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Malignant Transformation of High-Grade Osteoblastoma of the Petrous Apex with Subcutaneous Metastasis

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Abstract

Objectives—To present the clinical presentation, management, and pathological findings of a patient with osteosarcoma of the petrous apex with atypical metastasis to the lower abdominal wall.

Methods—Retrospective review of the records of a case of osteosarcoma of the petrous apex.

Results—A 49-year old man with facial pain is diagnosed with a right petrous apex lesion with biopsy demonstrating high-grade osteoblastoma. Multiple attempts at en bloc resection were not curative, therefore radiation and chemotherapy was recommended after two surgical attempts. The patient subsequently developed a cutaneous, lower abdominal wall mass which demonstrated osteosarcoma. Petrous apex tumor growth progressed despite treatment until the patient expired from burden of disease.

Conclusions—Temporal bone osteoblastoma and osteosarcoma are both extremely rare, and can be difficult to differentiate histologically. The present case illustrates these difficulties and demonstrates the possibility of malignant conversion from high-grade osteobastoma to osteosarcoma.

Keywords

osteoblastoma; osteosarcoma; cutaneous metastasis; petrous apex

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Conflict of Interest: None

INTRODUCTION

Primary neoplasms arising within the petrous apex are a rare clinical entity. The differential diagnosis for petrous apex tumors include chondrosarcoma, schwanomma, paraganglioma, and primary bony tumors, such as osteoblastoma and osteosarcoma. We report a case of a 49-year old man with a petrous apex osteoblastoma with presumptive malignant degeneration into osteosarcoma and subsequent distant metastasis. The particular rarity of this clinical scenario as well as the noted difficulty of histologically differentiating osteoblastoma, osteosarcoma, and malignant degeneration between the two is what prompted this case report.

CASE REPORT

The patient was a 49-year old male who presented with facial pain which was progressive over the course of 2.5 months. Prior to referral, he had been treated unsuccessfully for presumed chronic rhinosinusitis with oral antibiotics and non-steroidal anti-inflammatory drugsHis pain was characterized as predominantly over the right maxilla as well as radiating into the temple and forehead. Notably, the patient also had a history of sudden moderate right-sided sensorineural hearing loss 1.5 years prior to presentation. An MRI at that time was unremarkable. He noted a subjective progression of the right-sided hearing loss over the same time-frame as the facial pain. Previous medical history was notable only for ulcerative colitis. On examination, a right-sided middle ear mass was noted, which abutted the undersurface of the anterior 25% of the tympanic membrane. Facial nerve function was normal. Audiometric evaluation demonstrated a profound right-sided sensorineural hearing loss and subsequent MRI and CT demonstrated a destructive right petrous apex lesion with extension into the carotid canal and the medial epitympanum, up to Meckel’s cave. There was associated bony erosion of the otic capsule. The lesion was isointense to brain on T1-weighted images, hypointense on T2-weighted images, and demonstrated enhancement post-Gadolinium infusion (Figure 1).

A transcanal middle ear exploration and biopsy was performed. The mass was noted to be composed of both fibrous soft tissue and calcific spiculations with the appearance of moth eaten wood. Histologic examination showed a proliferative bony lesion. Expert pathologic consultation was obtained from multiple head and neck pathologists, including consultation with the Armed Forces Institute of Pathology (AFIP), and consensus opinion was that the lesion represented a high-grade osteoblastoma. The patient underwent gross total resection of the tumor via a transcochlear approach. The neoplasm was found to fill the entire petrous apexand involve the intrapetrous portion of the carotid canal, but was easily dissected from the internal carotid artery. All gross tumor was removed. There were no postoperative complications and the patient was discharged home on post-operative day three. Pathology was again confirmed by multiple head & neck pathologists to be high-grade osteoblastoma (Figure 2).

One month post-operatively, the patient awoke with sudden-onset right-sided facial weakness, which completely responded to oral corticosteroid therapy. A temporal bone CT showed only expected post-operative changes. howeverone month later, he developed

progressive right-sided facial numbness. MRI imaging showed a mildly contrast-enhancing soft tissue filling the petrous apex which was felt to represent an inflammatory response or indolent petrous apicitis versus residual tumor. A follow-up MRI performed 1 month later showed no change.

Four months post-operatively, the patient again developed a right-sided facial paresis that rapidly progressed to a total paralysis. Repeat CT and MR imaging showed dramatic interval mineralization of the previously noted soft tissue within the petrous apex with new bony destruction at the petroclival junction.

The patient proceeded to the OR for re-resection via the transcochlear approach. Based on intraoperative nerve stimulation, the facial nerve appeared to have a conduction block at the geniculate ganglion and facial nerve decompression was performed. The tumor was not amenable to en bloc resection due to internal carotid artery involvement and anterior inferior cerebellar artery involvement. Frozen section analysis returned positive for osteoblastoma and the decision was made to leave gross disease behind given the significant risk of morbidity with further dissection. There were no intraoperative or postoperative complications and the patient was discharged home on post-operative day two. Final surgical pathology was again read by multiple head & neck pathologists as high-grade osteoblastoma.

Due to the continued growth of the tumor, primary chemotherapy with doxorubicin combined with alternating ifosfamide and cisplatinum was recommended. The patient completed 4 months of treatment. Right facial nerve function remained a House-Brackmann grade VI and post-treatment CT and MRI showed new bony destruction near the basocciput region, as well as destruction of the narrow space and the cortex adjacent to the foramen magnum with associated new soft tissue enhancement. Surgery was not recommended because of the inability to completely remove the tumor without sacrificing the internal carotid artery and the lower cranial nerves. Proton irradiation was recommended and completed instead.

During radiation therapy, the patient developed a soft tissue nodule in the lower left abdominal quadrant at the fat graft harvest site of the initial resection. The abdominal fat graft site was treated as a separate surgical site during the initial operation and there was no sharing of instruments. Fine needle aspiration of the nodule was negative for carcinoma, however, there was continued growth of the mass and it was therefore subsequently excised for pathological examination. The histology was discussed and examined with several pathologists who all agreed the lesion definitively represented osteosarcoma, rather than osteoblastoma (Figure 3).

The patient underwent further resection to achieve negative margins of his abdominal wall mass. It was recommended that he undergo additional chemotherapy with high-dose methotrexate, which was tolerated poorly by the patient and discontinued early.

Post-treatment MRI showed progression of the lesion into the posterior fossa with compression of the brainstem and cerebellum. The patient was enrolled in a trial of tetrathiomolydate and had initial symptom improvement, however serial MR imaging

showed continued growth of the primary tumor. The patient became increasingly ataxic and lethargic and passed away due to burden of disease two years after initial diagnosis (Figure 4).

DISCUSSION

Osteoblastoma is a rare, benign bone tumor that was first categorized as distinct from osteoid osteoma in 1956, simultaneously by both Jaffe and Lichtenstein.[1] Although histologically similar, osteoblastoma and osteoid osteoma are distinguished by the size of the lesion.[1,2] Osteoblastomas most commonly form in the vertebral column, long and flat bones, and metacarpals and metatarsals.[2,3,4] Osteoblastoma of the temporal bone is exceptionally rare, with only 15 reported cases in the literature.[5] Complete excision is considered curative for these lesions, however subtotal resection or curettage is occasionally indicated in limb preservation surgery or if the primary site is not amenable to en bloc resection. A clinical phenomenon where the primary tumor involutes and ceases to grow has been observed after sub-total resection, which is believed to explain the low clinical recurrence rate of 10–20%. The role of adjuvant radiation therapy or chemotherapy has not been well studied. Although very rare, malignant degeneration of osteoblastoma following radiation therapy has been described.[6]

Osteosarcoma, while the most common malignant primary bone tumor, is also a rare clinical entity.[7] Diagnoses are most common among adolescents and primarily found in long bones, a demographic which mirrors osteoblastoma.[7] Primary osteosarcoma of the temporal bone is also an extremely rare clinical entity, with less than 10 cases described in the literature. Cutaneous metastases of osteosarcoma is also very rare, with one study identifying only 6 reported cases in the literature as of 2006.[8] For osteosarcoma of the extremities, treatment is generally wide local resection and adjuvant chemotherapy and radiation, which tends to be fairly successful.[9] In cases such as the one presented, complete resection is difficult because of the proximity of vital structures.

Histopathologic differentiation of osteoblastoma and osteosarcoma can be very difficult. Borderline tumors with radiographic and histologic features resembling both osteoblastoma and osteosarcoma occur for which Dorfman and Weiss and Lucas et al have adopted the term “aggressive osteoblastoma” or “high-grade osteoblastoma.” These lesions are characterized by histologic findings of epithelioid osteoblasts, increased mitotic activity within the stromal component, and disordered osteioid matrix. These lesions are felt to carry a higher rate of recurrence but no metastatic potential. To further complicate matters, there have been reports of malignant change in aggressive osteoblastoma, though the rate of transformation has not been studied due to the clinical rarity of these tumors.[10]

CONCLUSION

This case illustrates the diagnostic dilemma presented by such borderline tumors. It is possible that the initial tumor represented a mis-diagnosed osteosarcoma at the initial and second resection. However, multidisciplinary discussion favors that the initial tumor was a high-grade osteoblastoma which underwent malignant degeneration. This malignant

degeneration could have been radiation-induced or spontaneous, as both have been described. The subsequent subcutaneous metastasis has not been previously described in the literature. Pathologic review by a head & neck pathologist of bony tumors of the petrous apex may help mitigate these diagnostic challenges and help to tailor therapy appropriately early in care. Primary bone neoplasms, both benign and malignant, should remain in the differential of petrous apex lesions.

Acknowledgments

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SUMMARY

- Differentiating osteosarcoma vs. high-grade osteoblastoma histologically can be a challenging diagnosis
- Review by head and neck pathologists can help mitigate this challenge.
- Neoplasm should remain in the differential for patients with petrous apex lesions.

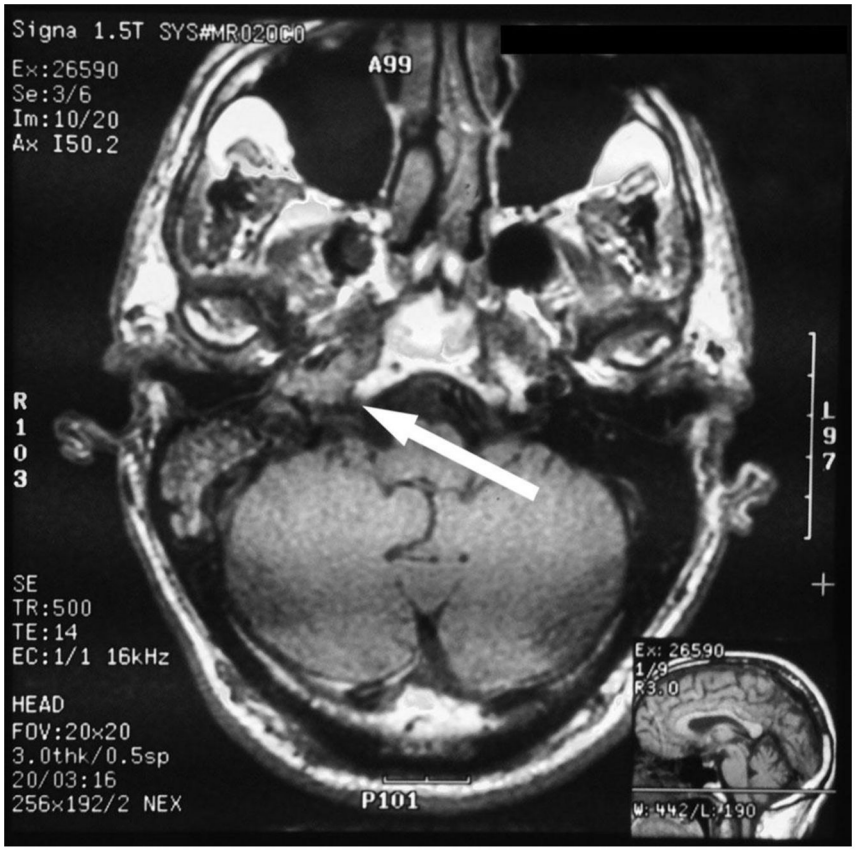

Figure 1.

Initial MRI (T1-weighted, post gadolinium) at time of diagnosis of right petrous apex lesion, showing invasion into the carotid canal, Meckel's cave, and the medial epitympanum.

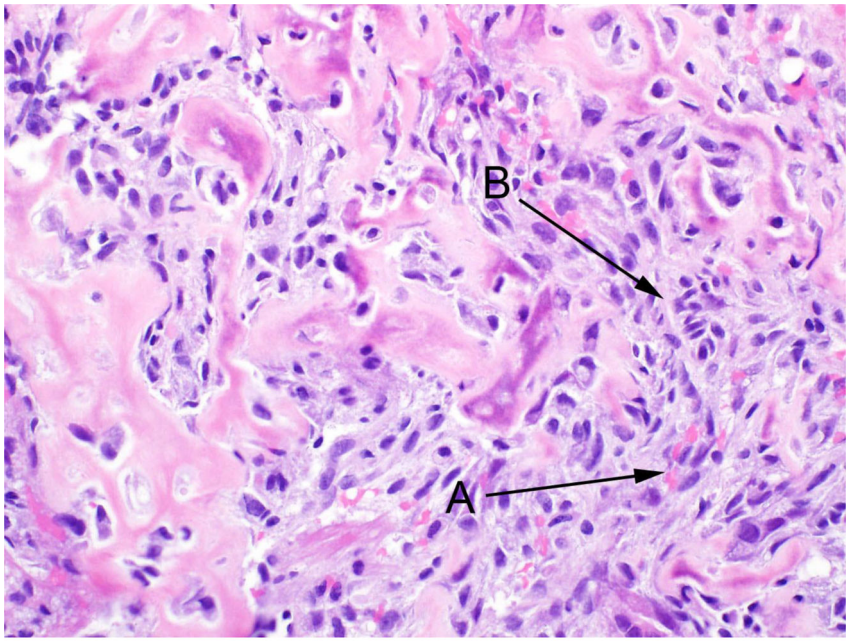

Figure 2.

High-power view of the primary excisional specimen from the petrous apex. Note the highly-vascularized stoma (A) as well increased cellularity of the stroma (B) and disorganization of the bony matrix which favors a diagnosis of aggressive osteoblastoma.

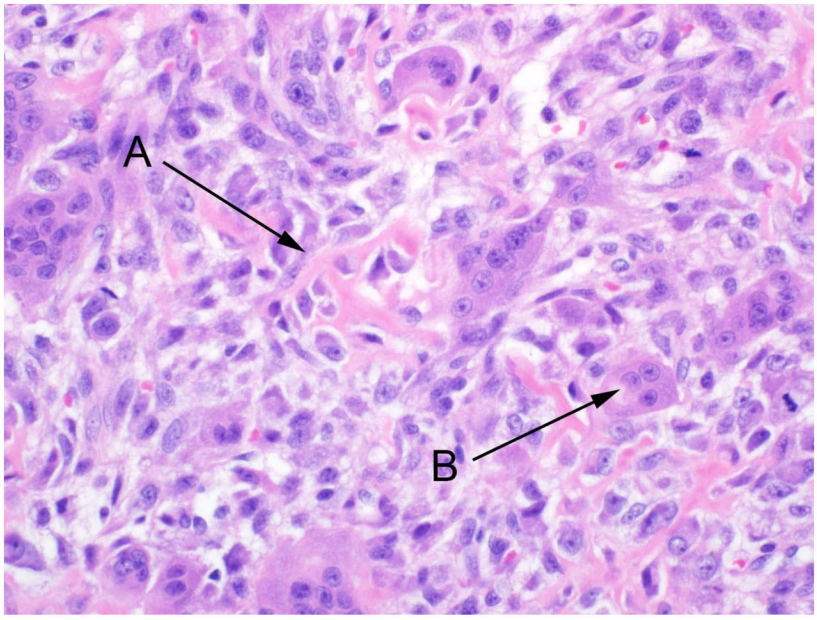

Figure 3.

High-power view of the excisional specimen from the patient's left lower cutaneous abdominal quadrant. Note the bizarre nuclei with vacuolated cystoplasm of the osteoblasts (A) and coarse, lacelike pattern of the neoplastic bone (B). Final surgical pathology read as osteosarcoma.

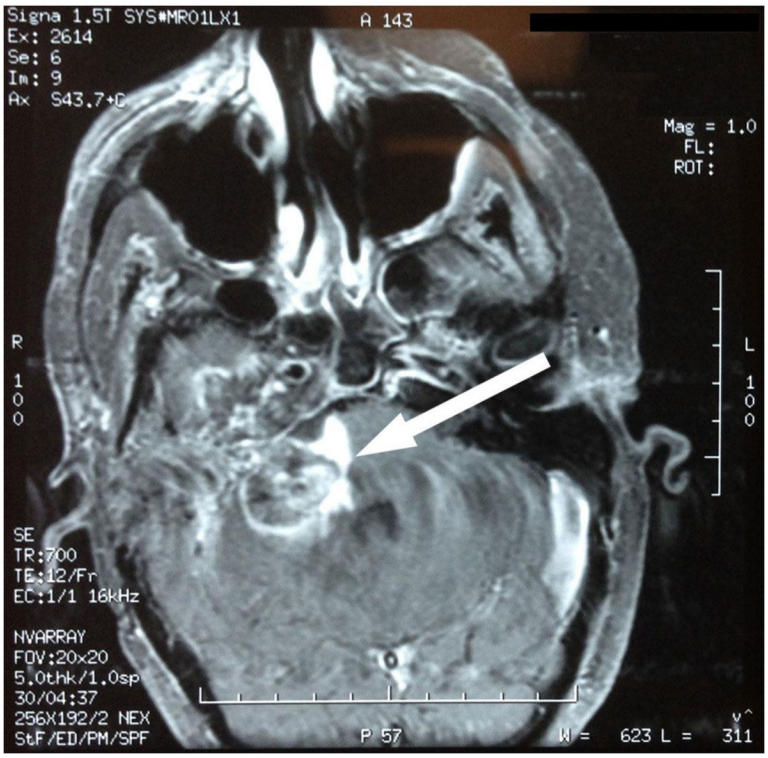

Figure 4.
Final MRI (T1-weighted, post gadolinium) of lesion prior to patient demise.