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CASE REPORT

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Giant pilomatrix carcinoma of the scalp with intracranial extension: A case report

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Key Clinical Message

A very rare skin cancer. Malignant variant of pilomatricoma. It is unique because of its highly aggressive behavior. Responds poorly to chemotherapy and radiotherapy. Treated with a wide surgical excision with safe margins.

Abstract

Pilomatrix carcinoma is uncommon and locally aggressive tumor of the hair follicle matrix. It is a malignant variant of pilomatricoma. If left untreated for a long time may attain massive size and has the potential for distant spread. Only few cases with brain extension have been reported in the literature. Diagnosis of these tumors is established by histopathology. Although pilomatrixoma and pilomatrix carcinoma are well-recognized lesions, clinically they are frequently misdiagnosed as other skin conditions. The tumors respond poorly to chemotherapy and radiotherapy. Thus, recommended treatment is a wide surgical excision with safe margins. Herein, we report the case of a 39-year-old African woman with pilomatrix carcinoma of the scalp eroding the skull bone with intracranial extension.

KEYWORDS

case report, malignant pilomatrixoma, pilomatrix carcinoma, recurrence, skin cancer, wide local excision

1 | INTRODUCTION

Pilomatricoma also known as pilomatrixoma is a benign skin adnexal tumor originating from hair matrix and cortex. Pilomatricoma is uncommon cutaneous appendage tumor in patients 20 years of age or younger.[1] On the other hand, pilomatrix carcinoma (PC) is an extremely rare malignant skin tumor. PC arises de novo or through malignant transformation of a pilomatrixoma. PC is locally aggressive with high rate of local recurrences and metastases. Only few cases of PC with massive size arising from the scalp with intracranial extension have been reported in the English literature.[2] These tumors are observed frequently in the white male over 50 years old.[3] Histopathological diagnosis may be challenging especially in small specimens. Treatment consists of a wide surgical excision. Mohs Micrographic Surgery seems to be a good modality to limit margins. Herein, we report the case of a 39-year-old woman with PC of the scalp extending to supra auricular area a brief review of the literature.

2 | CASE PRESENTATION

A 39-year-old female presented with a 4-year history of expansive scalp mass at left temporal region. The mass was extending to the parotid region and was associated with left sided facial pain and weakness along with difficulty in opening the mouth and chewing. She also reported loss of hearing of the left ear. During the course of her illness, she initially attended local dispensaries, herbalists as well as over-the-counter medications with no relief. On physical examination, she was ill looking, wasted, with trismus grade 2, as well as facial nerve palsy grade 3. She was a peasant, non-smoker, and non-alcoholic. She had no familial history of cancers, her past medical and family history were essentially unremarkable. On local examination, a huge left scalp mass measuring 15 cm by 20 cm was seen. The mass was non-tender, soft, and warm with tense shiny skin on inferolateral aspect. The left external auditory canal was obliterated (Figure 1). CT scan and MRI of the head and neck showed a destructive mass eroding the skull bone with some intracranial extension (Figure 2). CT scan of the chest and abdomino-pelvis for staging and to rule out distant metastasis were performed. The results came back as normal. An impression of malignant scalp tumor was entertained. Incisional biopsy from the mass was suggestive of basal cell carcinoma. The patient was scheduled for debulking surgery.

2.1 | Surgical procedure

A tracheostomy was fashioned to facilitate endotracheal intubation to administer general anesthesia and maintain the airway. Blunt dissection was used to separate temporalis fascia from muscles. Under general anesthesia and in sterile manner, a transversal incision was made on temporal bone skin and flap elevated. Multiple cystic masses with pocket of pus and serosanguinous fluid were observed and the necrotic tumor had lysed the squamous part of temporal bone with brain meninges intact. Debulking surgery of the tumor mass was done with scant tissue retained due severe bleeding (Figure 3A–C). Brain meninges were reserved and hemostasis achieved. Despite the huge tumor size, the defect was successfully repaired in layers without a need for skin graft and drainage was inserted. Reasonable functional and esthetics were obtained (Figure 3D–F). The resected tumor was submitted for histopathology. The patient was nursed in Intensive Care Unit for 48 h then was transferred to the ENT ward. There, she spent 10 days before being discharged. Grossly, the submitted tumor specimen was fragmented (in piecemeal) with aggregate dimensions of 18 × 13 × 6 cm. Histopathology results demonstrated the tumor composed of combination of atypical basaloid cells with transition into ghost cells, mitoses, and necrosis. The morphology was consistent with PC (Figure 4A,B) with differential diagnosis of basal cell carcinoma with metrical differentiation. The patient was discussed in multi-discipline tumor board where the decision of adjuvant radiotherapy was suggested. Currently, the patient is undergoing adjuvant radiotherapy.

3 | DISCUSSION

Pilomatrix carcinoma (PC) is a malignant variant of pilomatricoma. It is also known as pilomatrical carcinoma or malignant pilomatrixoma. PC plays a minor role among dermal tumors due to its rarity. For the same reason, its biology and natural course of evolution are still poorly defined.[1] It is unclear if these lesions arise de novo or through malignant transformation of a pre-existing benign pilomatrixoma. It is believed that pilomatrixoma and PC are two extremes of a progression towards malignancy.[4,5]

As documented in the literature, PC is clinically aggressive presenting as an exophytic tumor that is

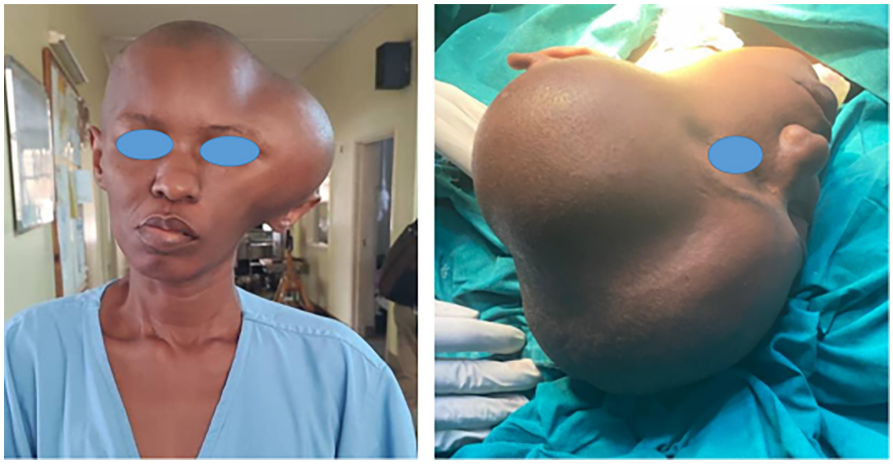

FIGURE 1 Photograph of the patient showing a huge left scalp mass which was non-tender, soft and tense on inferolateral aspect obliterating left ear auricular canal.

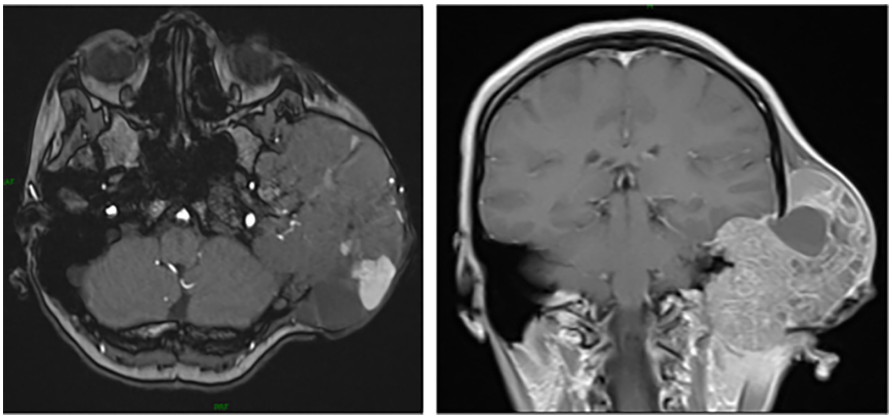

FIGURE 2 MRI and CT scan of the head and neck showing a destructive mass eroding the skull bone with intracranial extension.

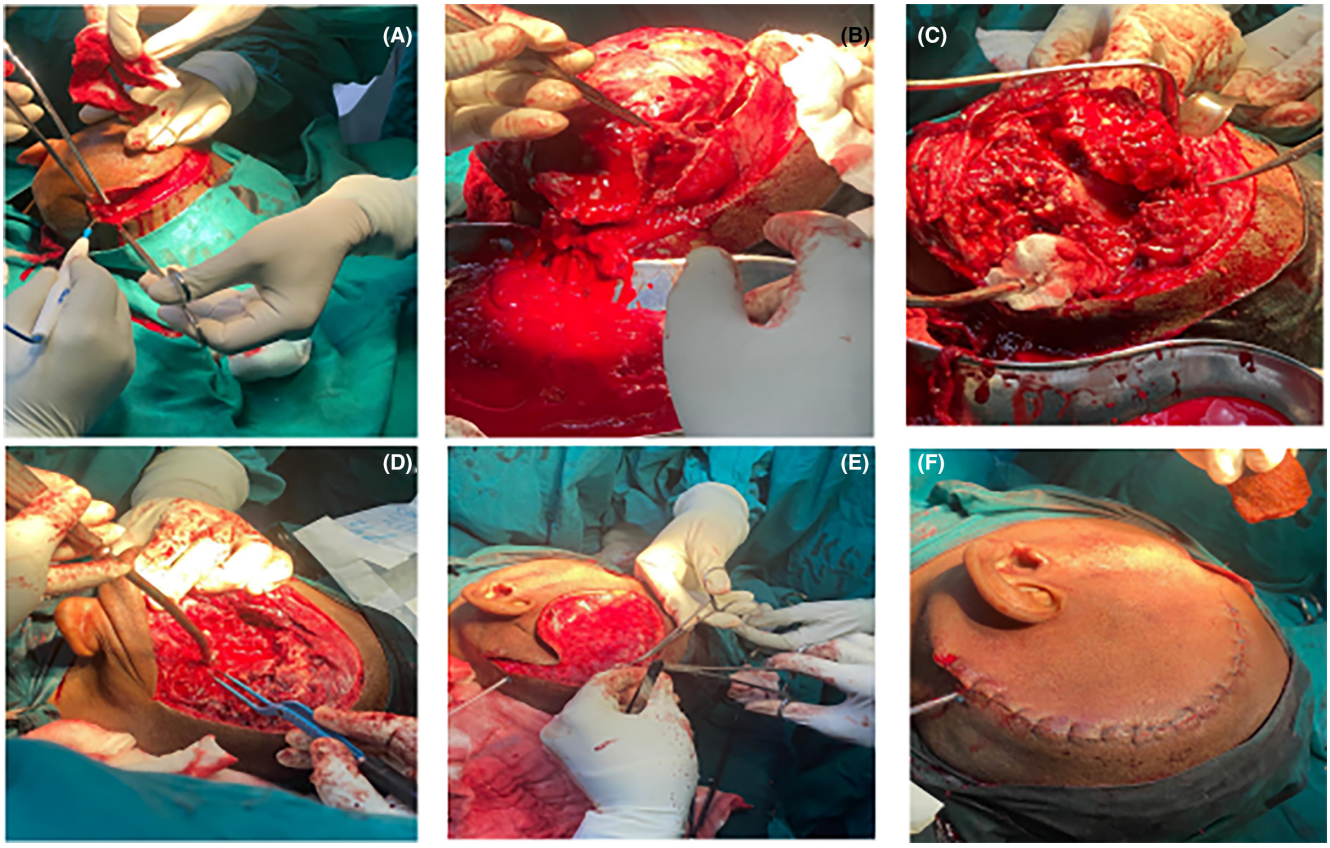

FIGURE 3 Photographs of the patient intraoperatively demonstrating a transversal incision on temporal bone skin and an elevated flap (A), the tumor debulking procedure associated with excessive bleeding (B), the solid and cystic tumor components, with a serosanguinous fluid (C), the erosion of the squamous part of the temporal bone by the tumor, but the brain meninges were intact (D), achievement of hemostasis and the successful repair of the defect in layers without a need for a skin graft (E), appearance of the patient immediately after surgery and a drainage tube (F).

infiltrating subcutaneous fat. Contrary to the index case, the mean age at diagnosis in most patients is 60 years, and men are more affected than females. The tumor has predilection to the head and neck region particularly in the preauricular area, scalp, posterior neck, and upper back as compared to other parts of the body.[6,7] Clinical differential diagnosis includes not only pilomatricoma, epidermal cyst, basal cell carcinoma, and squamous cell carcinoma, but also malignant melanoma, vascular lesions among others.[8,9] The tumor can recur in more than 60% of cases treated by simple excision. Surprisingly, these tumors were previously considered to be low grade and slowly growing with a very poor potential of distant spread.[2] At present, as it has been lighted in the recent literature and in this case, PC is a highly locally aggressive neoplasm, with strong tendency to relapse and with the possibility of secondary spread.[5–7]

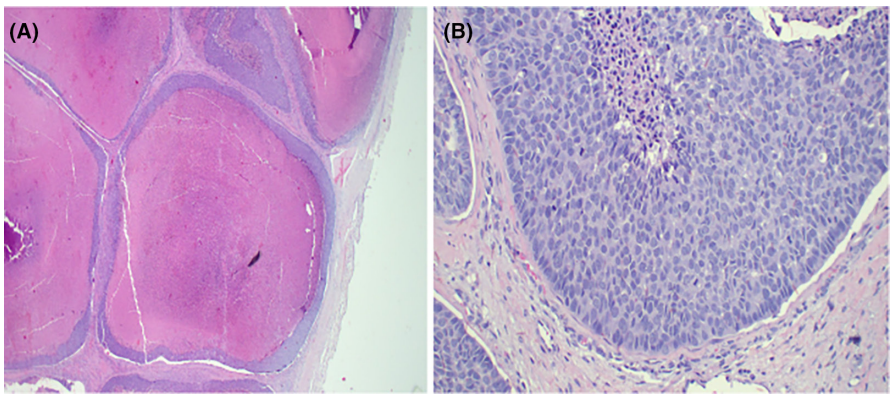

FIGURE 4 Histopathology displaying low-power microscopic view of pilomatrix carcinoma consisting of intradermal tumor lobules made up of basaloid cells transitioning into ghost cells with central necrosis and shadow cells, H&E staining, 20× original magnification (A), high power microscopic view highlighting infiltrating atypical basaloid cells with frequent mitoses fitting the diagnosis of pilomatrix carcinoma, H&E staining 400× original magnification (B).

As it was mirrored in our case, PC typically demonstrates the following clinico-pathological characteristics: (i) tumor diameter often larger than 1 cm, (ii) asymmetry and indistinct borders, (iii) tumor cell nests of unequal size and shape and tend to fuse, (iv) necrotic tissue often seen within the tumor mass, (v) tumors are composed of acidophilic and basophilic cells, but basophilic cells predominate, (vi) basophilic cells display nuclei containing chromatin that is rich and atypical, and frequently exhibit mitotic figures with abnormal nuclear fission, and (vii) lymphocytic and inflammatory cell infiltrates.[8] Definitive diagnosis of PC is established on histological evaluation and at times, it may not be easy especially in small biopsies. Some authors reported diagnosis of these tumors to be somehow subjective particularly with small biopsies.[9] Some cases of PC can be easily confused with pilomatrixoma therefore, lead to under-diagnosis.[8,9] Some of the pilomatricoma cases that show locally aggressive growth pattern were over diagnosed as PC. Unfortunately, immunohistochemistry is not contributory to reliably distinguish between benign and malignant hair matrical tumors. Morphologically, as it was evidenced in the index case, PC is characterized by infiltrative growth pattern. The tumor cells display features of pilomatrixoma (ghost cells and metrical cells) with severe cytological atypia. In addition, the tumor cells are pleomorphic, hyperchromatic basaloid cells with areas of squamous metaplasia and exhibit prominent nucleoli. High mitoses including atypical ones are often encountered along with necrosis, lymphovascular, and perineural invasion.[9–12]

Case reports of PC with brain metastasis have rarely been reported. For instance, Flynn et al.[13] documented a case of recurrent scalp PC in a 24-year-old female with brain metastasis. Despite the prompt neurosurgery and radiotherapy, the patient' final outcome was fatal. Similarly, in their study Toffoli et al.[14] found that distant metastases were uncommon, occurring in 10%–13% of patients, of which two (4%) had metastases at the first tumor staging following primary tumor excision, and six (12%) had metastases during follow-up. Most common sites were lung and regional lymph nodes, followed by local bone infiltration, skull bones, brain, and parotid gland. Only one patient had diffuse metastases to the lung, pleura, liver, and bones.[14] The role of chemotherapy is still debated.[9,12] Different protocols and drugs have been tried, but none of them has been demonstrated to significantly modify the course of this disease. Although the role of radiotherapy has yet to be fully elucidated however, adjuvant radiotherapy following excision of primary as applied in the index case, has been reported to provide adequate local tumor control.[10] Since it is poorly responsive to chemotherapy, it must be treated with a wide surgical excision. The tumor has the tendency to recur locally after incomplete excision and can metastasize to bone, regional lymph nodes, abdominal, and thoracic visceral organs.

4 | CONCLUSION

PC is a rare malignant tumor with high rate of disease relapse. Surgical procedure with wide margins is recommended to avoid recurrence. Given its aggressive nature, the tumor has a high propensity for recurrence after excision. It is important to perform wide local excision to avoid an incomplete resection and higher recurrence rates. Further studies are needed to create a clearer standard of treatment and evaluate the role of adjuvant chemotherapy and radiotherapy.

AUTHOR CONTRIBUTIONS

Kenneth Mlay: Conceptualization; data curation; writing – review and editing. **Peter Shija:** Conceptualization;

data curation; writing – review and editing. **Marco Magwizi:** Data curation; writing – review and editing. **Ezekiel G. Karuga:** Data curation; writing – review and editing. **Philbert Mtenga:** Data curation; writing – review and editing. **Alex Mremi:** Conceptualization; data curation; investigation; writing – original draft; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

All authors have declared that no competing interests exist.

DATA AVAILABILITY STATEMENT

There are no data generated from this study.

ETHICS STATEMENT

The patient provided written informed consent to allow for her de-identified medical information to be used in this publication. A waiver for ethical approval was obtained from the authors' institution review board committee.

CONSENT

Written informed consent for publication of clinical details and images was obtained from the patient.

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REFERENCES

1. Siddha M, Budrukkar A, Shet T, et al. Malignant Pilar tumor of the scalp: a case report and review of literature. *J Cancer Res Ther*. 2007;3(4):240-243. doi:10.4103/0973-1482.39001
2. Papadakis M, de Bree E, Floros N, Giannikaki E, Xekalou A, Manios A. Pilomatrix carcinoma: more malignant biological behavior than was considered in the past. *Mol Clin Oncol*. 2017;6(3):415-418. doi:10.3892/mco.2017.1148
3. Martin S, DeJesus J, Jacob A, et al. Pilomatrix carcinoma of the right postauricular region: a case report and literature review. *Int J Surg Case Rep*. 2019;65:284-287. doi:10.1016/j.ijscr.2019.10.087
4. Sorin T, Eluecque H, Gauchotte G, et al. Pilomatrix carcinoma of the scalp. A case report and review of the literature. *Ann Chir Plast Esthet*. 2015;60(3):242-246. doi:10.1016/j.anplas.2014.06.005
5. Autelitano L, Biglioli F, Migliori G, Colletti G. Pilomatrix carcinoma with visceral metastases: case report and review of the literature. *J Plast Reconstr Aesthet Surg*. 2009;62(12):e574-e577. doi:10.1016/j.bjps.2008.08.024
6. Bremnes RM, Kvamme JM, Stalsberg H, Jacobsen EA. Pilomatrix carcinoma with multiple metastases: report of a case and review of the literature. *Eur J Cancer*. 1999;35(3):433-437. doi:10.1016/s0959-8049(98)00299-8
7. Saussez S, Mahillon V, Blaivie C, Haller A, Chantrain G, Thill MP. Aggressive pilomatrixoma of the infra-auricular area: a case report. *Auris Nasus Larynx*. 2005;32(4):407-410. doi:10.1016/j.anl.2005.07.012
8. Lee & Febiger. Ackerman's Histologic Diagnosis of Neoplastic Skin Disease: A Method by Pattern Analysis. Philadelphia/London 1993, 661–675.
9. Wierzbicka M, Kraiński P, Bartochowska A. Challenges in the diagnosis and treatment of the malignant adnexal neoplasms of the head and neck. *Curr Opin Otolaryngol Head Neck Surg*. 2023;31(2):134-145. doi:10.1097/MOO.0000000000000872
10. Hardisson D, Linares MD, Cuevas-Santos J, Contreras F. Pilomatrix carcinoma: a clinicopathologic study of six cases and review of the literature. *Am J Dermatopathol*. 2001;23(5):394-401. doi:10.1097/00000372-200110000-00002
11. Jones C, Twoon M, Ho W, Portelli M, Robertson BF, Anderson W. Pilomatrix carcinoma: 12-year experience and review of the literature. *J Cutan Pathol*. 2018;45(1):33-38. doi:10.1111/cup.13046
12. Tselis N, Heyd R, Vogt HG, Zamboglou N. Pilomatrix carcinoma with lymph node and pulmonary metastases. *Strahlenther Onkol*. 2006;182(12):727-732. doi:10.1007/s00066-006-1600-2
13. Flynn A, Agastyaraju AD, Sunitha N, Harrison A. Malignant Pilomatricoma: a report of two cases and review of literature. *J Clin Diagn Res*. 2017;11(7):ED27-ED28. doi:10.7860/JCDR/2017/27589.10260
14. Toffoli L, Bazzacco G, Conforti C, et al. Pilomatrix carcinoma: report of two cases of the head and review of the literature. *Curr Oncol*. 2023;30(2):1426-1438. doi:10.3390/curroncol30020109

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