

High Grade Hemangioendothelioma of the Temporal Bone in a Child: A Case Report

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Hemangioendothelioma is a rare vascular tumor characterized by endothelial tumor cells and variable malignant behavior, and it's not common for this lesion to involve the bone. Although there are a few reports of cranial involvement by hemangioendothelioma, only rare cases arising in temporal bone have been published. We present the radiologic findings of a 7-year-old boy who had a high grade hemangioendothelioma involving the temporal bone with intracranial extension. Evidence of flow voids on MR images suggested a tumor of vascular origin, and the ill-defined margins, cortical destruction and intracranial extension on the CT and MR images were correlated with the tumor's high histologic grade.

Index terms:

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Hemangioendothelioma is a rare vascular tumor composed of endothelial cells having a range of malignant behavior, and this tumor can involve soft tissue or bone (1). The incidence of the hemangioendothelioma has been reported as 0.5 - 1.0% of primary malignant bone tumors (2). Although this tumor usually involves the long bones in the cases of bony involvement, relatively rare cases arising in skull have been reported. To the best of our knowledge, the radiologic findings of only few cases involving the temporal bone in children have been published (2, 3). We present here a case of high grade hemangioendothelioma of the temporal bone in a child, and we describe the plain film, CT, MR and angiographic appearance of this tumor, as well as its histopathologic findings.

CASE REPORT

A 7-year-old boy visited to our hospital complaining of a slowly growing mass in his right posterior auricular region for 3 months. On physical examination, about a 3 cm sized pulsating soft mass in the posterior auricular region was palpated. There was no definite abnormality on the laboratory findings.

Plain radiographs of the skull revealed a relatively well-defined, osteolytic lesion in the right temporal bone. No evidence of sclerotic margin or periosteal reaction was found (Fig. 1A). Temporal bone CT revealed about a 5 cm sized, soft tissue density mass with marked bone destruction that mainly involved the mastoid portion of the right temporal bone (Fig. 1B). The mass showed dense heterogeneous enhancement and intracranial extension with a large area of necrosis.

On T1-weighted MR images (TR/TE, 509/14), the mass was heterogeneously hypointense with some high signal foci and signal void dots (Fig. 1C). The T2-weighted MR images (TR/TE, 4225/100) revealed a heterogeneous signal intensity mass with multiple signal void dots (Fig. 1D). After an intravenous infusion of contrast media, the main mass involving the temporal bone was intensely enhanced (Fig. 1E). This

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lesion appeared to extend intracranially at its superior aspect, and there was associated peritumoral edema in the adjacent temporal lobe. The intracranial portion of the mass showed intermediate signal intensity on the T1-weighted images and homogenous high signal intensity on the T2-weighted images, and there was peripheral rim enhancement that represented necrosis (Fig. 1E).

A large, ill-defined, hypervascular mass involving the right temporal region was seen on the early arterial phase of the external carotid arteriogram, and the mass was mainly supplied by petrosal branches of the middle meningeal artery (Fig. 1F). The lesion still remained hypervascular on the late venous phase of the arteriogram, and this was due to delayed washout of contrast media

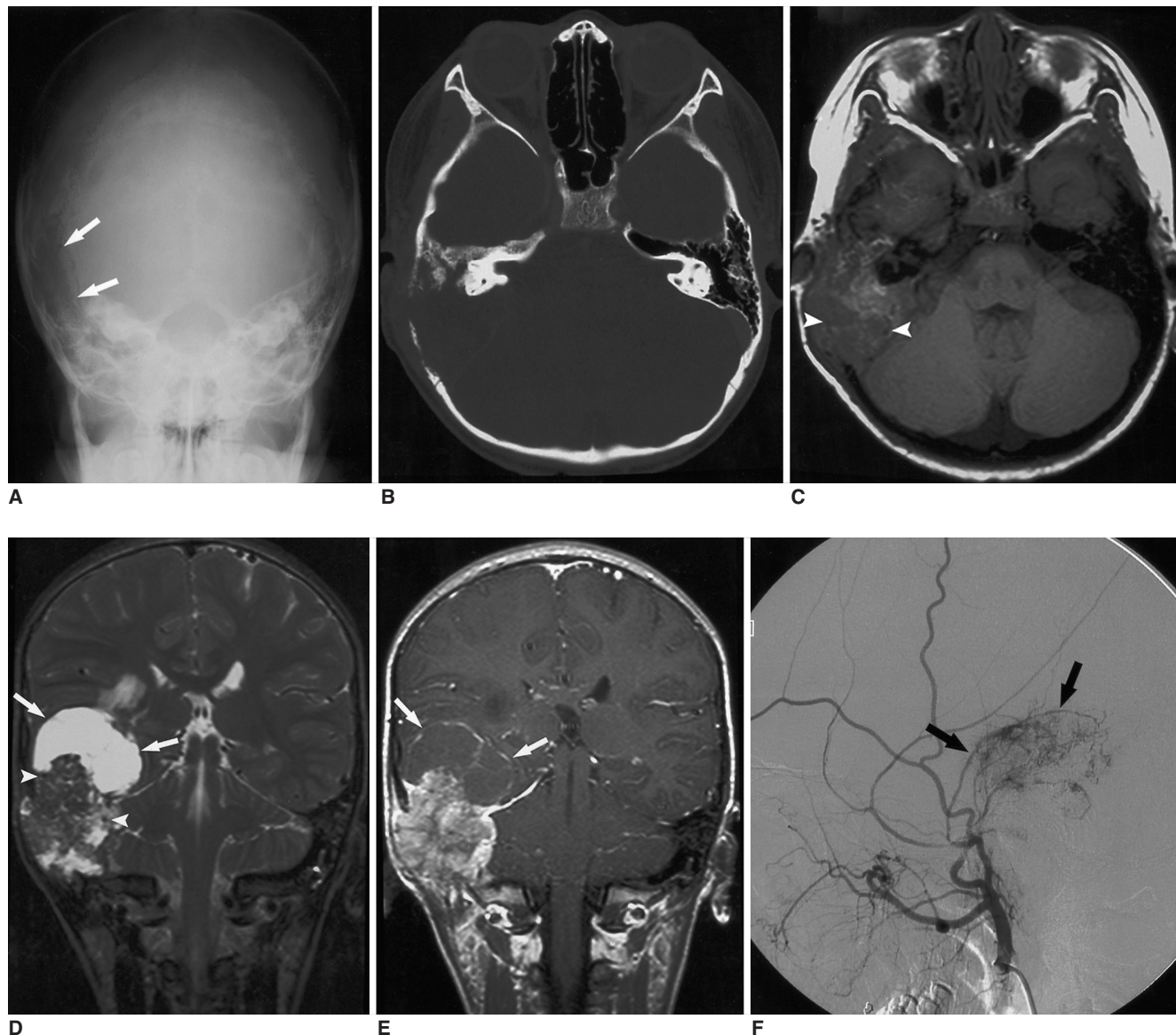


Fig. 1. A 7-year-old boy with high grade hemangioendothelioma of the right temporal bone.

A. Plain radiograph shows a relatively well-defined osteolytic lesion in the right temporal bone (arrows).

B. Axial CT scan with bone window shows the marked bone destruction, with ill-defined margins, involving the mastoid portion of the right temporal bone.

C. Axial T1-weighted MR image (TR/TE, 509/14) reveals the main mass involving the right temporal bone with heterogeneous signal intensity and signal void dots (arrowheads).

D. Coronal T2-weighted image (TR/TE, 4225/100) shows temporal portion of the mass to be heterogeneously hypointense with multiple signal void dots (arrowheads), and the supratentorial portion to be homogeneously hyperintense (arrows).

E. The coronal contrast-enhanced T1-weighted MR image (TR/TE, 509/14) reveals heterogeneous enhancement of the main mass in the temporal bone and peripheral enhancement of the intracranial portion (arrows).

F. Lateral view of the right external carotid angiogram shows a large, ill-defined hypervascular mass in the right temporal region, which is mainly supplied by petrosal branches of the middle meningeal artery (arrows).

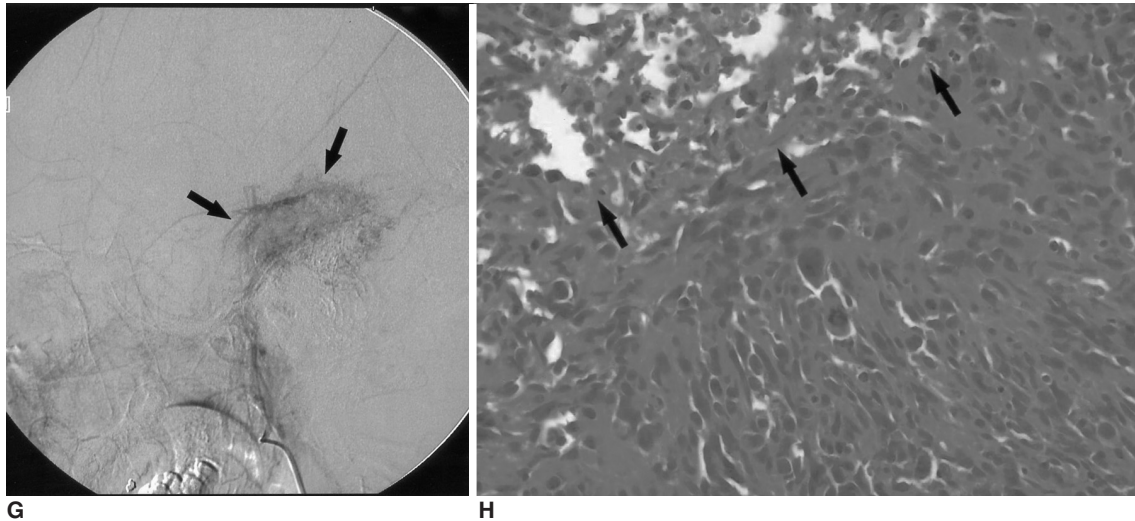


Fig. 1. **G.** The lesion still remained hypervascular on the late venous phase due to delayed washout of contrast media. Arteriovenous shunt was not demonstrated.

H. Photomicrograph shows the pleomorphic, highly atypical spindle cells with focal luminal differentiation filled with erythrocytes, and the irregular anastomosing vascular channels (arrows) (hematoxylin-eosin staining, $\times 200$).

(Fig. 1G). No arteriovenous shunting could be observed.

The preoperative diagnosis was malignant tumor of a vascular origin and the differential diagnosis included other sarcomas such as rhabdomyosarcoma.

The patient underwent an incomplete tumor resection due to massive bleeding via the right temporal approach. The removed tumor was a brown colored fragile soft tissue mass that measured up to $4.5 \times 5.0 \times 6.5$ cm. The intracranial portion of the mass was noted to have hemorrhagic necrosis. Histologically, the tumor was composed of short strands or solid nests of pleomorphic and highly atypical spindle cells. The cells showed relatively abundant eosinophilic cytoplasm, bizarre nuclei and frequent mitotic figures including the atypical forms. The tumor showed focal luminal differentiation filled with erythrocytes (Fig. 1H). A vascular origin for these cells was confirmed by the positive test for CD34, a specific endothelial marker. This tumor was finally diagnosed histologically as a grade III hemangioendothelioma.

Following the surgery, the patient underwent radiotherapy. At one year after the initial disease presentation, the boy ominously returned the emergency room due to dyspnea, and the chest radiograph we took showed bilateral pneumothorax with multiple cavitary lung metastases. Sadly, two years after the initial disease manifestation, our patient succumbed to his illness despite our best efforts using radiotherapy and concurrent chemotherapy.

DISCUSSION

Hemangioendothelial tumors are rare vascular tumors, so there remains confusion concerning the naming of them (4). Some authors have referred to these endothelial tumors of bone by various names such as angiosarcoma, hemangioendothelial tumor and hemangioendothelial sarcoma (4). However, malignant endothelial tumors of bone have generally been described as hemangioendothelioma of bone, and hemangioendothelioma is divided into three grade according to tumor differentiation (4, 5).

Hemangioendothelioma can affect any bone, but involvement of the skull including the temporal bone is extremely rare. Pain is usual a presenting symptom of this tumor, and it is observed as a local mass or swelling in the case of skull involvement (5, 6). Most of these tumors arise in the third decade, although they has been reported to occur in almost all age group (4, 5). It is more frequently seen in men than in women (4, 5), and it has a tendency of multicentricity in 22% of patients (4).

Microscopically, the essential feature of this tumor is a neoplastic vasoformative appearance, with the mass of the tumor being composed of vascular channels in various stages of angiogenesis and epithelial cells with abundant eosinophilic cytoplasm (4, 5).

As was previously mentioned, Unni et al. divided the tumors into three distinct histologic grades based on the degree of the vasoformative appearance, pleomorphism of the neoplastic cells and the mitotic figures.

Hemangioendotheliomas are graded on a scale I to II, and

grade III represent anaplastic tumors, which also known as angiosarcoma (7). The histologic grade is the most important indicator of prognosis, and overall survival rates for patients with grade I, II and III lesions were 95%, 63% and 20%, respectively (4). For ambiguous cases, immunohistochemistry is helpful for demonstrating the vascular nature of tumor cells; these types of cells will express factor VIII associated protein, CD 34 and ULEX europaeus (8). In our case, the tumor cells were positive for CD 34.

The most frequent radiographic finding of hemangioendothelioma is osteolytic lesion (9). Calcifications and periosteal reactions are unusual (4). The MR findings of hemangioendothelioma are nonspecific (10). The signal intensity of these vascular structures may display as either high flow (low signal intensity on images of all pulse sequences) or low flow (high signal intensity on the T2-weighted images) (8). The other sarcomas, even though they are hypervascular, rarely show the definable prominent vessels on MR imaging (10). Rodolfo et al. (2) has reported on grade II hemangioendothelioma of the temporal bone with flow voids on the MR images. In our case, multifocal signal void dots within the tumor mass on MR images were also noted, and this raised the possibility of a tumor of vascular origin.

Our case revealed an ill-defined lesion that involved the temporal bone with marked bony destruction, intracranial involvement and distant metastasis; all of these factors correlated well with the histologic grade. Radiologic findings for one case of grade III hemangioendotheliomas of the skull have been reported as a well-enhancing expansile mass in the calvarium with extension into the brain (9). However, that particular case did not include signal void dots or large area of necrosis, like our case.

The treatment of the hemangioendothelioma usually consists of wide surgical excision and postoperative radiotherapy, and the therapy should be dictated by the grade and location of tumor. Radiation may play an important role for the therapy of patients with multicentric

low grade tumors or surgically inaccessible tumors (4). Chemotherapy may be useful for treating patients with grade III tumor, but too few patients have been treated to be able to generalize on the final results (4).

In conclusion, the occurrence of hemangioendothelioma in the temporal bone in a child is extremely rare. Prominent serpentine vessels on the MR images should suggest neoplasm of a vascular origin, and marked osseous destruction and an associated intracranial extension with area of necrosis can suggest the possibility of high grade lesion.

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