Huge Piolomatrix Carcinoma on the Scalp: Case Report and Review of the Literature

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Abstract: Pilomatrix neoplasm is a proliferative that is related to the hair follicles (matrix). It is considered one of the cutaneous adnexal tumors.(1) Although there are many reports document that pilomatrix neoplasm can be presented in different parts of the body, as in the scalp, different parts of the face, supraclavicular region, back, spine, upper extremities, vulva, clitoris & knees. The head and neck are considered as the most common site in 60% of patients.(2) Diagnosis of such cases is not easy to be reached clinically due to the variety of the differential diagnoses, which requires a histological evidence to be established and to base on it the optimal management.
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1. Introduction:

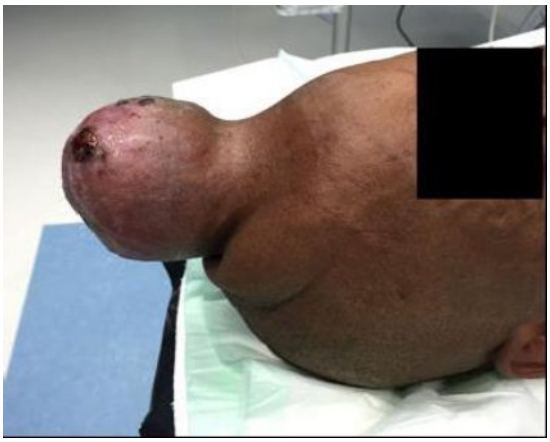
Pilomatrix neoplasm is a skin tumor originated from basaloid cells of the hair follicles. Histopathologically, pilomatrix neoplasm is one of the follicular tumors that are addressed under the cutaneous adnexal tumors. Pilomatrixoma is a common head and neck neoplasm among children. On the contrary pilomatrix carcinoma is rare and its poor prognostic metastatic grade is exceptional to be found, with high risk of local recurrence. This type of carcinoma presented with asymptomatic masses involving the dermal and subcutaneous tissues.(3, 4)The patients are usually in the middle age with 4:1 male: female incidence ratio. (5)

Our report is about the first case to be published in Saudi Arabia of a 37-year-old male Ethiopian patient presented with frontal scalp swelling.

2. Case Report:

A 37 years old Ethiopian male shepherd, medically free presented with painless frontal scalp mass for 5 months ago.**(Figure 1&2)** The mass started as a small nodule with progressive increase in size, Associated with Headache and blurred vision. There was no history of previous trauma or weight loss. Other systemic reviews were unremarkable. Patient had history of brucellosis one year ago and treated medically.

On physical examination: There is frontal scalp swelling 5x10x15 cm circular in shape. Surface looks nodular and ulcerated in the anterior aspect of the mass 1x1 cm with discharging serous fluid from it. The swelling is highly mobile, soft in consistency. Other systems were unremarkable.



(Figure 1)

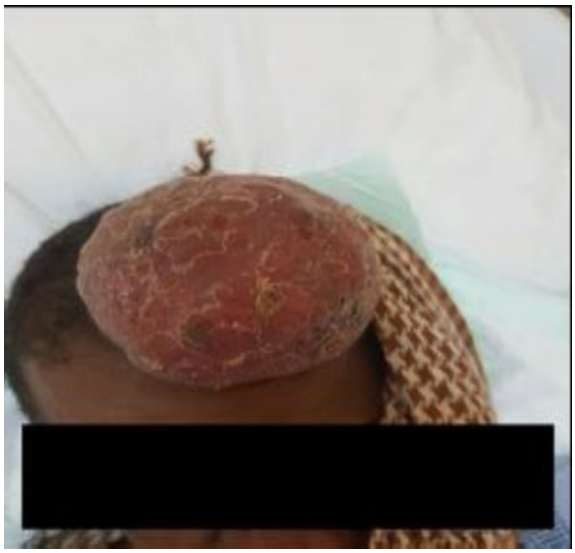
3. Discussion:

Pilomatrix carcinoma is a rare tumor of the hair follicles, where head and neck represents 60% of the reported cases in the scalp, cervical, frontal, temporal, peri-orbital, parotid, mandibular, submental and peri-oricular regions. The pilomatrix neoplasms are classified into benign and malignant tumors, which had been referred to by different terms. The benign class of the tumor was the first one to be reported about in 1880 by Malherbe and Chenantais, as a calcifying epithelioma and they conclude that it is originated from the sebaceous gland. But in 1949 there was another suggestion that it originates from the hair matrix cell by Lever and Griesemer. In 1961, the benign class was termed for the first time by pilomatrixoma by Forbis and Helwig.(3) Another term found in the review was pilomatricoma and calcifying epithelioma of Malherbe. However, in 1927, Gromiko reported the first case of pailomatrix neoplasm with aggressive behavior. Then a similar case was

presented to Lopansri and Mihmin 1980, and a literature review was reported and they termed it as calcifying epitheliocarcinoma of Malherbe or pilomatrix carcinoma.(4, 6)

It usually presents as painless mass, starts as a small nodule that is irregular, growth maybe slow or rapid with local invasion and can metastasize to lungs, bone and abdominal viscera with high recurrence rate 46-60%.(7)

Our case presented with painless frontal scalp mass for 5 months ago, the mass was progressively increased in size, associated with headache and blurred vision. No history of fever, weight loss, nor previous history of trauma.

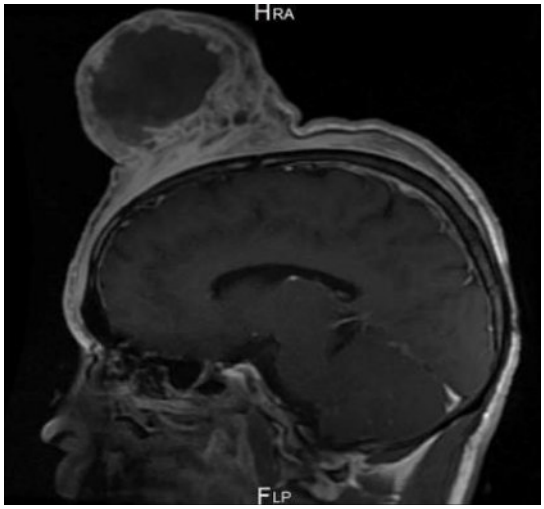


(Figure 2)

Patient was admitted as a case of sebaceous cyst for excision, after further investigations, CT head was done first and reported that part of the mass shows soft tissue and cystic component with calcifications matrix no bony destruction or intracrinal extension. The differential diagnosis includes dermatofibrosarcoma protuberance, soft tissue sarcoma or fibroma for further evaluation by MRI. Then MRI was done and showed large soft tissue mass seen below the scalp in the frontal region superiorly, and measures 9.5 x 8.5 x 7 cm in size. Hemorrhages were noticed. Lesion is well defined and limited to the scalp. No intracranial extension noted. **(Figure 3)**

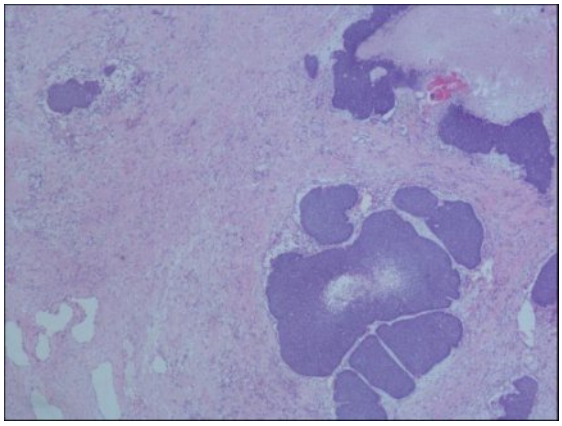
Biopsy was considered but since it was highly hemorrhagic, we progressed into surgical excision with safety margin of 1 cm and the mass was sent for histopathology evaluation.

CT neck, chest, and abdomen were done in order to rule out metastasize.

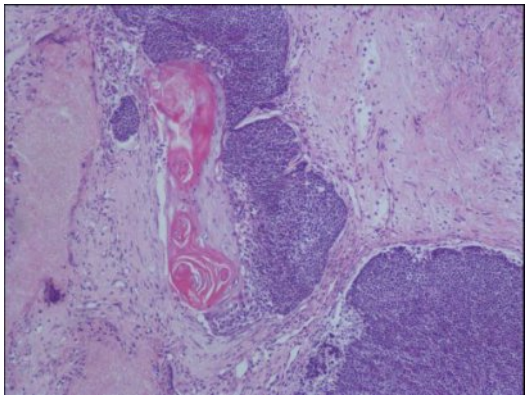


(Figure 3)

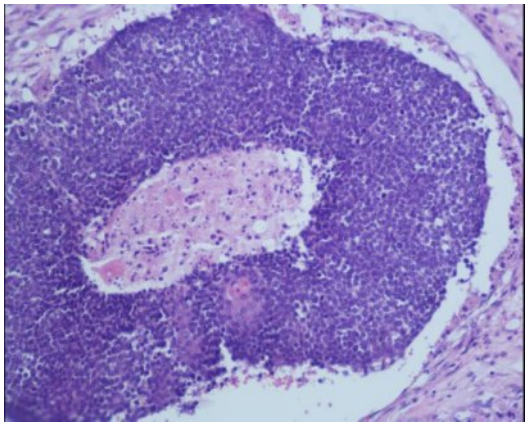
(Figure 4)



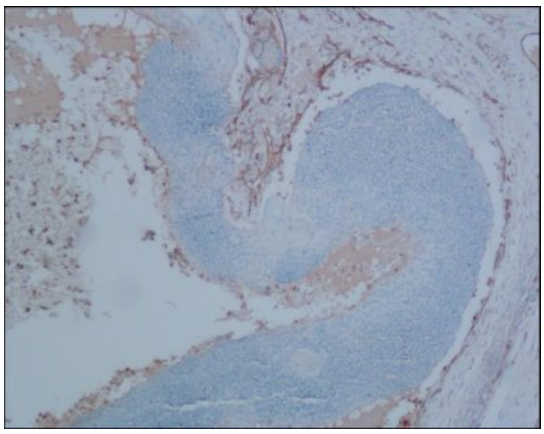
(Figure 5)
Infiltrative nests of Neoplastic Basaloid Epithelium



(Figure 6)
Focal Squamous Differentiation & Keratinization



(Figure 7)
Central Necrosis in doughnut shaped infiltrative nest



(Figure 8)
Vascular Invasion endothelial lining highlighted by CD31 Immunization

MRI: Showing large soft tissue mass involving the frontal region well defined limited below the scalp. No breech of the cranial vault. No intracranial extensions were noted.

Histopathology report was:

Gross findings:

Dome-shaped, fungating soft large forehead mass. **(Figure 4)**

Microscopic findings:

Histopathology reported that there is infiltrative growth of highly proliferative masses and nests of basaloid epithelium. **(Figure 5)** The neoplastic epithelium is matted with shadows of squamous epithelium and showing focal squamous cell differentiation with prominent eosinophilic nucleoli. **(Figure 6)** Large central hemorrhagic ischemic-type necrosis. **(Figure 7)** Vascular invasion was noted. **(Figure 8)**

The basal cutting surface is totally involved by tumor infiltration an advised adequate safety margin to be excised as possible from the skin edge and bed of tumor.

So rextensive excision was done up to the periosteum with safety margin more than 1 cm all around. Wound was closed by flap covered with the split thickness skin graft taken from the left inner upper arm.

Biopsy was taken again in order to investigate for negative margin.

4. Conclusion:

This case presentation of pilomatrix carcinoma/ malignant pilomatrixoma represents an offensive type of malignant carcinoma with low grade, aggressive biological behavior and infiltrative nature, in addition to its tendency of recurrence and metastasis. Must carefully manage this type of carcinoma by taking adequate safety margins. Also, we conducted that the benign ”Piolmatrixoma /calcifying epithelioma of Malerlabe ’’is not just a hematoma, but a real neoplasm as many cases reported as ‘piolomatrix carcinoma’ already started with small long standing nodule and this evolution could be happened in our patient. We prefered to name the tumor... of the benign counterpart. The aim of this study is to emphasize the need for more studies to be conducted and cases to be reported for better understanding of the disease presentation, investigation, management and prevention of future metastasis.

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