REPORT

An 8-year-old female child presented to us with complaints of swelling over the back of the neck for 2 months. There was no history of fever, cough or contact with a known case of tuberculosis. On examination, a well-defined swelling was seen over the back of the neck measuring 1.2 cm which was tender on palpation. Other system examinations were normal. The differential diagnostic possibilities considered clinically were dermoid/calcified sebaceous cyst. Ultrasound done showed a 1.1 × 0.5 cm partially calcified lesion in the subcutaneous plane and skin of the right nape of the neck and suggested possibilities of calcified sebaceous cyst or calcified trichilemmal inclusion cyst. The lesion was excised under local anaesthesia and the specimen was sent for histopathological examination.

On gross examination, it was a well-circumscribed grey white soft-tissue mass, hard to feel measuring 1.2 × 1 × 0.8 cm. The specimen was gritty to cut and the cut section was grey-white

with flecks of calcified areas in-between [Figure 1]. Microscopic examination showed predominantly trichilemmal keratinisation, and ghost cells [Figure 2] along with extensive areas of calcification. Interstitial fibroblastic proliferation with occasional foreign body giant cell reaction and admixed chronic inflammatory infiltrates were seen. On searching one, focus revealed an occasional collection of negligible basaloid cells in the periphery undergoing abrupt trichilemmal keratinisation [Figures 3 and 4]. Based on the histopathological findings, a diagnosis of Pilomatricoma was made.

DISCUSSION

Pilomatricoma or calcifying epithelioma of Malherbe was first described by Malherbe and Chenantais in 1880. Later, in 1922, Cazenave and Dubreuilh reported the classic histopathologic features of pilomatricoma as islands of basaloid cells and shadow (ghost) cells. It can present at any age but demonstrates bimodal peak presentation during the first and sixth decades of life with 40% occurring in patients younger than 10 years and 60% occurring before the second decade of life.[3] There is a slight female preponderance with the female to male ratio being 1.5:1. The incidence of this benign cutaneous tumour is approximately 1 in 2000 surgical specimens. The most common sites of pilomatricoma include the head and neck, followed by upper limbs, trunk, and lower extremities.[4] Our case as said in the literature is a female in the first decade with swelling over the neck region, a usual site for the occurrence of pilomatricoma.

Pilomatricoma most often presents clinically as a solitary cutaneous nodule with an average size of 1 cm and rarely exceeds 2 cm in diameter. It is covered by normal or

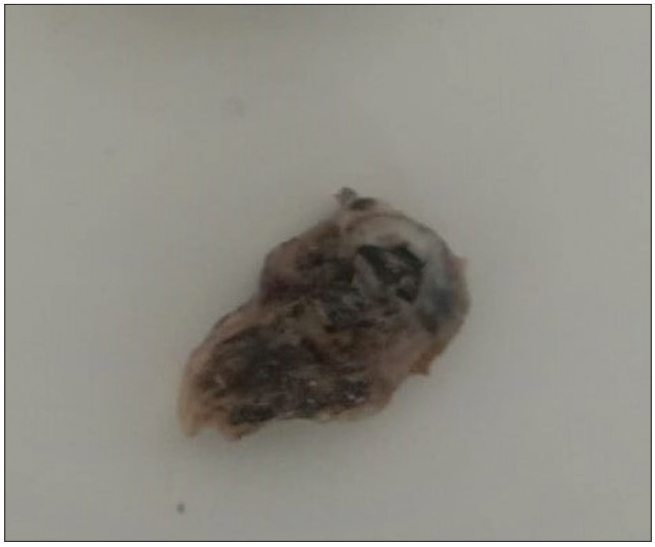

Figure 1: Gross showing the grey-white tumour.

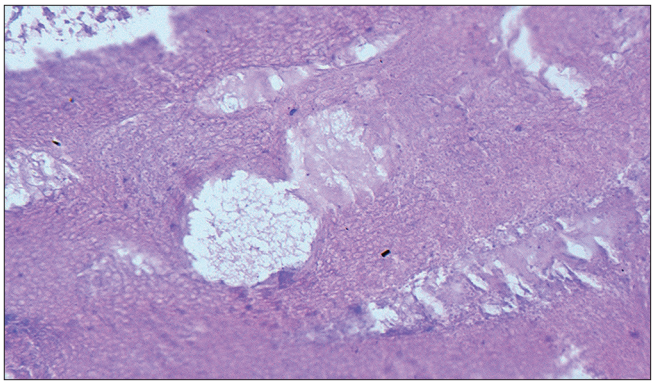

Figure 2: Microphotograph showing sheets of ghost cells without any basaloid cells (H&E, ×100).

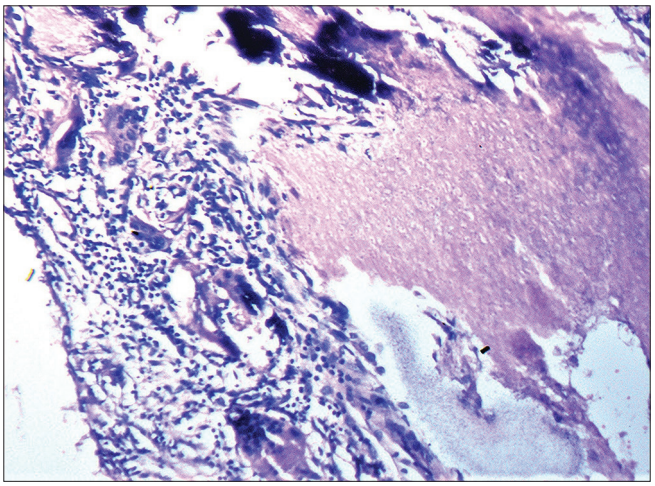

Figure 3: Microphotograph showing sheets of ghost cells with foreign body giant cell reaction and calcification (H&E, ×100).

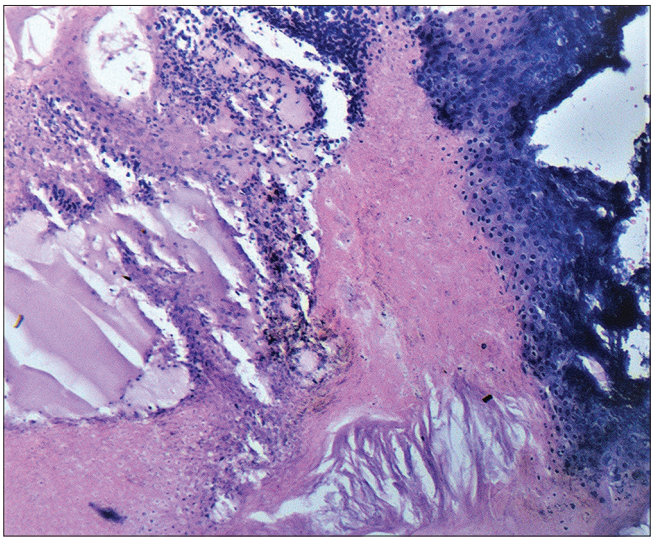

Figure 4: Microphotograph showing negligible basaloid cells in periphery undergoing abrupt trichilemmal keratinisation, ghost cells (H&E, ×400).

hyperaemic skin. They are asymptomatic, firm, non-tender, deep-seated, and adherent to overlying skin but not fixed to underlying tissue. Stretching of skin over tumour produces 'tent sign' with multiple angles and facets which are pathognomonic of pilomatricoma. On applying pressure on one edge of the tumour, the opposite edge protrudes like a 'teeter-totter'. The other classic feature is the blue-red discoloration of overlying skin which helps to differentiate pilomatricoma from its common clinical differential – epidermal inclusion or dermoid cyst.[4] In our case, the size of the lesion is <2 cm and it is a little deeply seated without any fixation on the underlying tissue.

Familial occurrence and multiple lesions are known and are associated with Turner syndrome, Gardner's syndrome, Steinert disease, Sarcoidosis, Rubinstein–Taybi syndrome, and Myotonic dystrophy.[3] Due to the unfamiliarity of this tumour among clinicians, the accuracy of pre-operative diagnosis is between 0% and 30%. Radiology and fine-needle aspiration cytology have a limited role in the diagnosis of pilomatricoma.[4]

Histologically, pilomatricoma is an encapsulated tumour composed of basaloid cells and ghost (shadow) cells. Early lesions show predominantly basophilic basaloid cells. As the tumour matures, the basaloid cells undergo abrupt trichilemmal keratinisation with loss of nuclei and become shadow (ghost) cells, which mainly occupy the central portion of the tumour. There can be also areas of calcification, ossification, melanin deposition, mononuclear infiltration, and foreign body reaction near ghost cells. Haemorrhage, myxoid change, ossification, oedema, and stromal fibrosis may occur as secondary changes.[5] Based on the tumour evolution from a matrix cyst to a nodule with no recognizable basaloid cells, four morphological stages are described: (i) Early: small, cystic lesions, (ii) fully developed: large cystic lesion, (iii) early regressive: foci of basaloid cells, shadow cells and lymphocytic infiltrate with multinucleated giant cells and (iv) Late regressive: predominant shadow cells, absence of basaloid cells and inflammatory cells.[6] Our case too was in stage iv with the predominance of shadow cells and negligible basaloid cells with foci of calcification. This non-classical feature posed a diagnostic challenge.

The aetiology of pilomatricoma is unknown, but, recently, it has been attributed to the overactivity of BCL-2 and mutation in CTNNB1.[7] Malignant transformation to pilomatrix carcinoma is rare. Since there is no possibility of spontaneous regression, surgical excision is the only treatment option available.[4]

CONCLUSION

Pilomatricoma is a rare skin tumour mainly seen in children and young adults. It is often misdiagnosed as a dermoid cyst as in our case. The presence of classical histopathological features makes the diagnosis easier. Histology too varies depending on the stage of maturation of tumour. The absence of basaloid cells and the predominance of shadow cells overshadowed the diagnosis of pilomatricoma and posed a diagnostic challenge. Thus, we presented this case to enlighten the pathologists about the varied histopathological features of pilomatricoma.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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