MALIGNANT PILOMATRIXOMA: CASE REPORT AND LITERATURE REVIEW

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INTRODUCTION

Educational Objective: Participants should be able to have a better understanding of the clinical course, pathology, and treatment of this rare disease.

Study Design: Report of a single case from a tertiary care institution and review of the literature.

METHODS: Report of a patient with malignant pilomatrixoma is presented with treatment course and histopathologic findings.

CASE HISTORY

A 60 year-old male presented with a history of a prior excision of a right temporal/pre-auricular pilomatrixoma. A 2x2 cm lesion recurred at the original site, and he was scheduled for elective re-resection 1 month later. When the patient presented to surgery, the lesion had tripled in size. Because of this rapid growth, the excision was deferred and repeat biopsy and further work-up with repeat imaging was performed. Biopsy was again consistent with pilomatrixoma.

The patient then underwent definitive surgery 2 ½ weeks later, including tympanomastoidectomy for facial nerve identification, parotidectomy approach through a modified Blair incision and wide-local resection of the temporal pilomatrixoma (Figure 1). Closure was completed with a full-thickness skin graft. Surgical final pathology revealed Malignant Pilomatrixoma (MP). The patient was scheduled for adjuvant radiation. On routine surveillance there has been no evidence of recurrence 4 ½ post-operatively.

BACKGROUND

- Pilomatrixoma: a slow growing dermo-hypodermic tumor arising from hair matrix cells. (younger age group, more than 60% before 3rd decade, F ≈ M 3:2).2,3
  - 1st described by Malherbe in 1880 as a calcifying epithelioma.4
  - Associated with Gardner’s Syndrome and Myotonic Dystrophy.4
  - Malignant Pilomatrixoma: less than 65 total cases reported. 1st case reported by Gromiko in 1927.4 More frequent in males > 3/1. Age range: 10-88, mean 45. Mean size at presentation: 4.6 cm.1,3
  - Sites of involvement reported: face, neck, back, upper extremities, scalp, breast, buttock, thigh.3
  - Predilection for sites is the same for the benign and malignant variants.2

DISCUSSION

- MP, a rare neoplasm, may arise from a transformation of a benign pilomatrixoma, or de novo.1
- The main indicators of malignancy appear to be the infiltration of skin and soft tissue, necrosis, ulceration, frequent mitoses, and pleomorphism, and MP frequently infiltrates more deeply and has shadow and basaloId cells with more nuclear abnormality than other disease processes.3
- Differentials include Basal Cell Carcinoma, which often has a sharp edge and may be separated from adjacent stroma by a gap, Proliferating Pilar Cyst, which lack shadow cells or basaloId cells, and Aggressive Pilomatrixoma.1
- Immunohistochemistry and flow cytometry have failed to differentiate the malignant form from its benign counterpart.2
- MP is characterized by frequent local recurrences. Lymph node and visceral metastasis has been reported, but is very rare. Therefore, MP is thought to be a low grade malignant tumor.1,2
- One study reported that of 17 patients with MP, 10 patients locally recurred within 5-18 months after surgery, and 3 of those had multiple recurrences.3 In another study of 55 patients, 21 recurred locally.2
- Locations of metastasis reported include the lungs, bone, and skin. Spread to base of skull/nasal cavity has been reported.2
- No recurrences have been reported after wide local excision, radiotherapy, or combination of these modalities. Chemotherapy has not been shown to have a response.2

PATHOLOGY

Pathological Findings
- Invasive features evident (Figure 2). Numerous mitotic figures with a high proliferative index (Figure 4).
- BasaloId cells are the main proliferating cells, which are usually ovoid or spindle-shaped with scant cytoplasm and prominent nucleoli. (Figures 3 & 5)

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REFERENCES