DOI: 10.7860/JCDR/2023/66420.18426

Case Report

Pathology Section

Aggressive Pilomatrixoma of the Scalp in a Geriatric Patient: A Rare Occurrence

RAKESH RAJIV PATKAR[1], VICKY JAIN[2], SHILPA MISHRA[3]

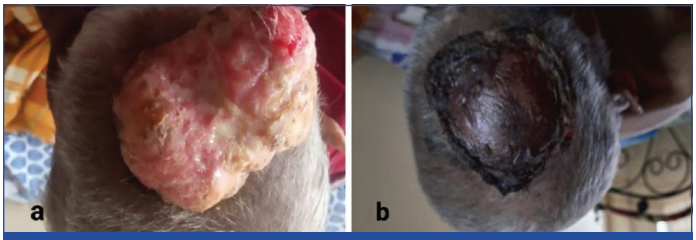
ABSTRACT

Pilomatrixoma is a benign cutaneous adnexal neoplasm that arises from the hair matrix. This lesion is typically observed in the first two decades of life, with most cases occurring before the age of 20. However, it is rare in the elderly. While aggressive and malignant variants of pilomatrixoma exist, they are rarely reported compared to their benign counterparts. In this case, we present an instance of aggressive pilomatrixoma in a 90-year-old female. The patient exhibited a large ulceroproliferative growth on the scalp. A preliminary biopsy was performed, followed by wide local excision. Both the biopsy and excision specimens exhibited characteristics of pilomatrixoma, with a focal infiltrative growth pattern and moderate nuclear atypia. Features indicative of malignancy, such as geographic necrosis, brisk mitosis, atypical mitosis, vascular invasion, and perineural invasion, were not observed. Therefore, the diagnosis of aggressive pilomatrixoma was made. Wide local excision and subsequent skin grafting were conducted, with no involvement of deeper subcutaneous tissue and intact pericranium. The patient is currently faring well during follow-up. This case underscores the importance of meticulous histopathological examination for distinguishing aggressive variants, as they carry a higher risk of recurrence and aid in distinguishing aggressive pilomatrixoma from carcinoma. Failure to recognise the aggressive variant may result in inadequate management.

Keywords: Cutaneous adnexal tumours, Skin grafting, Tumours of hair matrix

CASE REPORT

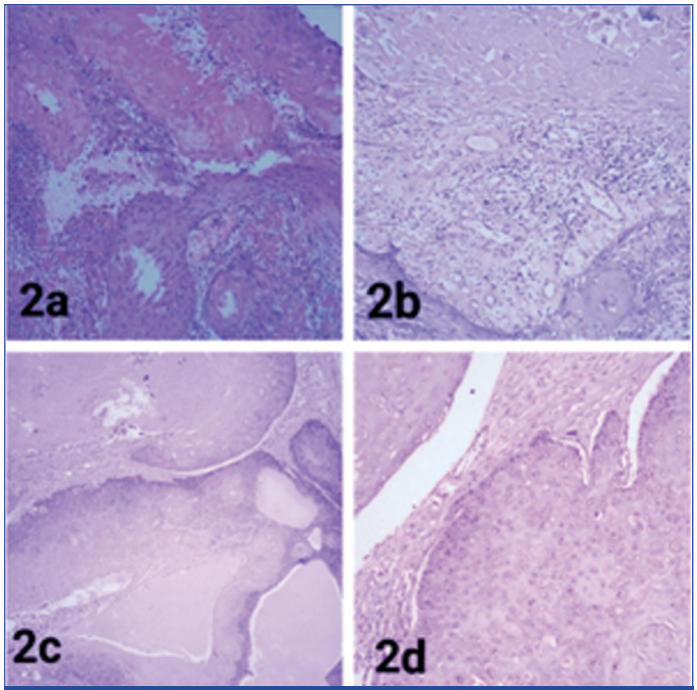
A 90-year-old female patient presented with a slow-growing scalp lesion. She first noticed the lesion approximately 20 years ago. There was a rapid increase in the size of the lesion for 4-5 months, and it started bleeding intermittently with scab formation. She decided to undergo surgery due to the discomfort. Clinically, it appeared as an ulceroproliferative growth [Table/Fig-1a]. The patient did not have any other significant medical or surgical history. Her preoperative clinical examination and routine blood tests did not show any abnormalities. She did not have similar lesions elsewhere in her body or any family history of similar lesions. Initially, a preoperative biopsy was performed, revealing basaloid cells in nests and sheets with abundant cytoplasm and moderately pleomorphic vesicular nuclei. Trichilemmal-type keratinisation was observed. Mitotic activity was minimal, leading to a diagnosis of aggressive pilomatrixoma. There were no features of malignancy. Following the biopsy, a local excision and skin grafting were performed [Table/Fig-1b] as the bone was not exposed and the pericranium was intact. The surgery was done under sedation with a total scalp block.



[Table/Fig-1]: a) Scalp mass with superficial ulceration; b) After wide local excision and skin grafting.

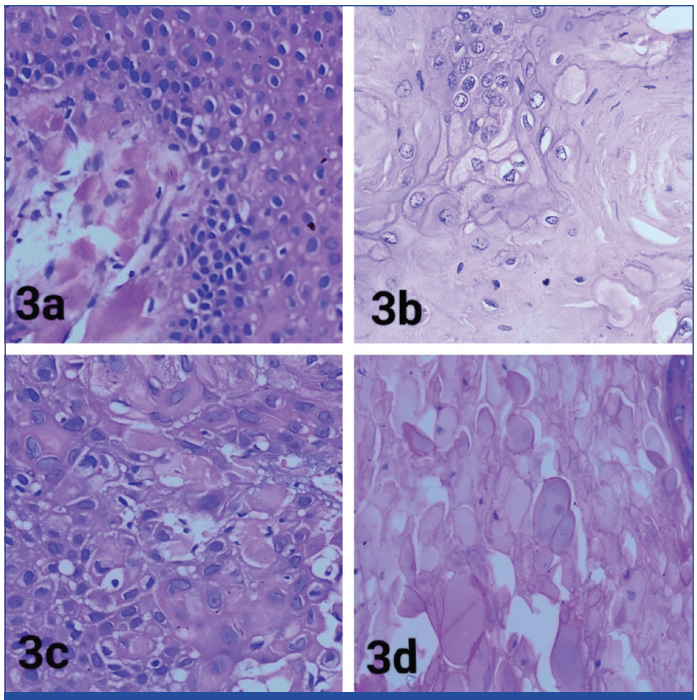
The excision specimen was examined, revealing a large fleshy white mass attached to the scalp skin, measuring 17×14×9 cm. Gross examination showed that the lesion was away from the margins. Representative sections were taken, and slides were stained with Haematoxylin and Eosin (H&E).

Microscopy revealed a partially circumscribed neoplasm. The lesion exhibited an infiltrative pattern in some areas, reaching up to the epidermis [Table/Fig-2]. There were solid nests of basaloid cells in the dermis, undergoing abrupt trichilemmal-type keratinisation [Table/Fig-3a]. The basaloid cells had scanty to moderate amounts of cytoplasm and ovoid hyperchromatic nuclei with minimal pleomorphism. Cells with squamous differentiation displayed abundant eosinophilic cytoplasm and hyperchromatic to vesicular nuclei [Table/Fig-3b,c]. Occasional mitotic activity was observed. Sheets of anucleate ghost cells and calcification were present [Table/Fig-3d]. Prominent nucleoli were noted [Table/Fig-4]. Multinucleated giant cells with moderate mixed inflammation were seen [Table/Fig-5]. Focal necrosis was also observed. Atypical mitosis, perineural invasion, or vascular invasion were not seen. Surgical margins

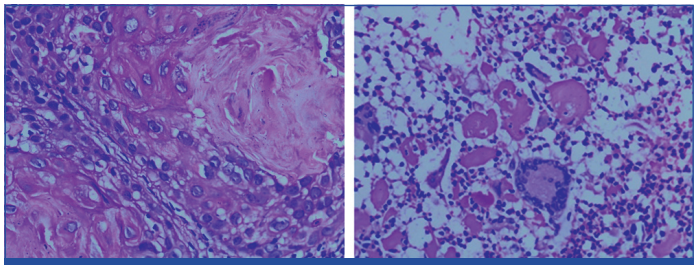


[Table/Fig-2]: a-c) The tumour with infiltrative pattern with surrounding fibro connective tissue (H&E x100); d) Cells arranged in sheets with ample amounts of eosinophilic cytoplasm.

were free from the tumour. Due to the presence of basaloid nests, ghost cells, and abrupt trichilemmal keratinisation, the diagnosis of a tumour originating from the hair matrix was considered. The term "aggressive pilomatrixoma" was assigned as the lesion was larger, locally infiltrative, and exhibited nuclear atypia. The diagnosis of pilomatrix carcinoma was ruled out due to the absence of rapid growth, geographical tumour necrosis, and atypical mitosis.



[Table/Fig-3]: a) The tumour showing sheets of basaloid cells with abrupt trichilemmal type keratinisation (H&E x400); b) Cells with squamoid morphology (H&E x400); c) Cells with moderate nuclear atypia, vesicular chromatin and nucleoli (H&E x400); d) Ghost cells (H&E x400).



[Table/Fig-4]: Cells with vesicular nuclei and prominent nucleoli (H&E x400).
[Table/Fig-5]: Multinucleated giant cell with moderate mixed inflammation (H&E x400).
(Images from left to right)

An expert pathologist reviewed the findings due to the subjective variation in such cases regarding clinical behaviour. The patient was subsequently followed-up and did not show any recurrence for more than two years. This was consistent with the histopathological diagnosis.

DISCUSSION

Pilomatrixoma is a benign dermal adnexal tumour with differentiation toward the matrix of hair, the hair follicle's inner sheath, and hair cortex [1]. This entity was first described as "calcifying epithelioma of Malherbe" by Malherbe and Chenantais in the 1880s. Later, the term pilomatrixoma was used by Forbis R and Helwig EB in 1961 [1]. These lesions mostly occur in children and young adults, with a female preponderance. Forty percent of the cases occur in the first decade, 60% in the second decade, and it is very rarely reported in the elderly [2]. In the current case, the patient was a 90-year-old lady.

They clinically present as solitary lesions. Familial patterns may be associated with multiple lesions, and some cases may have an association with myotonic dystrophy [3]. The current case did not have any other lesions or a family history of such lesions.

Pilomatrixoma was earlier known as calcifying epithelioma of Malherbe. At that time, it was thought to arise from sebaceous glands [4]. Forbis R and Helwig EB described that the neoplasm was derived from primitive basal cells with differentiation towards hair matrix, following electron microscopy and histochemical stains [1].

On clinical examination, pilomatrixomas are small, nodular lesions ranging in size from 5-30 mm and reddish-blue in colour [4]. In the current case, the lesion was much larger than the usual pilomatrixoma. Histopathology reveals predominantly basaloid cells, ghost cells (anucleate shadow cells), nucleated squamous cells, calcification, and foreign body type giant cells [5]. The above features were also seen in the present case.

Pilomatrixoma is a common lesion, and aggressive pilomatrixomas are very rare [5]. The term aggressive pilomatrixoma is used when histopathology shows atypical features and a local infiltrative pattern. However, features of carcinoma, such as abnormal mitotic activity, perineural and vascular invasion, are not identified [4,5]. Comparative features of pilomatrixoma, aggressive pilomatrixoma, and pilomatrix carcinoma are described in [Table/Fig-6]. The novelty of this case lies in understanding the fact that these tumours are part of a spectrum ranging from benign pilomatrixoma to pilomatrix carcinoma [4,5].

Type of lesion and histopathology	Basaloid cells and shadow cells	N:C ratio	Local invasion	Metastasis	PNI and LVI	Central necrosis
Pilomatrixoma	Yes	Low	No	No	No	No
Aggressive pilomatrixoma	Yes	High	Yes	No	No	Yes
Pilomatrix carcinoma	Yes	High	Yes	Yes	Yes	Yes

[Table/Fig-6]: Comparative features of pilomatrixoma, aggressive pilomatrixoma and pilomatrix carcinoma [5].
N:C ratio: Nucleo-cytoplasmic ratio; PNI: Perineural invasion; LVI: Lymphovascular invasion

Aggressive pilomatrixoma is also commonly observed in children [6]. The term 'aggressive pilomatrixoma' was first used by Nield DV et al., [7]. They described a case of an eight-year-old male with recurring lesions. Microscopic examination revealed large, uniform proliferating basaloid cells with brisk mitosis, keratinous nests, a more pronounced infiltrating pattern, perineural invasion, and vascular invasion [7]. In the present case, basal cell proliferation, a focal infiltrative pattern, and nuclear atypia were observed. However, vascular invasion and perineural invasion were not seen. Due to the presence of proliferating basaloid cells, some authors have referred to this variant as 'proliferating pilomatrixoma' [8,9]. Kaddu S et al., described nine cases of pilomatrixoma with distinct histopathological features in elderly patients [8]. In their study, they referred to these lesions as the proliferating variant of pilomatrixoma. The tumours were well-circumscribed, and the basaloid cells exhibited matrical and supramatrical differentiation with nuclear atypia and mitosis [8]. Although this lesion does not meet the criteria for malignancy, the risk of recurrence has been reported [6]. Nadershah M et al., reported a case of giant recurrent pilomatrixoma in which the tumour was unusually large and demonstrated multiple recurrences [10]. However, the present case has not shown any recurrence for more than two years. It is established that the recurrence rate for pilomatrixoma is low (0-3%) [10]. However, data on the recurrence rate of aggressive pilomatrixoma are insufficient, and further studies with regular follow-up are needed [9].

Other histological differential diagnoses for aggressive pilomatrixoma can include pilomatrixcarcinoma, proliferating pilar tumour, and basal cell carcinoma with matricial differentiation. However, with meticulous histopathological examination, features of malignancy can be ruled out [4,5]. In the present case, there was no evidence of comedo-necrosis, geographic necrosis, prominent peripheral

nuclear palisading, stromal clefts, or brisk and atypical mitosis when compared to the above differentials.

The best treatment for aggressive pilomatrixoma is wide surgical excision. The role of radiation therapy remains ambiguous, as not much research has been done in this regard [5]. Considering previous studies, regular close follow-up after wide local excision remains the ideal treatment for aggressive pilomatrixoma [3,5].

CONCLUSION(S)

Stringent histological criteria have not been formulated for the diagnosis of aggressive pilomatrixoma. This lesion represents a spectrum ranging from benign pilomatrixoma to pilomatrix carcinoma. Differentiating it from carcinoma remains a diagnostic challenge and is often subjective, requiring close follow-up and regular communication with the clinician involved in patient care, as well as the patient. Detailed histopathological examination is of utmost importance for distinguishing it from typical pilomatrixoma and pilomatrix carcinoma. The best treatment for aggressive pilomatrixoma remains wide surgical excision, followed by regular patient follow-up.

REFERENCES

[1] Forbis R Jr, Helwig EB. Pilomatrixoma (calcifying epithelioma). Arch Dermatol. 1961;83:606-18.
[2] Simon Cypel TK, Vijayasekaran V, Somers GR, Zuker RM. Pilomatricoma: Experience of the hospital for sick children. Can J Plast Surg. 2007;15(3):159-61.
[3] Missak M, Haig A, Gabril M. Pilomatrixoma with atypical features: A case report. Case Rep Dermatol. 2021;13:98-102.
[4] Gupta M, Bansal R, Tiwari G, Sharma S. Aggressive pilomatrixoma: A diagnostic dilemma on fine-needle aspiration cytology with review of literature. Diagn Cytopathol. 2014;42(10):906-11.
[5] 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):407-10.
[6] Inglefield CJ, Muir IF, Gray ES. Aggressive pilomatricoma in childhood. Ann Plast Surg. 1994;33(6):656-58.
[7] Nield DV, Saad MN, Ali MH. Aggressive pilomatrixoma in a child: A case report. Br J Plast Surg. 1986;39(1):139-41.
[8] Kaddu S, Soyer HP, Wolf IH, Kerl H. Proliferating pilomatricoma. A histopathologic simulator of matrical carcinoma. J Cutan Pathol. 1997;24:228-34.
[9] Kondo RN, Pontello Junior R, Belinetti FM, Cilião C, Vasconcellos VR, Grimaldi DM. Proliferating pilomatricoma--case report. An Bras Dermatol. 2015;90(3 Suppl 1):94-96.
[10] Nadershah M, Alshadwi A, Salama A. Recurrent giant pilomatrixoma of the face: A case report and review of the literature. Case Rep Dent. 2012;2012:197273.

PARTICULARS OF CONTRIBUTORS:
1. Head and Consultant Pathologist, Department of Histopathology, Microcraft-Oncquest Laboratories Limited, Mumbai, Maharashtra, India.
2. Consultant, Department of Plastic Surgery, The Esthetique Clinique, Mumbai, Maharashtra, India.
3. Director, Department of Histopathology, Microcraft-Oncquest laboratories Limited, Mumbai, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Shilpa Mishra,
25, Firuz Ara-East Wing, MK Road, Nariman Point,
Mumbai-400021, Maharashtra, India.
E-mail: mishra.sm012@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Jul 13, 2023
- Manual Googling: Jul 24, 2023
- iThenticate Software: Aug 02, 2023 (5%)

ETYMOLOGY: Author Origin

EMENDATIONS: 5

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Jul 06, 2023**
Date of Peer Review: **Jul 19, 2023**
Date of Acceptance: **Aug 04, 2023**
Date of Publishing: **Sep 01, 2023**