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Hemangioendothelioma of the Sphenoid Bone : A Case Report

Hemangioendothelioma is borderline or intermediate type of vascular neoplasm. Hemangioendothelioma is rare lesion that constitutes less than 0.5% of the malignant tumors of bone. We present a case of low-grade hemagioendothelioma of the skull in a 29-yr-old woman. She had pain, diplopia and exophthalmos of the left eye. Radiographic images showed a relatively well-demarcated, expansile osteolytic lesion with irregularly thickened trabeculae and calcifications in the left greater wing of sphenoid bone. Histologically, the tumor was an infiltrative vasoformative lesion. The vessels are generally well-formed with open or compressed lumina surrounded by endothelial cells showing mild atypia. It lacked frequent mitotic figures and severe atypia. Although excessive bleeding occurred during the operation, the mass was totally resected. Postoperative radiation was not necessary. She is free of disease and well 6 months postoperatively.

Key Words: *Hemangioendothelioma; Vascular Neoplasms; Skull*

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INTRODUCTION

Hemangioendotheliomas are intermediate or borderline malignant tumors of vascular origin and well known as a superficial or deep soft tissue tumor of the extremities and retroperitoneum (1). Hemangioendothelioma of the bone is a rare primary neoplasm occurring in the bone, and in many series, its incidences have been reported to be less than 1% of primary bone tumors (2-5). The neoplasm is found most frequently in the long bones of the extremities (2, 3, 5, 6). Only rare cases arising in the skull have been published (7-11). We present a case of hemangioendothelioma of the sphenoid bone in a 29-yr-old woman with ophthalmologic manifestations.

CASE REPORT

A 29-yr-old pregnant woman complained of pain, diplopia and exophthalmos of the left eye that had slowly progressed during several months. There was no history of trauma or concurrent illness. Ophthalmic examination showed limitation of lateral gaze and decreased visual acuity.

Simple antero-posterior view of the skull showed obliteration of the left oblique orbital line and unsharpness of left sphenoid ridge, suggesting destructive lesion at the left greater wing of sphenoid bone. CT showed an expansile osteolytic lesion with irregularly thickened trabeculae and calcifications in the left greater wing of sphenoid bone, displacing left eyeball anteriorly. The cortical margin was partially thickened and also partially destroyed

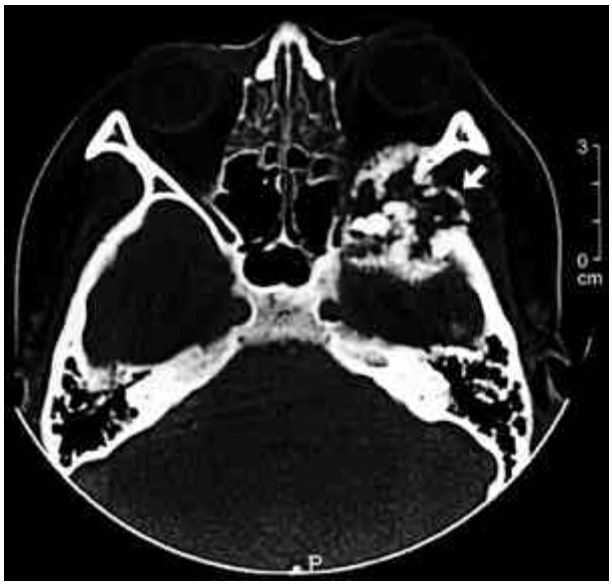

Fig. 1. On bone setting of orbital CT, expansile osteolytic lesion (arrow) is seen on the left greater wing of sphenoid bone, displacing left eyeball anteriorly. Multiple irregularly thickened trabeculae are also noted inside the lesion, as well as focal cortical defect.

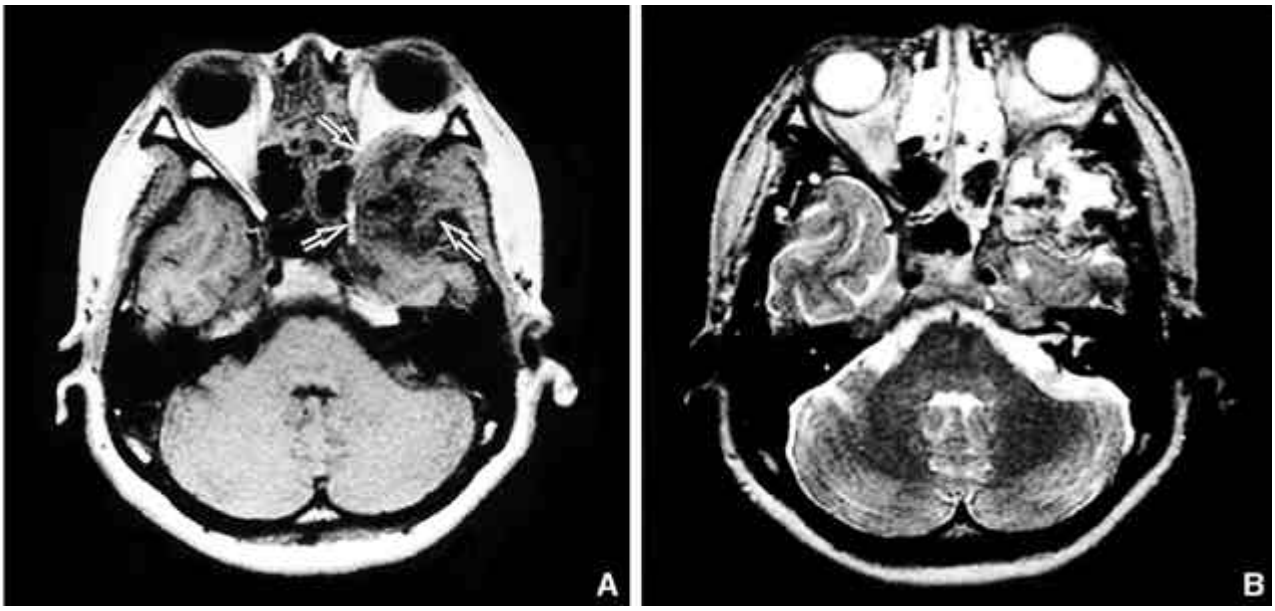

Fig. 2. Axial T1-weighted MR image (A) shows a heterogeneously hypointense lesion on the left greater wing of sphenoid bone (arrow), which is hyperintense with multiple signal voids on T2-weighted image (B).

(Fig. 1). The lesion was heterogeneously hypointense on T1-weighted MR image, hyperintense with multiple signal voids on T2-weighted MR image and well enhanced on contrast-enhanced T1-weighted image (Fig. 2). Differential diagnoses included low-grade chondrosarcoma, aneurysmal bone cyst and primary vascular tumors of bone.

Excisional biopsy was performed. The tumor mass was confined to the sphenoid bone, and the thin cortical bone was easily fractured. Incision of the capsule to obtain biopsies resulted in brisk bleeding and the tumor tissue was a hemorrhagic, spongy and lobular mass, packed between sclerotic bony trabeculae. She refused blood transfusion and complete resection was impossible due to massive bleeding. Biopsy disclosed the pathologic diagnosis of hemangioendothelioma. Subsequently, she was reoperated on 10 days after the first operation and the tumor was completely excised. Further imaging studies including bone scan were performed to search for another lesion, but not found.

Histologically, the tumor was a definite vasoformative lesion showing infiltrative growth. The vessels were generally well-formed with open lumina. The endothelial cells lining the vessels showed mild hyperchromasia, slightly variation in size and frequent cytoplasmic vacuolation. These cells were larger and plumper than those of hemangioma, and intravascular papillary tufting was focally noted. Neither cellular anaplasia nor mitotic figure was found. (Fig. 3) The tumor was diffusely infiltrating into the marrow spaces and exuberant reactive new bone formation was noted throughout the lesion (Fig. 4). Immunohistochemical staining revealed positive reactivity for vimentin and Factor VIII-related antigen. There was no immunoreactivity for cytokeratin, smooth muscle actin, or S-100 protein. By electron microscopy, the cells showed characteristics of endothelium, including continuous and multilayered basal lamina, tight junctions, pinocytotic vesicles and villus-like projections on their

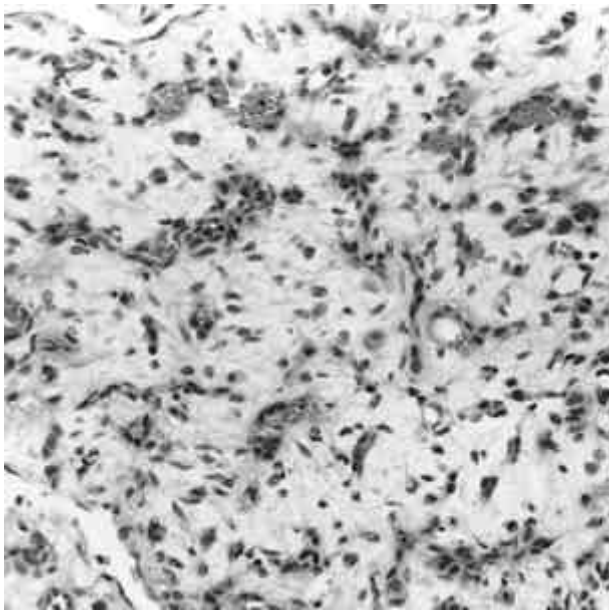

Fig. 3. The tumor consists of abundant well-formed vascular spaces lined by crowded endothelial cells showing mild cytologic atypia. The neoplastic endothelial cells show mild hyperchromasia, slight variation in size and frequent cytoplasmic vacuolation, and there is no mitotic activity (H&E, ×200).

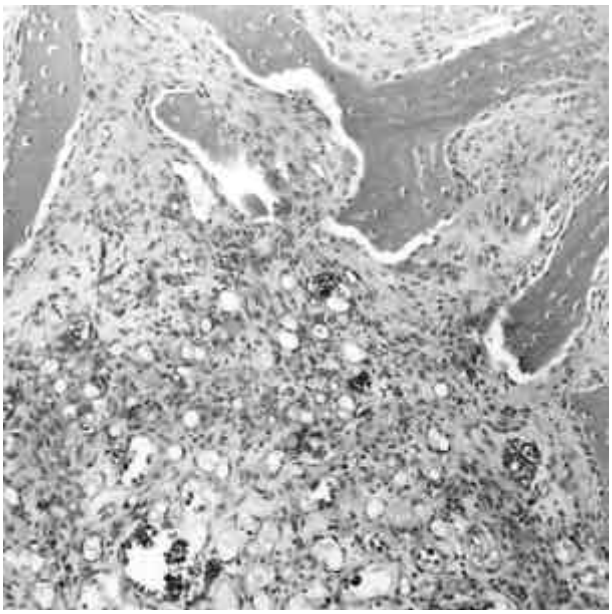

Fig. 4. Reactive new bone formation is seen throughout the lesion (H&E, ×100).

luminal aspect. They also contained moderate amounts of microfilaments, some rough endoplasmic reticulum and rare Weibel-palade bodies.

DISCUSSION

The term hemangioendothelioma is used to denote vascular neoplasms showing histologic features and clinical behavior intermediate between hemangioma and angiosarcoma (1). They frequently occur in soft tissue, skin, lung and liver, and uncommonly involve the bone (1-5). Because of it rarity, the literature is confusing regarding the nomenclature of these tumors. Term such as angiosarcoma, angioendothelioma, hemangiosarcoma, hemangioendothelial sarcoma, and angioblastic sarcoma have been used to designate this neoplasm (5, 13). Recently, authors tend to call all of these tumors hemangioendothelioma (5, 7-14).

Hemangioendothelioma may affect any portion of the skeleton but most often in the axial skeleton (2, 3, 5, 6, 12). Involvement of the cranium is rare. We have found less than twenty reported cases involving the skull in English literature (7-12). It may occur at any age, but it occurs most common between the second and third decades of life (3-5, 13). There is a slight male predominance (3, 4). The usual presenting symptom of lesions not involving the skull is pain, whereas in the skull it is a local mass or swelling (8). Multicentric tumors were present in approximately 25% of cases (5, 13). This case is solitary lesion occurring in the skull of a young, pregnant woman complained of pain, diplopia and exophthalmos, reflecting localized mass effect.

Radiologically, the vast majority of hemangioendothelioma of bone are purely osteolytic, however, the radiologic findings can be nonspecific (15, 16). When the lesions are multicentric, they commonly have a distinctive radiologic appearance, purely osteolytic lesions involving the cortex of the tubular bones (17). In contrast, if the lesion is solitary and unusually located, as in this case, there is a broad spectrum of differential diagnosis (17). Moreover, this case showed a mixed pattern of lysis and sclerosis and new bone formation throughout the lesion. These features suggested another possibility of bone forming tumor. In general, radiologic findings are well correlated with the degree of histologic differentiation; low-grade lesions show well-marginated areas of lysis, sometimes associated with a sclerotic rim and prominent new bone formation, whereas high-grade lesions are commonly associated with cortical destruction and the production of soft tissue mass (5). Retrospectively, radiologic findings of this case are thought to be compatible with those of low-grade hemangioendothelioma.

Hemangioendotheliomas are characterized by infiltrative proliferation of rich vascular spaces lined by endothelial cells with modest cytologic atypia (18). Immunohistochemical study is helpful in confirming the diagnosis by identifying factor VIII-related antigen, which is a marker for vascular endothelial cells (9). Also, ultrastructural studies confirm the endothelial origin of the tumor. Hemangioendothelioma is distinguished from hemangioma by the more primitive vasoformative appearance and infiltrative growth (5, 13, 18). Angiosarcoma is separated from this tumor by nuclear anaplasia, conspicuous anastomosing vascular channels and brisk mitotic activity (5, 13, 18). The histologic features of this case, such as mild nuclear atypism of endothelial cells, infiltrative growth pattern, relatively well vasoformative appearance and absence of mitosis, are compatible with low-grade hemangioendothelioma. Unni et al. (4), in a review of 22 cases of hemangioendothelioma of bone, divided the tumors into three distinct histologic grades and stated that the most important indicator of prognosis was the grade of anaplasia. Three grades have been delineated based on the degree of vasoformative appearance, the pleomorphism of neoplastic cells and mitotic figures (4, 5, 13). Although patients were treated with a variety of modalities and comparisons are difficult, there was a definite decrease in disease-free survival with increasing grade (grade 1, 95%; grade 2, 62%; and grade 3, 20%) (19).

Initial ablative surgical procedure with en bloc resection is generally considered to be the treatment of choice (5, 8, 10), but sometimes it is impossible because of

extensive bleeding during surgery (7, 10, 11). This patient was operated on twice and the tumor was completely resected. Postoperative radiation was deferred because the tumor was completely excised and the histologic grade of the lesion was mainly grade 1. She is free of disease and has normal visual fixation and ocular motility 6 months postoperatively.

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