Article is available online at http://www.webio.hu/por/2006/12/4/0251

CASE REPORT

Malignant Pilomatricoma in the Parietal Area

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A 27-year-old Japanese woman presented with a 2.5-cm nodular subcutaneous lesion in the parietal area. The nodule was well demarcated and situated in the dermis and subcutis. Histologically, the tumor was diagnosed as malignant pilomatricoma. The tumor was excised, the postoperative course was uneventful, no evidence of local recurrence or distant metastasis was observed, and the patient continues to be under close follow-up. Malignant pilomatricoma, a locally aggressive counterpart of benign pilomatricoma, is also referred to as pilomatrix carcinoma. Most cases are excised as benign tumors; however, when the excision is incomplete local recurrence is likely, and distant metastases have also been reported. Histologically, the diagnosis can be challenging because no clear histologic criteria are available. Because of the rarity of malignant pilomatricoma, no well-defined standards in the surgical management of this neoplasm have been established. Moreover, since distant metastases have been described, close follow-up of the lesion is requisite. (Pathology Oncology Research Vol 12, No 4, 251–253)

Key words: malignant pilomatricoma, pilomatrix carcinoma, basophilic cells, distant metastasis

Introduction

Malignant pilomatricoma,[6] a locally aggressive counterpart of benign pilomatricoma, is also referred to as pilomatrix carcinoma. Most cases are excised as benign tumors; however, when the excision is incomplete local recurrence is likely, and distant metastases have also been reported.[4,10] Herein, we present a case of malignant pilomatricoma in the parietal area and discuss it.

Case report

A 27-year-old Japanese woman presented with a 2.5-cm nodular subcutaneous lesion in the parietal area. A physical examination demonstrated no lymph node involvement. A chest radiograph and computed tomography of the head and chest showed no findings suggestive of metastases. The clinical diagnosis was schwannoma. The resected specimen showed a 2.5 cm, well-defined, soft tan dermal and subcutaneous mass containing pale caseous material. The nodule was well demarcated (*Figure 1*), situated in the dermis and subcutis, with a predominantly pushing margin. On the other hand, smaller cords of tumor cells also infiltrated the stroma in the border region of the tumor (*Figure 2*).

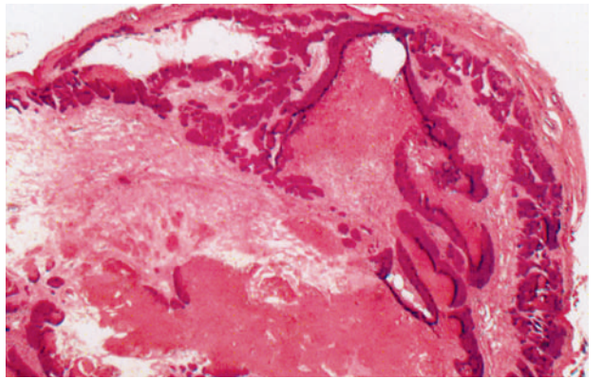

Figure 1. *On scanning magnification, the nodule is well demarcated. (HE staining, x20)*

Received: Jan 11, 2006; *accepted:* Sept 15, 2006

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Toward the center, the tumor showed abrupt transition from basophilic cells to anucleate, eosinophilic ghost cells (*Figure 3*). Poorly-differentiated basophilic cells showed prominent nucleoli, nuclear pleomorphism, and frequent abnormal mitotic figures (*Figure 4*). Histologic analyses revealed no definite vascular or lymphatic invasion, and no direct attachment to the epidermis was observed. Consequently, the tumor was diagnosed as malignant pilomatricoma. The postoperative course was uneventful, the patient showed no evidence of local recurrence or distant metastasis after the initial excision, and continues to be under close follow-up.

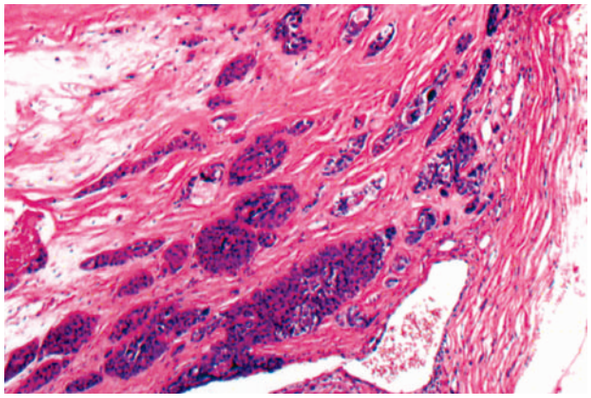

Figure 2. *In the border region of the tumor, stromal invasion is identifiable. (HE staining, x100)*

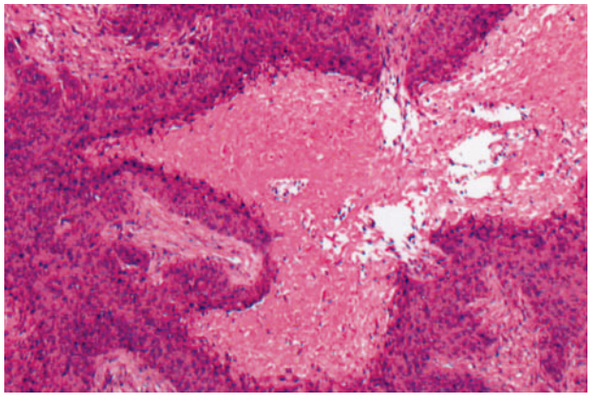

Figure 3. *The tumor shows abrupt transition from basophilic cells to anucleate, eosinophilic ghost cells. (HE staining, x100)*

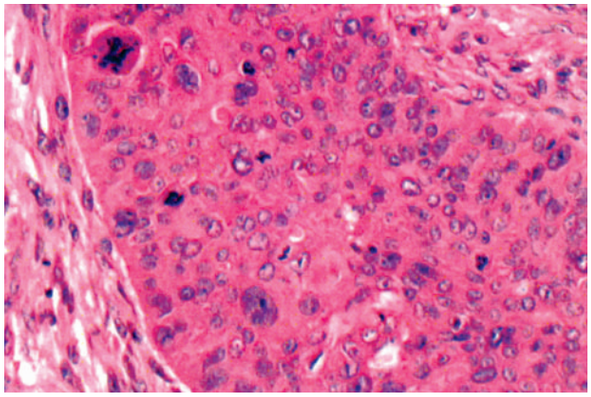

Figure 4. *Poorly-differentiated basaloid cells show prominent nucleoli, nuclear pleomorphism, and frequent abnormal mitotic figures. (HE staining, x400)*

Discussion

Pilomatricoma, a common adnexal tumor that differentiates towards hair cortex cells, is usually benign and rarely invasive. A malignant variant of pilomatricoma was first described as malignant pilomatricoma or calcifying epitheliocarcinoma of Malherbe.[6] Most lesions occur on the scalp, face and neck in previously healthy skin,[11] as in the present case. Lesions generally occur as an asymptomatic mass and cannot be reliably distinguished from benign adenexal tumors by clinical appearance.[7] Metastases of malignant pilomatricoma have been documented, most of which recur locally prior to the occurrence of distant metastasis.[1]

Histologically, numerous areas, especially at the periphery of the tumor, show proliferation of large, anaplastic, hyperchromatic basophilic cells with numerous mitoses.[2] Toward the center of the tumor nests, abrupt transformation of basophilic cells into eosinophilic shadow cells of the type seen in benign pilomatricomas, or possibly a large cystic center containing necrotic debris,[2] is observed. The histological diagnosis can be challenging, especially that no clear histologic criteria distinguishing this neoplasm from other matrical tumors[11] have been established. Immunohistochemical investigations have not, so far, delineated a specific marker for this tumor.[8]

In this case, a wide excision including the surrounding skin was not carried out. Because of the rarity of malignant pilomatricoma, there are no well-defined standards in the surgical management of this neoplasm and no clear recommendations for the width of the surgical margins.[3] According to the data from the literature, wide excision of the lesion is associated with a low rate of tumor recurrence, whereas malignant pilomatricoma recurs in more than 50% of cases in which simple excision is carried out.[9,12] A minimum of 5 mm in the surgical treatment of malignant pilomatricoma has been empirically recommended as an adequate margin.[5] Moreover, because distant metastases have been described, close follow-up of the lesion is requisite. Particular attention is therefore needed when treating patients with this rare tumor.

Acknowledgement

We thank Yuko Nishikawa for technical assistance.

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