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A RARE CASE OF PROLIFERATIVE TRICHILEMMAL TUMOR IN A 38-YEAR-OLD MALE MASQUERADING AS SCALP LIPOSARCOMA

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ABSTRACT

This is a case of a 38yr old male who presented with a history of a slowly growing scalp lesion initially noticed about 10 years ago. The mass was painless, and the patient has no constitutional symptoms. There was no preceding history of trauma. Plain radiograph of the skull showed a lobulated mass of soft tissue density in the left parietal scalp region close to the vertex. The underlying skull vault was intact. Cranial computed tomographic (CT) scan revealed a well-defined solid, lobulated mixed density mass within the skin and subcutaneous tissue in the left parietal region close to the vertex with multiple coarse calcifications within it; the mass showed heterogeneous contrast enhancement. No intracranial extension of the lesion was seen and there was no evidence of vault destruction. Histology following incisional biopsy revealed a rare proliferative trichilemmal tumor.

Keywords: Rare, Scalp, Trichilemmal Tumor, Liposarcoma

INTRODUCTION

Scalp lesions can be classified as congenital, traumatic, inflammatory, or neoplastic in origin.¹ The majority of scalp lesions are benign, with cystic lesions accounting for over 50% of all benign scalp lesions. Such lesions include trichilemmal cysts (pilar cysts), sebaceoma, epidermoid cysts, dermoid cysts, teratoid cysts and benign neoplasms of

the scalp, including melanocytic nevi, keratoacanthoma, pilomatricoma, neurofibroma, and lipoma.^{2,3} Malignant scalp tumors like liposarcomas are rare; however they carry a potential risk of delayed detection, resulting in poorer outcomes.¹⁻³ Most scalp lesions show nonspecific imaging findings, especially on plain radiograph, although some possess characteristic features on CT and Magnetic Resonance Imaging (MRI).

The proliferating trichilemmal tumor (PTT), also known as pilar tumor, is a slow growing lobulated solid-cystic lesion showing trichilemmal differentiation present in the isthmus of the hair follicle.^{4,5,6} It affects 5% to 10% of the population, with a female predominance, and can be inherited in an autosomal-dominant fashion.¹⁻⁶ Ninety percent are located on the scalp, but it can also affect the face, trunk and back (10%).^{3,5,7}

CASE REPORT

A 38-year-old male presented to the general outpatient department (GOPD) of Aminu Kano Teaching Hospital, kano, with a history of a slowly growing scalp lesion initially noticed about 10 years previously which started as pea-sized lesion but over the last 2 years, he noticed the mass was progressively enlarging. The mass was painless and the patient had no constitutional symptoms. There was no preceding history of trauma. No history of similar lesion in the family or close relations. Physical examination revealed a

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single, firm and slightly fluctuant mass in left parietal area of the scalp close to the vertex which measured about 6 x 4 cm in size. It was not fixed to the underlying bone. There was no regional or cervical lymphadenopathy. Systemic examination was essentially normal.

A plain radiograph of the skull, AP and Lateral (Figure 1) was requested which showed a lobulated mass of soft tissue density in the left parietal scalp region close to the vertex which was continuous with adjacent subcutaneous tissue. No focal area of calcification or lucency was seen within it. The underlying skull vault was intact. The remaining aspect of the cranial and facial bones were within normal limits. An impression of a scalp soft tissue lipoma to rule out epidermal inclusion cyst was made.

The patient was booked for surgical removal of the lesion but he became apprehensive and decided against it. About 4 months later, he re-presented history of recurrent headache over the preceding one month but had no history of fever. Examination showed a slight increase in size of the scalp lesion measuring 6x5cm in size. His blood pressure was 110/78 mmHg. Other systemic examinations were unremarkable. Relevant laboratory investigations prior to surgery were all within normal limits. Cranial computed tomography (CT) scan revealed a well-defined solid, lobulated mixed density mass within the skin and subcutaneous tissue with pre-contrast CT value ranging between >300HU for calcifications and 80-100HU for mixed density areas, 15-40HU for soft tissue and -8 to -15 for fatty tissue in the left parietal region close to the vertex with multiple coarse calcifications within it and the mass also showed heterogeneous contrast enhancement indicating mild vascularity of the lesion with no evidence of skull bone infiltration, suggestive of a lipomatous tumor. It measured 6.4 × 5.6 × 4.4 cm in size (Figure 2). The mass was not attached to the underlying skull bone (Figure 3) with no evidence of cranial vault destruction or intracranial extension. 3D volume rendered image confirmed intact cranium (Figure 4). Based on the CT findings the differential diagnosis were dermoid cyst, proliferating trichilemmal tumor,

pilomatricoma and liposarcoma with borderline malignant transformation.

Incisional biopsy results revealed cell nests composed of cells with large cytoplasm without atypia but with eosinophilic centers lined by walls of stratified squamous epithelium that demonstrated abrupt keratinization (trichilemmal keratinization) and ghost cells. A histologic diagnosis of proliferating trichilemmal tumor was made. The tumor was then completely resected with free surgical margins and the procedure was well tolerated with no post-operative complications. A year post-op, the patient was doing very well with no history of recurrence or complication.

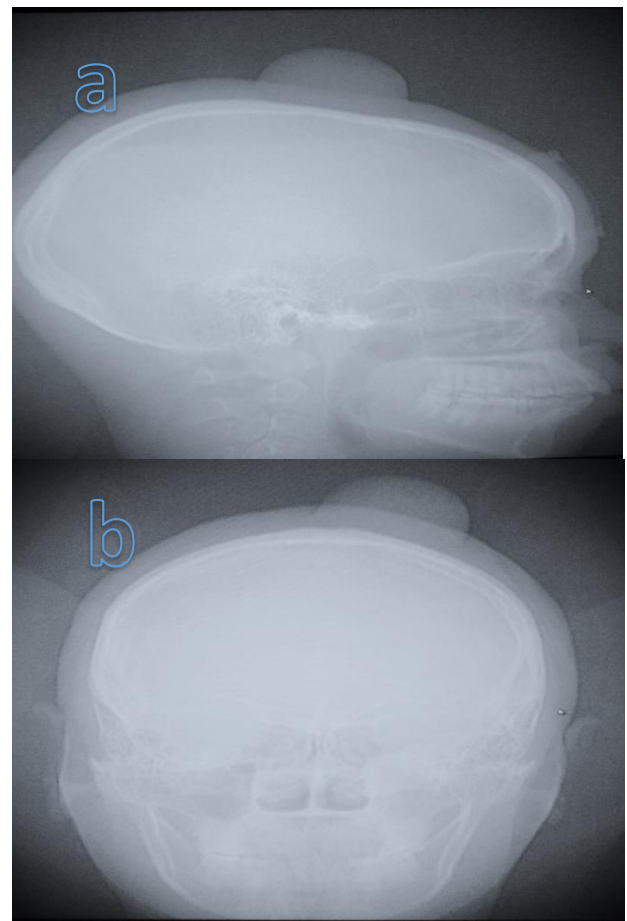


Figure 1: Plain skull radiograph showing a lobulated mass of soft tissue density in the left parietal scalp region close to the vertex that is continuous with adjacent subcutaneous tissue. No calcification or lucency is seen within it. The underlying skull vault is intact.

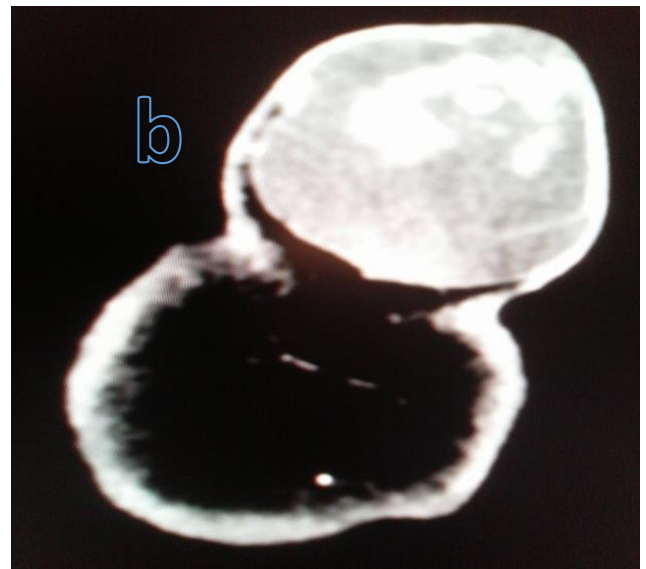
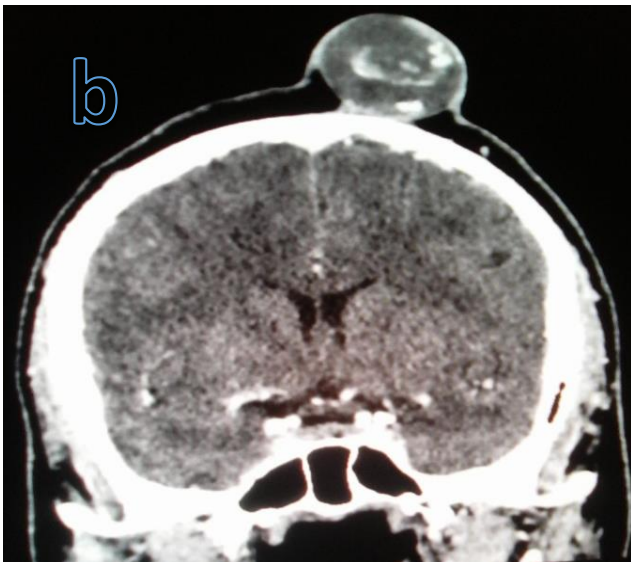
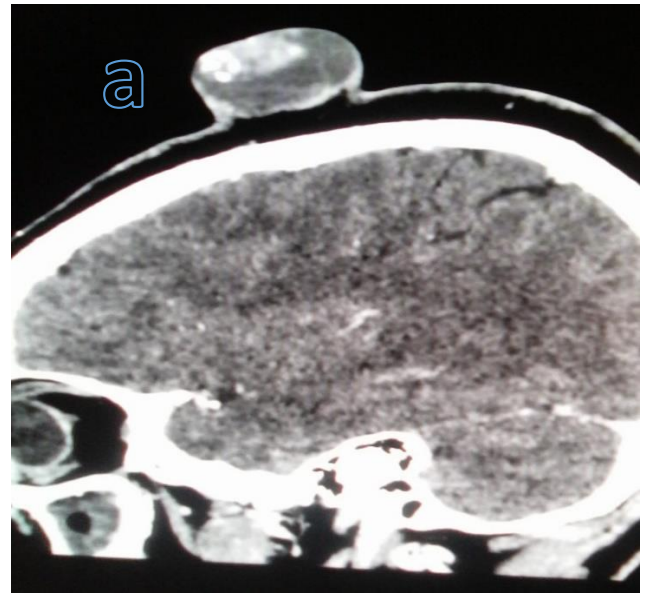
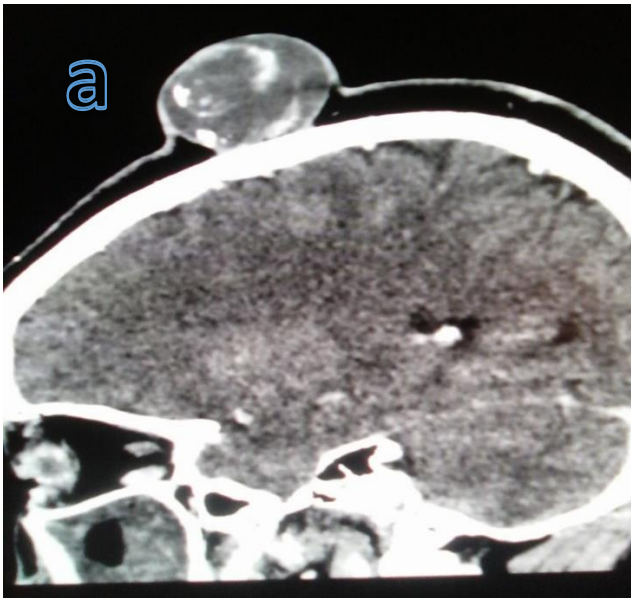


Figure 2: Sagittal (a) and Coronal (b) reconstructed, contrast enhanced, cranial computed tomographic images (brain window) showing a well-defined solid, lobulated mixed density mass within the skin and subcutaneous tissue in the left parietal region close to the vertex with multiple coarse calcifications noted within it. The mass showed heterogeneous contrast enhancement.

Figure 3: Sagittal reconstructed (a) and Axial (b), contrast enhanced, cranial computed tomographic images (brain window) showing non-attachment of the mass to the calvarium and subcutaneous location.

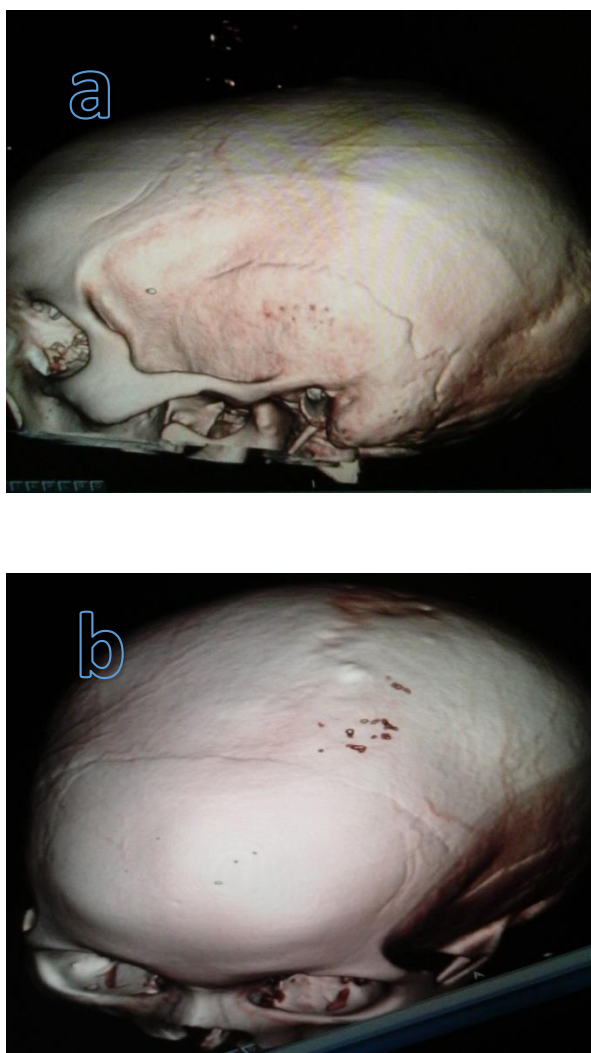


Figure 4: Left lateral (a) and Anterior left oblique (b) 3D Volume Rendered images showing left parietal location of the mass close to the vertex with underlying intact skull bone and the intralésional calcifications above.

DISCUSSION

Proliferating trichilemmal tumor is a rare tumor arising from the epithelium of hair follicle.⁵⁻⁹ It is more common in elderly female patients.¹⁻⁹ The index patient was a young man which is a rare finding. However, there are cases described in young patients, but they are mostly females and there are no report of such in local literature.^{5,9} Ninety percent of cases are found on the scalp and present as a slow-growing solitary nodule but may occur less frequently in other

topographies, such as the vulva, trunk, face, lips, buttocks and back.^{3,5,7,9} Proliferating trichilemmal tumor is usually a benign neoplasm but rarely may undergo malignant transformation in a step-wise manner starting with an adenomatous stage of the trichilemmal cyst to an epitheliomatous stage of the proliferating trichilemmal tumor evolving into the carcinomatous stage of the malignant proliferating trichilemmal tumor.^{7,8,9} Therefore, clinically and microscopically, proliferating trichilemmal tumor can simulate malignancy and the correct diagnosis is essential due to the indolent biological behavior of these lesions.^{5,8} In the case presented, the scalp lesion showed non-specific findings on plain radiographs initially suggestive of a lipoma and the CT findings were relatively more characteristic but still not diagnostic as CT features of the tumor are those of multiple complex subcutaneous solid or cystic masses.¹⁻⁴ The cystic components contain high density proteinaceous material which sometimes layers dependently. MRI is superior to C.T. in the evaluation of this tumor but, it was not available at the time the patient presented. However, microscopically, the appearance was consistent with proliferating trichilemmal tumor which showed proliferation of cell nets lined by stratified squamous epithelium with abrupt trichilemmal keratinization and ghost cells, without atypical cells, as described in the literature.^{5,7,8,9}

Because proliferating trichilemmal tumor has a benign biological behavior, the recommended treatment is complete resection with free surgical margins (surgical excision with a 1-cm margin of normal tissue).^{5,7,8,9} This was the treatment of choice performed for the index patient so as not to give a chance for possible recurrence or rarely malignant transformation, based on literature reports of cases of proliferating trichilemmal tumor with recurrence and aggressive clinical behavior and/or malignant transformation.^{5,7} The aggressive behavior is more common in tumors located out of the scalp, when it is fast-growing and infiltrative, tumors larger than 5 cm, presence of atypia and mitotic activity.^{5,9}

The index case showed no clinical and/or microscopic aspects related to the more aggressive biological behavior.

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