Case Report

Beyond the Matrix: A Rare Case of Pilomatrix Carcinoma with Histologic Insights

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Abstract

Pilomatrix carcinoma is rare entity which arises from matrical cells of hair follicle and misdiagnosed quite often. A 27 years female with presented with a swelling on right side of nape of neck for approximately 3 years which was painless, slowly progressive, soft to firm in consistency. Previously misdiagnosed as squamous cell carcinoma, the lesion recurred and was excised again in our institution and was diagnosed here as Pilomatrix carcinoma with regional lymph node metastasis. Since she had metastasis, the patient is currently undergoing radiotherapy. Considering the rarity of this carcinoma it was noteworthy to report this case as correct diagnosis aids patient's treatment protocol and improves prognosis.

Keywords: Pilomatrixoma; Pilomatrix carcinoma; Matrical cells; Skin adnexal tumor

Introduction

A rare carcinoma called *Pilomatrix carcinoma* develops when matrical cells in growing hair proliferate abnormally. It is an uncommon malignant form of *Pilomatrixoma* that is a locally aggressive tumor. The most common location of presentation is the head region and neck region, and a bimodal age distribution is observed with a female predisposition. There is no established treatment plan for the condition because the tumor is extremely uncommon. Nonetheless, Moh's micrographic method surgery has been regarded as the gold standard. We describe a 27-year-old woman who had recurrent neck swelling and was diagnosed *Pilomatrix carcinoma*.

Case Report

A 27-year-old female had a past history of lump on the right side of the nape of her neck, measuring approximately 3×2 cm, which had been slowly increasing in size over the past three years which was excised and was reported as metastatic squamous cell carcinoma in another tertiary care center. She later presented to our outpatient department with a recurrent swelling at the same location for 3 months after her excision of the previous lump. On examination, the swelling was firm, painless, mobile, and measured around 3×3 cm, with no attachment to the overlying skin or deeper tissues. The overlying skin did not show any significant changes. Right posterior cervical lymph node was also palpable. A contrast-enhanced CT scan of the head and neck was done which also showed multiple right level V lymphadenopathy which was suspicious of malignancy [1]. The patient subsequently underwent a wide local excision along with a right neck dissection.

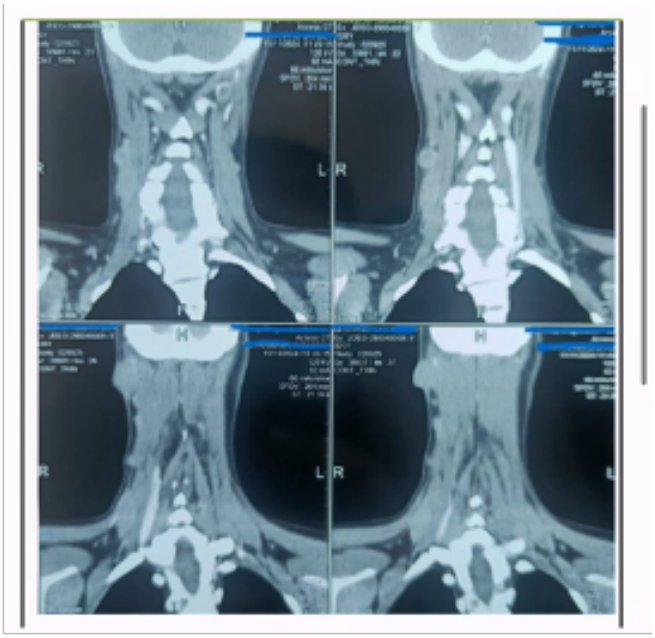

Figure 1: CECT head & neck showing neck swelling and neck nodes.

For histopathological examination we received a skin attached grey, white soft tissue. On sectioning the cut section showed round ill circumscribed grey, white lesion [2]. The margins appeared grossly free. On microscopic examination, the dermis

Figure 2: Gross picture of the lesion.

showed multiple poorly circumscribed and asymmetrical neoplasm. The tumor cells were arranged in organoid pattern with few showing central areas of necrosis [3]. These nests were separated by fibrous stroma. The neoplastic cells comprised of solid aggregates of immature basaloid cells extending up to the subcutaneous tissue. Cystic spaces filled with keratinous

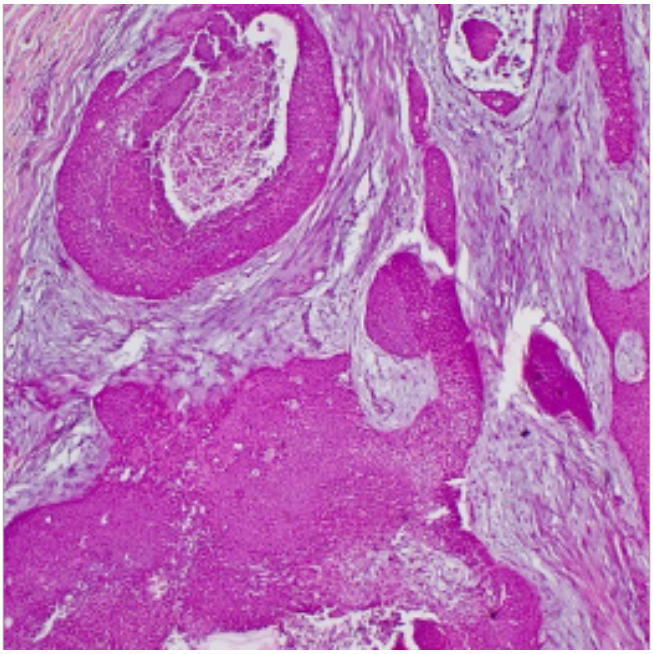

Figure 3: 10X, H&E showing tumor cells arranged in organoid pattern with central areas of necrosis.

material were observed between clusters of matrical cells. Mitosis was 10-12/10 HPF. There was no lymphovascular or perineural invasion seen in the sections examined from the main tumor and a diagnosis of *Pilomatrix Carcinoma* was made. All the margins were free and the tumor was 0.1 cm away from the base. 4 lymph nodes out of 15 lymph nodes submitted which were positive for malignancy. Immunohistochemistry was not done for the given case and the diagnosis was mostly based on the morphology.

Given the recurrent nature of the disease and involvement of lymph nodes, the patient was planned for adjuvant radiotherapy to the local site and regional neck nodes, as recommended by the multidisciplinary team. Unfortunately we could not get the follow up details on the patient after starting the therapy. This case is noteworthy due to its rarity and young age of presentation which often leads to diagnostic challenges.

Discussion

Since the initial reported in 1980, only approximately 206 cases have been documented in the available literature to date [1]. As there are no IHCs markers to distinguish this entity from their benign counterparts, the diagnosis of *Pilomatrix carcinoma* relies entirely on histopathological evaluation. Although their age of presentation is bimodal, it peaks during the fifth and seventh decade with head-neck region as the most frequent place [2]. It is unclear if pre-existing *pilomatrixomas* undergo malignant transformation or if *Pilomatrix carcinoma* arises de novo. This is sustained by the observation that many tumours classified as *Pilomatrix carcinoma* have a lengthy phase of latency before suddenly displaying rapid growth [2].

Wide local excision (WLE) is the most widely used treatment approach; nevertheless, Mohs micrographic surgery may offer lower risk of metastasis and recurrence [1].

Pilomatrix carcinoma often recurs locally, although it can also spread to other places. Myotonic dystrophy and a number of hereditary disorders, including xeroderma pigmentosum trisomy 9, Turner syndrome, Rubinstein-Taybi syndrome, Soto syndrome and Gardner syndrome are linked to multiple *pilomatrixomas* [3]. *CTNNB1* mutations seen on exon 3 which encodes *beta-catenin*, have been discovered in genetic studies of this entity. *Beta-catenin* affects the proliferation and differentiation of cells but its specificity is not yet determined [4]. There have been reports of distant metastases to the brain, liver, pancreas heart, and lungs [5].

Benign *pilomatrixoma*, in contrast to *Pilomatrix carcinoma*, is well circumscribed, composed of a mixture of basaloid cells transitioning to abundant shadow cells, with low mitotic activity and no infiltrative borders, necrosis, or perineural/vascular invasion. Squamous cell carcinoma (SCC) lacks matrical differentiation and does not contain shadow cells; instead, it shows keratin pearls, intercellular bridges, and marked keratinization, often with pronounced stromal desmoplasia. Thus, the key distinctions are shadow cells (present in benign and malignant matrical tumors, absent in SCC), infiltrative atypical basaloid proliferation with high mitoses (*pilomatrix carcinoma*), and well-circumscribed low-grade morphology (*pilomatrixoma*).

In cases that are enduring repeated surgical excision, radiation treatment can be employed as an adjuvant. According to Tselis et al., *Pilomatrix carcinoma* can be effectively treated with both external beam and interstitial brachytherapy [6].

Conclusion

Due to the possibility of distant metastases, pathologists and clinicians should be discerning about the existence of *Pilomatrix carcinoma*. *Pilomatrix carcinoma* is a malignant tumor of hair matrix cell origin showing bimodal distribution. It is commonly misdiagnosed preoperatively due to lack of clinical suspicion and rarity of the disease. However, the lesion can be properly treated if it is better understood. In our case, the lesion was diagnosed correctly which lead to proper treatment of the patient. This case report will open up new doors for further studies.

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