Pilomatrix Carcinoma of the Thoracic Spine: Case Report and Review of the Literature

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Abstract

Context: Pilomatrixoma is a common head and neck neoplasm in children. Its malignant counterpart, pilomatrix carcinoma, is rare and found more often in men.

Method: Case report of a 21-year-old man with pilomatrixoma of the thoracic spine that underwent malignant degeneration to pilomatrix carcinoma.

Findings: The appearance of a painless mobile axillary mass was followed by severe back pain 1 year later. Imaging revealed a compression fracture at the T5 level. The patient underwent resection of the axillary mass and spinal reconstruction of the fracture; the pathology was consistent with synchronous benign pilomatrixomas. Three months later he presented with a recurrence of the spinal lesion and underwent further surgical resection; the pathology was consistent with pilomatrix carcinoma. He received adjuvant radiotherapy and at his 1-year follow-up examination had no sign of recurrence.

Conclusion/Clinical Relevance: Pilomatrix carcinoma involving the spine is a rare occurrence. It has a high incidence of local recurrence, and wide excision may be necessary to reduce this risk. Radiotherapy may be a helpful adjuvant therapy. Clinicians should be aware of this entity because of its potential for distant metastasis.

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INTRODUCTION

Pilomatrixoma (pilomatricoma) is a benign neoplasm of hair follicle matrix cells. It was first described by Malherbe and Chenantais (1) in 1880 as a “calcifying epithelioma” derived from cells of the sebaceous glands. In 1961, Forbis and Helwig (2) published a review of the histochemistry and electron microscopy of 228 patients with the disease. They established that the tumor originated not from sebaceous glands but from the outer root sheath cell of the hair follicle and proposed the term “pilomatrixoma.”

In 1980, Lopansri and Mihm (3) reported a case of pilomatrixoma with malignant features and proposed the term “pilomatrix carcinoma” or “calcifying epitheliocarcinoma of Malherbe.” In 1999, Bremnes et al (4)

published a report of a case of pilomatrix carcinoma resistant to both chemotherapy and radiotherapy with multiple systemic metastases and reviewed 4 similar cases from the literature. Metastatic pilomatrix carcinoma remains rare, although more cases have been reported since its initial description in 1984 (4–12).

Involvement of the spine is exceedingly rare. To the authors' knowledge, only 3 such cases have been previously reported (Table 1). We present the case of a 21-year-old man with synchronous pilomatrixomas of the axilla and thoracic spine with malignant degeneration of the thoracic lesion to pilomatrix carcinoma.

CASE REPORT

A 21-year-old man initially presented to another institution with severe back pain and a 5-cm nontender mobile mass subcutaneously above the left scapula. The patient first noticed the mass approximately 1 year earlier. There was no discomfort associated with the mass, so he did not seek medical attention for it. The patient underwent computed tomography of the thoracic spine, which

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Table 1. Previously Published Reports of Pilomatrix Carcinoma With Spinal Involvement

Author	Age	Gender	Index Location	Spinal Involvement	Treatment	Outcome
Bremnes et al (4)	74	M	Left temporal cutaneous lesion	Thoracic bony metastases	Radiotherapy, chemotherapy	Distant metastases, death at 3 months
Campoy et al (19)	58	M	Midthoracic cutaneous lesion	Thoracic epidural extension	Radiotherapy, chemotherapy	Paraplegia, radiologic regression of tumor at 8 months follow up
Autelitano et al (2)	53	M	Posterior neck cutaneous lesion	T4 pathologic fracture	Surgical resection and reconstruction	Paraplegia, local relapse requiring reresection

revealed kyphotic angulation centered about a compression fracture of the T5 vertebral body (Figure 1A). A lesion involving the T5 body extending posteriorly into the canal with impingement on the spinal cord was demonstrated on magnetic resonance imaging (Figure 1B). The spine lesion was not contiguous on imaging with the subcutaneous scapular mass. All other testing and laboratory values were normal.

The patient underwent a biopsy of the subcutaneous lesion on the scapula. Pathology from that biopsy reportedly showed normal squamous cells. He also underwent computed tomography-guided needle biopsy of the T5 vertebral body lesion. The pathology was suggestive of squamous cell carcinoma. He was then transferred to our institution for further management of the compression fracture.

The neurologic examination performed on admission was normal. The patient reported significant, debilitating upper back pain that radiated around the chest. He was unable to ambulate secondary to the pain and was requiring high doses of intravenous narcotics. Given the

initial biopsy results, it was presumed that the 2 lesions represented metastatic squamous cell carcinoma. The estimated life expectancy with this interim diagnosis was poor, and the goal with initial surgery was therefore palliative spinal reconstruction with the hopes of alleviating the patient's pain and improving ambulation.

Initial Operation

The patient underwent a staged posterior-anterior decompression and fusion. A T5 laminectomy and pedicle screw fixation from T2 to T11 using Polaris instrumentation (Biomet, Wilrijk, Belgium) were performed in the prone position. A portion of tumor involving the posterior elements and pedicle was resected. The tumor was noted to be partially necrotic, tan-white, gritty, and well encapsulated. An iliac crest bone graft was harvested, morselized, and laid along decorticated bone surfaces to promote fusion. Connecting rods from T2 to T11, as well as cross-links at the T8–T9 and T3–T4 levels, were placed. Postoperative radiographs from the initial procedure are shown in Figure 2.

The patient was then repositioned from a Jackson table (Mizuho OSI, Union City, CA) in the prone position to a second operating table in the lateral decubitus position. A thoracotomy was performed with the assistance of a general surgeon. The patient's scapular lesion was circumferentially excised in the same setting, and the site was incorporated into the thoracotomy incision.

Upon exposure, the pathologic T5 vertebral body was identified, and the level was confirmed by intraoperative radiography. An en bloc resection of the T5 vertebral body and limited partial corpectomies of T4 and T6 were performed. Multiple specimens were sent for pathology. Endplates were prepared, and tricortical iliac crest allograft was used to reconstruct the T5 level. Morselized rib obtained during the anterior approach was used to supplement anterior fusion.

The patient had an unremarkable postoperative course and rapidly progressed to independent ambulation using a thoracolumbar orthosis. Final pathology from both the scapular lesion and the vertebral body was consistent with pilomatrixoma. No evidence of malignant degeneration was noted, with bland cytology and few mitoses described.

Figure 1. (A) Preoperative sagittal computed tomograph showing kyphotic angulation centered on a heterogeneously sclerotic compression fracture of the T5 vertebral body. (B) Sagittal T2-weighted magnetic resonance image demonstrating effacement of the thoracic thecal sac and compression of the spinal cord.

Figure 2. Lateral (A) and anterior-posterior (B) radiographs of the thoracic spine taken after the initial surgery showing segmental pedicle-screw instrumentation from T2 to T11 with connecting rods and cross-links.

Postoperative magnetic resonance imaging did not show residual enhancement, although the view was partly obscured by the hardware artifact. At 1-month follow up, the patient reported independent ambulation with a thoracolumbar orthosis and adequate pain control.

Three months after the initial surgery, the patient returned with a recurrence of debilitating back pain. He remained neurologically intact on exam. Computed tomography at this time revealed new bony destruction of the residual T4 vertebral body and rib, as well as an associated new soft-tissue density at the T5–T6 level (Figure 3). The findings were most consistent with recurrent tumor. Given the benign pathology from the initial surgery, the aggressive behavior of the tumor was concerning. It was recommended that the patient return to the operating room for another biopsy and, if possible, further resection. The possibility of postoperative adju-

vant treatment was also discussed. The patient and his family agreed to proceed.

Second Operation

The patient returned to the operating room for temporary removal of hardware and resection of the tumor. Based on preoperative imaging that showed the tumor was primarily lateral and posterior to the spinal cord, a single-stage, left, lateral extracavitary approach was chosen. The patient was positioned prone on a Jackson table. The connecting rods, cross-links, and left T4 pedicle screw were removed to allow better visualization of the tumor and spinal cord. A large amount of recurrent tumor was grossly evident on the left at the T4, T5, and T6 levels. A formal left T4 hemilaminectomy was performed, as well as left lateral costotransversectomies of the T4 and T5 vertebrae that appeared involved under gross inspection. All evident tumor was resected from the epidural space. The tricortical iliac crest graft placed at the time of initial surgery was left in place for anterior column support and fusion. Connecting rods and cross-links were reinserted and secured with cap nuts.

Postoperatively, the patient's pain was again much improved, and he progressed with physical rehabilitation to a level of independent function. Final pathology from the second procedure was consistent with pilomatix carcinoma. Increased cytologic atypia, cellularity, and proliferation of predominantly basophilic cells were noted. The pathology was confirmed by immunohistochemistry performed at an outside institution.

Immediate postoperative computed tomography with contrast revealed hyperdensities to the right of the thecal sac at the T3 to T5 levels, representing residual tumor. Four weeks after surgery, the patient underwent radiotherapy centered about the residual tumor with a suitable margin. A total of 3,750 centigray to the 95% isodose line in 250-centigray fractions over a 1-month period was administered.

Figure 3. Immediate postoperative (A) and 3-month follow-up (B) computed tomograph revealing progression of tumor posterior to the T4 and T5 vertebral bodies with increased vertebral destruction (white arrow).

Figure 4. Lateral and anterior-posterior radiographs of the thoracic spine at 1-year follow up, revealing good bony fusion.

Six weeks after radiation treatment, follow-up computed tomography and magnetic resonance imaging were performed and showed a marked regression of the tumor at T4 and T5 with marked improvement in the patient's symptoms and performance status. At 1-year follow up from initial presentation, the patient remained free of local recurrence and metastasis. Radiographs of the region revealed good bony fusion (Figure 4).

DISCUSSION

Pilomatrix carcinoma is a rare malignant variant of pilomatrixoma, an epithelial neoplasm of hair follicle matrix cells (13,14). Pilomatrixoma is one of the most common superficial head and neck masses found in children and young adults and has a female predominance (9,13). Pilomatrix carcinoma is found more commonly in men in the fourth and fifth decade. Its potential to be locally aggressive and resistant to radiotherapy has been well described (4,15).

Pilomatrixoma typically presents as a painless, firm mass found most commonly on the face, neck, back, and upper extremities (14). It is usually encapsulated by connective tissue and sharply demarcated from the surrounding normal tissue. Ulceration of the overlying epidermis is a common feature (14). Histologically, it is composed of eosinophilic ghost cells with basophilic matrical cells in the periphery. Keratinization, melano-

phages, and calcium deposits are often seen in the tumors as well (16).

Findings that distinguish pilomatrix carcinoma from its benign counterpart include large size, central necrosis, cytologic atypia, and high mitotic index (11,17,18). In this patient, proliferation of basophilic cells, increased cytologic atypia, increased cellularity, and increased mitotic activity were noted to establish the diagnosis of pilomatrix carcinoma (Figure 5).

Since its first description in 1984 by Gould et al (7), pilomatrix carcinoma with metastases has been infrequently reported in the world literature. Distant metastases to the brain, lungs, liver, heart, and pancreas have been reported (11). In their review of metastatic pilomatrix carcinoma, Bremnes et al (4) described a patient with metastases to the thoracic spine. The patient was a 74-year-old man with a left temporal epithelial lesion that also spread to the neck, both lungs, and abdomen within a few months of initial diagnosis (4). The patient succumbed to the disease, which was resistant to both chemotherapy and radiotherapy, 3 months after diagnosis. In addition to this case, 2 other reports of pilomatrix carcinoma with spinal involvement have been previously published (Table 1) (19,20).

It is most likely that the current patient presented with synchronous benign pilomatrixomas of the axillae

Figure 5. Photomicrographs of pathologic specimens of axillary and T5 vertebral body pilomatrix tumors. (A,B) Specimens of axillary mass. Basophilic cells (b) with abrupt keratinization to shadow cells (s). (Hematoxylin-eosin stain, original magnification A $\times 40$, B $\times 200$). (C) Specimen taken from resection of recurrent T5 tumor demonstrating a greater proportion of basophilic cells and a significantly increased mitotic rate (mitoses marked by arrows). (Hematoxylin-eosin stain, original magnification $\times 400$).

and thoracic spine with subsequent malignant degeneration of the spine lesion. This is corroborated by the absence of malignant features in pathology taken from the initial surgery and the observation that metastases are exceedingly rare with this tumor. The findings from the initial surgery also suggest that the biopsy results from the outside institution were incorrect. Postoperative imaging from the initial surgery did not reveal residual tumor, although the site was partially obscured by hardware artifact. Therefore, it is difficult to exclude the possibility that the patient's recurrence may have arisen from malignant degeneration of residual tumor. The possibility of sampling error with the original pathology and that the initial tumor itself was malignant given its rapid progression must also be considered, although this is less likely. The possibility that the thoracic lesion represented a metastasis from a primary lesion in the axilla must also be considered, but neither the pathologic findings nor the existing literature on this tumor, which suggests metastases are extremely rare, supports this theory. This patient underwent adjuvant radiotherapy and at 1-year follow up had no radiologic evidence of local recurrence or metastases. Further follow up will be necessary to gauge long-term results.

Approximately two thirds of patients who undergo simple excision will develop recurrences; local recurrence with malignant degeneration of pilomatrixoma has been previously reported (4,6,10,11,19). DeGalvez-Aranda et al (6) described the case of a woman who had undergone multiple resections of a cutaneous lesion on her knee over a 20-year period. On presentation to these investigators, the lesion demonstrated malignant features consistent with pilomatrix carcinoma. It eventually invaded the underlying bone and muscle and resulted in systemic metastases. Sassmannshausan and Chaffins (21) reported a 63-year-old woman who underwent excision of a scalp lesion consistent with benign pilomatrixoma. After 3 years, a rapidly progressive lesion consistent with pilomatrix carcinoma recurred at the same site. The locally aggressive nature of pilomatrixoma has led some investigators to suggest that wide excision with adequate margins is necessary, even if reoperation is required (16).

No chemotherapy has been shown effective in control of recurrent or metastatic disease (4,11,15). In the literature, several patients with pilomatrix carcinoma have been described who have undergone initial radiotherapy or surgery followed by adjuvant radiotherapy with no recurrence of disease (4). However, no combination of chemotherapy or radiotherapy has been effective in halting the progression of metastatic disease (15).

CONCLUSION

Pilomatrix carcinoma is a rare epithelial neoplasm of hair matrix cells. It is much less common and seen in a different population than its benign counterpart, pilomatrixoma. It has a high incidence of local recurrence, and

wide excision may be necessary to reduce this risk. Clinicians should be aware of this entity because of its potential for distant metastases. Although there is some evidence that radiotherapy may be a helpful adjuvant therapy, there is no evidence that any regimen of chemotherapy provides benefit. Further studies of this rare disease may elucidate its natural history and optimal treatment regimens.

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