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Malignant pilar tumor of the scalp: A case report and review of literature

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ABSTRACT
Pilar tumor is a rare neoplasm arising from the external root sheath of the hair follicle and is most commonly observed on the scalp. These tumors are largely benign, often cystic, and are characterized by trichilemmal keratinization. Wide local excision has been the standard treatment. Recent reports have described a rare malignant variant with an aggressive clinical course and a propensity for nodal and distant metastases which, therefore, merits aggressive treatment. In this report, we present a case of malignant pilar tumor of the scalp with multiple nodal metastases at presentation. Diagnostic and therapeutic considerations, in the form of adjuvant radiotherapy, are subsequently discussed.

KEY WORDS: Diagnosis, histopathology, malignant pilar tumor, management, radiotherapy, surgery

Case Report

A 50-year-old lady presented with a gradually increasing swelling over the scalp which was first noticed 3 months ago. It had occurred spontaneously and was not associated with any history of trauma; there was no past history of a similar tumor anywhere in the body. It was a painless, nontender swelling, with no associated hair loss, skin changes, or other symptoms. Initial biopsy from the lesion done at the referring hospital was suggestive of squamous cell carcinoma. She was referred to our center for further evaluation and treatment.

On examination, she had a 7 × 7 cm fungated growth on her scalp, over the left parietooccipital region, with ulceration of the overlying skin. The tumor was not fixed to the underlying skull or periosteum. Multiple enlarged, discrete, mobile, soft, and nontender lymph nodes were palpable bilaterally in the suboccipital region and left upper cervical region.

Earlier reports classified this tumor as pseudop epitheliomatous hyperplasia rather than as a neoplasm. However Mann et al. reported metastasis from pilar tumors and hence considered it to be a genuine neoplasm. In addition, a few other reports have also described a rare malignant variant of the pilar tumor, with an aggressive clinical course and a propensity for nodal and distant metastases. However, metastasis from pilar tumors is rare. It has been reported after treatment of the primary. Recently, proliferating pilar tumors have been classified into three groups: benign, locally aggressive, and malignant.

Due to the rarity of this histopathological entity there are no guidelines available for the management of these tumors. Till date, the standard treatment has been wide local excision. The role of radiation therapy and chemotherapy, especially in the malignant variant, is not established. Most of the case reports till now have focused on the pathological aspects, with minimal emphasis on clinical behaviour and management issues.

We report a patient who was diagnosed as having a malignant pilar tumor of the scalp and had metastases to the lymph nodes at presentation. The aim of this report is to create awareness regarding this particular entity, considering its rare presentation and the need to differentiate these tumors from squamous cell carcinoma of head and neck region.

CASE REPORT

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Contrast-enhanced computerized tomography (CT) scan of the head and neck region revealed a heterogeneously enhancing, extracranial, subgaleal, high parietal, soft tissue swelling without any obvious bone erosion. Magnetic resonance imaging (MRI) showed a 5.2 cm scalp tumor infiltrating the deepest layer of the scalp, with a thickness of 2.5 cm. The outer table of the skull was not eroded. She was evaluated in the head and neck joint clinic, where the decision for surgical management was made by a surgeon, radiation oncologist, and medical oncologist.

A wide excision of the scalp tumor along with posterolateral neck dissection on both sides was performed under general anesthesia. Peroperatively, the tumor was firm to hard in consistency, with fungation of the overlying skin; it was moderately vascular and well demarcated, with discrete borders. It was free from the underlying skull and periosteum. Complete surgical excision was achieved. Reconstruction of the defect was done with a skin flap from the left thigh.

Histopathological evaluation showed a variegated tumor with distinct areas displaying characteristic histopathological features. The tumor was 6.7 × 6.5 cm in size, with a thickness of 3.8 cm [Figures 1-3]. The major portion of the tumor was formed by small cells with a high nucleocytoplasmic ratio arranged in confluent nodules. Most of these nodules demonstrated central keratinization. The nuclei were hyperchromatic and pleomorphic and several multinucleated tumor giant cells were present. Mitosis was frequently noted, indicating the malignant nature of the tumor. Also notable was a distinct area, resembling a typical benign pilar tumor, composed of interlacing nodules of smaller peripheral cells that were palisaded and matured into larger central cells with central keratinization. The final histopathological diagnosis was of a malignant pilar tumor. All the cut margins were free and the base was 1 mm away from the tumor. Lymphovascular and perineural invasion were absent. Four out of seventy-three lymph nodes sampled in the neck dissection of the left upper cervical region showed metastases from the malignant pilar tumor with perinodal extension. In view of the malignant nature of the tumor and the multiple lymph node involvement with perinodal extension, she was considered for adjuvant radiotherapy to the primary tumor and neck nodal region after discussion in the multidisciplinary clinic.

After healing of the surgical wound, adjuvant radiation was delivered to a localized area of the scalp covering the original site of the tumor with 2 cm margins, using 9 MeV electrons with an en face electron portal after assessing tumor thickness from the preoperative MRI. The left neck was treated with mixed 6 MV photons and 9 MeV electron beams. A total dose of 6000 cGy was delivered over 30 fractions in 50 days, treating once daily for 5 days a week. The patient tolerated the treatment well, having only mild hyperpigmentation over the treated regions along with mild dysphagia as acute reactions during the course of treatment. Treatment was completed without any gap and the patient was advised...
DISCUSSION

Tumors of adnexal components of the skin are exceptionally rare and were first reported in 1966 by Wilson-Jones as an entity which can simulate squamous cell carcinoma. They are also known as tricholemmomas or trichoclamydocarcinomas (or invasive hair matrix tumor). These tumors are more common in women and most scalp lesions are benign. Lanugo hair follicles of the bald scalp and follicles of other areas devoid of non-terminal hair are unlikely to produce these tumors. Therefore pilar tumors are not seen in the bald scalp, being more common in areas with excess hair growth. These pilar tumors arise through increased epithelial proliferation within pilar or sebaceous cysts. Hence, patients frequently give a history of a long-standing cyst at the same site.

Previous reports of trichilemmal tumors have been infrequent, comprising only 0.1% of skin biopsies, and the malignant variety is even rarer. Despite the characteristic histological features of these tumors, earlier reports have emphasized the need to differentiate this entity from well-differentiated carcinoma. Confirmation of the diagnosis is made on the histological appearance of the clear cells, which exhibit marked pleomorphism and atypical mitosis, and its origin from the outer root sheath of hair. Characteristically nodular trabecular formation of keratinocytes with a sharp border between parenchyma and stroma is seen. Malignant change in pilar tumors conforms to two growth patterns: circumscribed nodular and diffuse spindle cell type. Malignant pilar tumors need to be differentiated from metastases of spindle cell tumors with regions of squamous differentiation which tend to mimic their histopathological appearance. Differential diagnosis includes sebaceous carcinoma, clear-cell hidradenocarcinoma, and cutaneous metastasis of renal cell carcinomas.

The criteria of malignancy of these solid tumors of the scalp vary with different authors. In the majority of the cases reported earlier, the tumor had been present for many years before progression. Metastasis from malignant pilar tumors has only rarely been reported. Seff and Berkowitz reported two cases which metastasized several times to neck nodes, the patient subsequently dying of disseminated disease. Some authors tried to predict the behavior of the pilar tumor on the basis of the invasive nature of the tumor. Jay studied 76 patients and categorized them into three groups based on histopathological characteristics. Group I included those patients who showed no infiltration of surrounding stroma and minimal nuclear atypia; they showed trichilemmal keratinization, stromal invasion with a mononuclear infiltrate of plasma cells and lymphocytes, and dystrophic calcification and were considered as benign tumors. Group II were early invasive, with modest cytological abnormalities, eosinophilic cytoplasm, and loose edematous stroma with minimal to moderate infiltration by mononuclear inflammatory cells; they were considered locally aggressive. Group III tumors were invasive; they were cytologically anaplastic and were therefore considered malignant. Statistically significant difference in terms of local control and lymph node metastatic potential was found between the groups in their study.

In our patient, the diagnosis was clinched with the identification of the characteristic histopathological features. However, this case was unusual for its aggressive behavior since the tumor demonstrated involvement of multiple regional lymph nodes at presentation. Meta-analysis of 185 patients of 8 reported series confirmed the presence of the aggressive variant of the malignant pilar tumor, which has tendency to recur and metastasize distantly. All patients with aggressive tumors in these 8 series were not given any type of adjuvant treatment, which led to an unfavorable treatment outcome. Similarly, in the series by Ye et al., 4 out of 8 patients with malignant pilar tumors recurred within period range of 2-14 months. All these patients were not given any adjuvant treatment. Based on this evidence, we decided to give adjuvant radiotherapy to the operated tumor bed so as to achieve favorable locoregional control.

The role of adjuvant radiation therapy in pilar tumor, especially in the malignant variant, is not very clear. This is mainly due to the rarity of the disease. However, considering the aggressive nature of the malignant variant and high rates of locoregional as well as distant failures in previous series, adjuvant radiotherapy is justified.

Unlike head and neck squamous cell carcinomas which are locoregional, pilar tumors are primarily local only. For this reason they can be managed with wide local excision. The clinical presentation of the tumor as a large fungated mass and the presence of multiple histopathologically-proven metastatic nodes with perinodal extension suggested that a select subset of these rarely malignant neoplasms merit aggressive treatment. The case reported here documents unequivocal regional metastases from a clearly malignant tumor. Further study of similar tumors belonging to the malignant variant subset of pilar neoplasms is necessary to determine whether the addition of adjuvant radiotherapy improves locoregional control in this rare variant.

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