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Original Research Paper

Pathology

PILOMATRIX CARCINOMA- A RARE MALIGNANT HAIR FOLLICULAR TUMOR

Dr. Monika Gathwal	Professor, Department of Pathology, Bhagat Phool Singh Government Medical College Khanpur Kalan, Sonipat, Haryana.
Dr. Chiranjeev Gathwal	Professor and HOD, Department of Radiology, Bhagat Phool Singh Government Medical College Khanpur Kalan, Sonipat, Haryana.
Dr. Ruchi Agarwal	Professor, Department of Pathology, Bhagat Phool Singh Government Medical College Khanpur Kalan, Sonipat, Haryana.
Dr. Tanu Priya*	Post Graduate, Department of Pathology, Bhagat Phool Singh Government Medical College Khanpur Kalan, Sonipat, Haryana. *Corresponding Author

ABSTRACT Pilomatrix carcinoma is an extremely rare, locally aggressive tumor arising from basaloid cells in the hair follicle. Recurrence and distant metastases have been reported, with pulmonary lesions being the most frequent manifestation. Similar to pilomatrixoma, pilomatrix carcinoma typically presents as a nontender, firm dermal swelling and is found most commonly in the head and neck region. Although pilomatrixomas and pilomatrix carcinoma are well-recognized lesions, clinically they are frequently misdiagnosed as other skin conditions. Diagnosis is made by histopathology and when discovered, wide local excision has been shown to have the best results. Here we are presenting a case of 35 year old male who presented with infraauricular swelling which was later diagnosed as pilomatrix carcinoma on histopathology.

KEYWORDS : pilomatrix carcinoma, pilomatrixoma, recur, excision

INTRODUCTION

Hair follicle matrix cells give rise to pilocatrixoma, often referred to as calcifying epithelioma of Malherbe, a superficial benign skin tumor. It was first described in 1880 by Malherbe and Chenantais as a calcified epithelioma of sebaceous origin.(1) Pilomatrix carcinoma (PC) corresponds to the malignant variant of pilomatricoma and is locally aggressive with a high rate of local recurrence after surgical excision.(2) Most frequently observed in the head and neck region, typically occurs in fifth to seventh decades of life and are seen more frequently in males (3:1 ratio). To the best of our knowledge only around 125 cases were reported in literature till date.(3) Given its rarity, a gold standard treatment regimen has not been established, however, wide local excision, with or without adjuvant radiation, is advised by the majority of studies. Although pilomatrixomas and pilomatrix carcinoma are well-recognized lesions, clinically they are frequently misdiagnosed as other skin conditions.(1) The diagnosis of PC is made only by histopathology. Here we are presenting a case report of pilomatrix carcinoma in 35 year old male.

CASE REPORT

A 35 year old male presented with slowly progressive painless mass in right infraauricular region since 8 months. On examination, the swelling is well defined, soft to firm in consistency, mobile and non tender. Complete blood count, serum electrolytes, and biochemical profile were normal. On ultrasound, a 3x1.8cm hypoechoic lesion with well circumscribed margins seen in subcutaneous plane. On MRI, there is evidence of a 2.6x2.1x1.8cm sized T2/ STIR heterointense and T1 hypointense predominantly solid mass lesion seen arising from posterior right lateral subcutaneous fascia in the infraauricular region. Clinical diagnosis of salivary gland neoplasm was made. **FNACsmear shows neutrophils, lymphocytes and cluster of round shaped cells with basophilic cytoplasm in an amorphous background. Impression of inflamed cystic lesion was made.** Patient underwent excision biopsy and tissue submitted for histopathological examination. We received a grey brown globular soft tissue measuring 4x3x1.5cm and cut section is grey white and soft to firm in consistency. Microscopically, tumor composed of shadow cells , giant cells, calcification and nests of atypical basaloid cells with increased mitotic activity and focal areas of necrosis. Histomorphological features are suggestive of PC. Immunohistochemical expression is positive for CK5 and CK6 and increased activity of Ki67 is seen. No cervical lymphadenopathy and metastasis is noted. Hence, patient is kept on follow up.

DISCUSSION

PC is a rare malignant and locally aggressive hair follicle tumor with a high propensity for recurrence after excision. The genesis of these lesions—whether they develop from scratch or as a result of a benign pilomatrixoma turning malignant—remains largely unexplained. Malherbe and Chenantais initially defined pilomatrixoma in 1880 as a "calcifying epithelioma" that was assumed to originate from the sebaceous gland. Gromiko's 1927 description is the earliest documented account of the malignant counterpart. A literature review by Christopher et al. examined 125 recorded cases of pilomatrix carcinoma. Of those, 10 arose in a background of histologically confirmed benign pilomatrixoma.(2)

Both the clinical and histological distinctions between benign and malignant pilomatrixoma are challenging given their similar presentation and cytological constituents, and the lack of immunohistochemical markers to differentiate them.(1) When comparing benign and malignant pilomatrixomas, both can be well-circumscribed with nucleated basaloid cells at the periphery transitioning to anucleated shadow or ghost cells in the center. Both may exhibit foreign body giant cell reactivity and calcification, which indicate a granulomatous reaction to the keratinized material. One distinguishing feature of pilomatrix carcinoma is the proliferation of hyperchromatic basaloid cells with pleomorphism, accompanied by regions of squamous metaplasia, frequent mitoses, and large nucleoli. Pilomatrix carcinomas also have an infiltrative growth pattern and can have necrosis and lymphovascular invasion.(4)

Due to the rarity of pilomatrix carcinoma, there are no well defined standards for the surgical management of this malignant lesion.(1) Herman et al, their series of cases reported a 23% recurrence rate in tumors that were widely excised, in contrast to an 83% recurrence rate in tumors that were simply excised. Hence, wide local excision is the most frequent therapeutic option due to the relative high recurrence

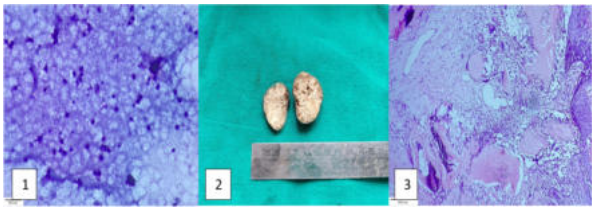
rate reported in the literature.(5)

Distant metastases have been reported in approximately 10% of cases, usually to the lung and regional lymph nodes and rarely to bones and viscera. Since neither chemotherapy nor radiation has been proven to significantly alter the course of the disease, their respective roles are still up for debate. Adjuvant radiation therapy is advised to be saved for cases of recurrent illness or remaining macroscopic disease.(1)

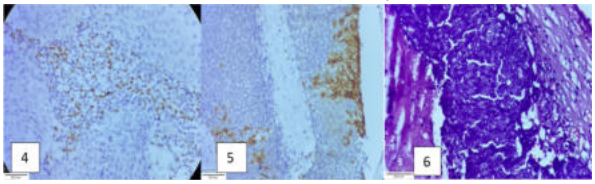
The diagnosis of the suspicious lesion and the initial treatment strategy are the primary factors that determine the overall outcome in PC. Due to the high frequency of local recurrence and the risk of metastasis, it is recommended that patients with confirmed cases of PC be scheduled for total-body skin examination 2 to 3 times annually.(5)

CONCLUSION

It is important to be aware of PC as it can arise from pre-existing pilomatricoma, hence every case should be sectioned properly to look for PC. Wide local excision is recommended. Close follow up of patients after excision of PC is advised in order to detect eventual recurrence and metastasis.



1. Pap smear shows inflammatory cells in amorphous background (100x)
2. Gross specimen- cut surface is grey white with areas of necrosis and calcification
3. Tumor composed of shadow cells, trichelemmal type of keratinization, calcification and atypical basaloid cells with mitosis and focal areas of necrosis.(H&E, 40x)



4. Membranous positivity for CK5/6 (400x)
5. Increased Ki67 (400x)
6. Atypical basaloid cells (H&E, 400x)

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