CASE REPORT

Metastatic Alveolar Soft-Part Sarcoma of the Intracranial Skull Base: Case Report

David B. Cohen, M.D.,[1] David M. Jones, M.D.,[2] Allan H. Fergus, M.D.,[3] Jiang Qian, M.D.,[2] and Marc S. Schwartz, M.D.[3]

ABSTRACT

A patient with an intracranial skull base alveolar soft-part sarcoma, thought on preoperative imaging to be a meningioma, is presented. The mass was removed and identified by pathological evaluation to be an alveolar soft-part sarcoma. Postoperative investigation revealed widespread systemic disease. The histologic and clinical characteristics of this unusual tumor and the implications for prognosis and treatment are discussed.

KEYWORDS: Brain metastases, sarcoma, skull base tumor

Accounting for fewer than 1% of all sarcomas, alveolar soft-part sarcomas (ASPSs) are an extremely rare type of tumor.[1] These tumors most often arise in the extremities, head and neck region, or trunk and rarely develop as an intracranial mass.[2,3] On initial presentation, an ASPS may be difficult to distinguish from other malignant or benign tumors. We present a patient with a metastatic ASPS that mimicked a skull base meningioma.[4] Because the primary tumor and other metastases were asymptomatic, the lesions were not detected until after a craniotomy had been performed.

CASE REPORT

A 28-year-old right-handed man had a 6-month history of progressively worsening, intermittent, severe headaches and a 2-week history of declining vision in the right eye. His medical and surgical history were unremarkable.

A physical examination revealed decreased visual acuity in the right eye. The right optic disc showed mild atrophy. His neurologic examination otherwise was normal, with no gross motor or sensory defects. Gadolinium-enhanced T1-weighted

Skull Base, volume 12, number 1, 2002. Address for correspondence and reprint requests: Marc S. Schwartz, M.D., Division of Neurosurgery, MC-61, Albany Medical College, 47 New Scotland Avenue, Albany, NY 12208. E-mail: schwarm@mail.amc.edu. [1]Department of Neurosurgery, Allegheny University, Pittsburgh, Pennsylvania. [2]Department of Pathology, and [3]Division of Neurosurgery, Albany Medical College, Albany, New York. Tel: +1(212) 584-4662. 1531-5010,p;2002,12,01,033,038,ftx, en;sbs00250x.

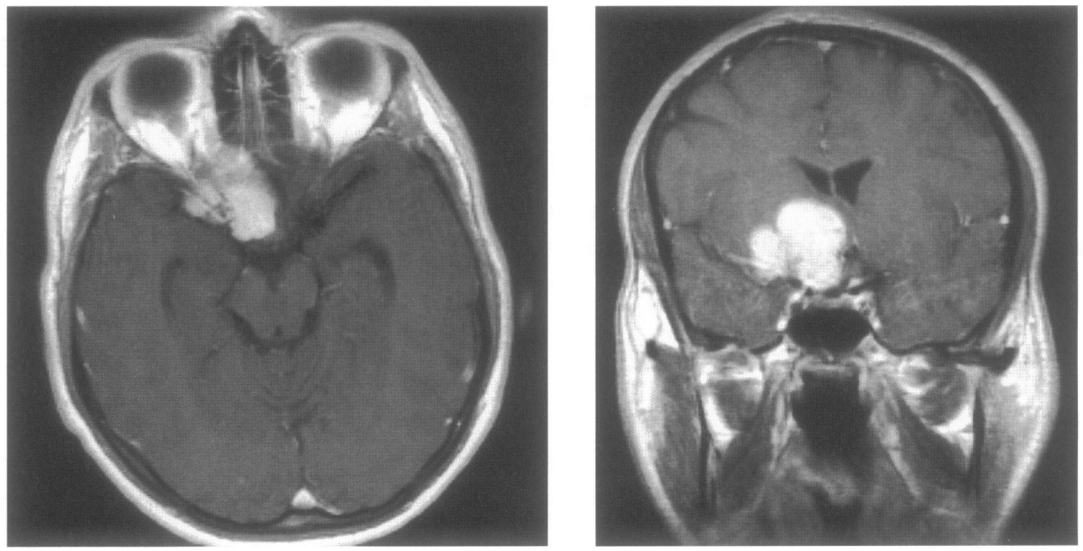

Figure 1 Initial magnetic resonance appearance of the intracranial skull base alveolar soft-part sarcoma. Note the lack of hyperostosis and the relatively small dural tail best seen on the axial image (A). The full size of the tumor is best appreciated on the coronal image (B).

magnetic resonance imaging (MRI) revealed a well-circumscribed, 3.5-cm, homogeneously enhancing, dural-based mass in the right suprasellar region compressing the optic chiasm, optic nerve, and optic tract (Fig. 1). The patient was seen by an ophthalmologist, and his visual acuity was 20/200 OD with a field cut in the inferior nasal field. His heart and lung were normal. Because the patient was young, no chest radiograph was obtained.

A right frontotemporal craniotomy with orbital osteotomy, extradural optic nerve decompression, and anterior clinoidectomy was performed. The tumor did not have the typical appearance of a meningioma: It was smooth, friable, and extremely vascular. Frozen-section examination identified the lesion as a probable meningioma. The tumor was attached to dura over the region of the clinoid and tuberculum; however, this attachment was relatively loose, and the underlying dura appeared normal. The tumor was separated completely from the underlying dura, but its vascularity did not decrease. It appeared to be receiving blood from small arteries along the entire interface between tumor and brain. A gross total resection of the tumor was achieved despite profuse bleeding that continued until the last pieces were removed.

After surgery, the patient remained neurologically stable. MRI revealed no residual tumor. After discharge from the hospital, the patient's headaches improved. His vision improved to 20/30 OD. Despite the frozen-section diagnosis of the lesion, later pathologic analysis diagnosed the lesion to be an ASPS (Fig. 2). At a follow-up examination 2 weeks after surgery, the pathologic diagnosis and its implications were described to the patient and he developed chest pain. A chest radiograph showed multiple pulmonary nodules. A full metastatic evaluation followed and revealed a large soft-tissue mass in the anterior left shoulder.

Because the disease was widespread and chemoresistant, the patient was followed with quarterly serial scans only. The pulmonary nodules remained unchanged. Nine months later, however, he again developed headaches. MRI showed multiple small, enhancing cerebral masses but no evidence of tumor regrowth at the original intracranial site (Fig. 3). He was treated with fractionated whole-brain radiation and enrolled in an investigative phase 1 liposomal chemotherapy regimen (NX-211). He remains neurologically stable with no clinical or radiographic evidence of disease progression 12 months after diagnosis of the multiple

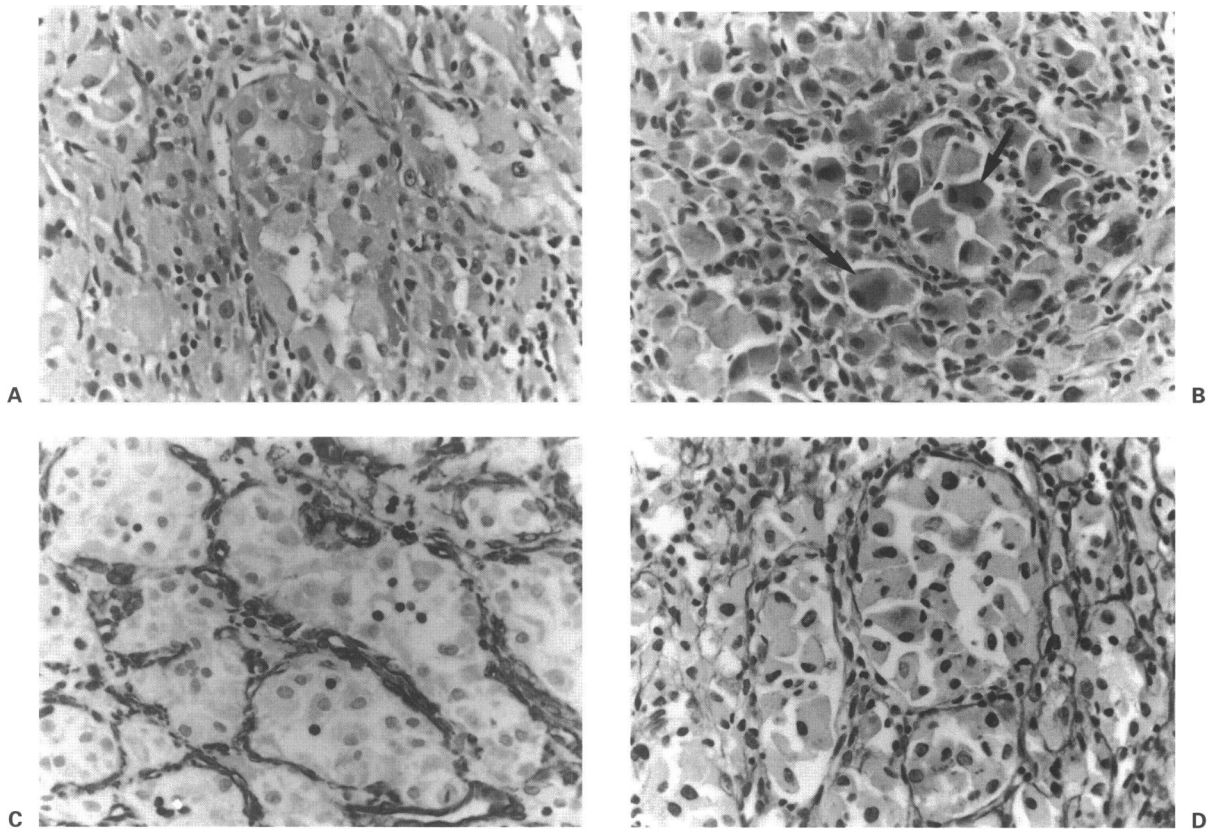

Figure 2 Photomicrographs of the dural-based intracranial metastasis of the alveolar soft-part sarcoma. (A) Large, round, and polygonal tumor cells are arranged in an alveolar pattern (hematoxylin and eosin). (B) Periodic acid Schiff (PAS)-positive and diastase-resistant intracytoplasmic crystalline granules are indicated by arrows (PAS-D stain). (C) Vimentin and CD34-positive fibrovascular septa surround the tumor nests (immunohistochemical stain for vimentin). (D) The alveolar/nested pattern of the tumor is highlighted by reticulin-positive fibers (reticulum stain). Original magnification, x 400 for all figures.

intracranial metastases and 21 months after his initial presentation. There continues to be no evidence of recurrent disease at the site of the original intracranial tumor, but the size of the multiple cerebral masses remains unchanged.

DISCUSSION

First described by Christopherson and Stewart in 1952,[5] ASPSs are exceedingly rare tumors that account for fewer than 1% of all sarcomas. They most commonly arise in the second and third decades of life and are more common in females when diagnosed earlier in life.[1,6,7] ASPSs most often occur in the extremities, followed by the head and neck region and trunk.[2] ASPSs also have been involved with the mediastinum,[8] pituitary gland,[9] stomach,[10] spinal column,[11] pulmonary vasculature,[12] lung,[13–15] breast,[16] and female genital tract.[17] Tumors in the head and neck are most prevalent in children.[18]

The original report assigned this tumor its descriptive name because of its uncertain histogenesis.[5] The term *alveolar* refers not to a pulmonary origin but to the appearance of irregular spaces within groups of cells when viewed through light microscopy.[2] The most distinctive histologic features of these tumors are periodic acid Schiff (PAS)-positive, diastase-resistant, rhomboid and

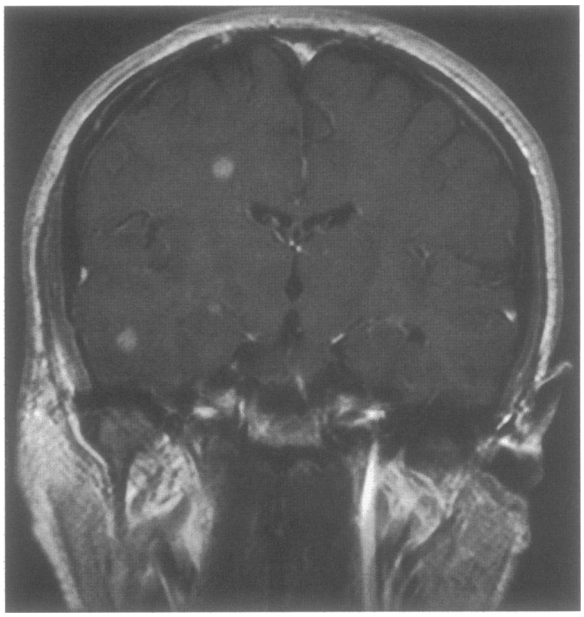

Figure 3 Follow-up magnetic resonance imaging showing multiple small, enhancing intraparenchymal metastases. No residual or recurrent tumor is seen at the original site of intracranial involvement.

rod-shaped crystals in a sheaflike arrangement in the cytoplasm.

The histogenesis of ASPS has been a source of considerable debate. Two major theories propose either a neural or muscular origin for ASPS. Although early evidence supported a neural origin,[19,20] more recent evidence points toward an origin from muscular tissue.[21] Nakano[22] found evidence of a skeletal-muscle origin, finding actin in the granules of ASPS cells. Furthermore, several antibodies to skeletal muscle reacted with ASPS cells. Wang et al.[7] and Christopherson and Stewart[5] have suggested that ASPS may originate from the muscle spindle, which contains both intrafusal muscle fibers and nerve tissue.

Usually, ASPSs are painless, slow-growing masses. If symptoms are present, they tend to be related to the direct mass effect of the tumor.[2] Unlike most sarcomas, these tumors frequently metastasize, and metastasis to the lung or brain may be the initial indication of disease. The primary tumor is seldom diagnosed before metastases occur.[23] The presence of metastases at diagnosis makes a marked difference in prognosis. The median survival for patients with no metastases is 11 years compared with 3 years for those with metastases.[6] These figures suggest that metastases during the course of the disease are the rule but usually take many years to develop and follow an indolent course of progression.[2]

If no metastases are present, resection of the primary tumor is the best treatment. In a series by Lieberman and co-workers,[6] the median survival of patients who underwent excision was 218 months compared with 63.5 months for those who did not undergo excision. When cancer metastasizes to the brain, removal of the metastases tends to increase survival.[24] A study of patients with brain metastases from various types of sarcomas found that removing brain metastases from ASPS was associated with a relatively good prognosis. These researchers suggest that this outcome may be related to the slow-growing, indolent nature of ASPS.[25] Resection of pulmonary metastases also can increase survival.[26] No significant benefit for survival has been demonstrated with adjuvant chemotherapy, but adjuvant radiation therapy may be of some benefit.[1,27]

Our patient presented with a single mass lesion attached to the dura and with perhaps a suggestion of a dural tail. Radiographic analysis was most consistent with the diagnosis of meningioma, and we approached surgical treatment with the expectation of encountering this type of tumor. In retrospect, several features of the case, including the relatively quick onset of symptoms and radiographic studies that showed no hyperostosis and a relatively small area of dural attachment, may have suggested the more worrisome diagnosis. Including sarcoma in the differential diagnosis probably would not have altered the decision to operate or the perioperative course, but it may have been useful in terms of the patient's expectations before and immediately after surgery.

Several examples show how consideration of ASPS or other sarcomas in the differential diagnosis can affect treatment. Preoperatively, awareness of the presence of other systemic disease could affect the decision to operate. Given our patient's symptoms and the location of the disease, resection was needed despite the presence of systemic dis-

ease, but this may not be the case for all patients. Intraoperatively, the specifics of surgical strategy also could change. We had expected to devascularize the tumor by separating it from the dura, but resection was made more difficult by the continued profuse bleeding from the tumor. If malignant disease is possible, further postoperative evaluation may be necessary. Our patient underwent an immediate postoperative MRI only because his intraoperative findings were unusual. Although our patient did not undergo early adjuvant treatment, this option could be an important consideration.

Unlike even malignant meningiomas or hemangiopericytomas, which typically are included in the differential diagnosis of meningioma, the diagnosis of ASPS is associated with the possibility of metastatic disease. Although rare, ASPSs and other types of sarcomas can occur as dural-based lesions that resemble meningiomas. Especially in younger patients or in those with unusual clinical radiographic features, ASPS should be considered in the differential diagnosis. In some cases, considering ASPSs and other forms of sarcomas as a potential diagnosis could affect not only the perioperative course of treatment but also the decision to operate.

CONCLUSIONS

Alveolar soft-part sarcoma should be considered in the differential diagnosis of an intracranial mass with the radiographic characteristics of a meningioma, especially if clinical or radiographic findings are even marginally unusual. Inclusion of metastatic sarcoma in the differential diagnosis of meningioma could, in some cases, alter the approach to treatment.

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