Primary Hemangiopericytoma of the Parietal Bone: A Case Report

Parietal Kemikte Primer Hemanjioperistom: Olgu Sunumu

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Summary

Hemangiopericytomas are rare hypervascular tumors arising from Zimmerman’s pericytes. They usually occur in the soft tissue, and intraosseous lesions are very rare. Surgical excision is the first choice for treatment. Many studies show that patients should be monitored for some time following treatment because of a high rate of recurrence and metastasis after radical resection. This report introduces a 56-year-old patient with a hemangiopericytoma in his parietal bone.

Keywords: Parietal bone, Hemangiopericytoma

Özet


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Anahtar Kelimeler: Parietal kemik, Hemanjioperistoma, Primer
Introduction

Emangiopericytomas can develop wherever pericytes are present and are rare and potentially malignant vascular tumors. Stout and Murray first described these tumors in 1942 [1]. They usually develop in the extremities and pelvic and retroperitoneal regions, and primary bone localization is very rare. Symptoms depend on the tumor’s location, size and potential for malignancy [2]. This case report draws on relevant literature to present a very rare hemangiopericytoma located in the parietal bone.

Case Report

A 56-year-old man arrived in our neurosurgery clinic with a painful lesion on his head that had been present for two months. During the physical exam, a painful, fixed tumor was detected on his right parietal region. It appeared as a round and osteolithic region on an X-ray, and a CT scan revealed a 34 mm by 37 mm hypodense, osteolithic tumor with an extra-axial soft-tissue component lying within the epidural space (Fig. 1). There was no bone beneath the lesion. Radiological findings showed that the lesion was primarily derived from bone, and we decided to operate. Surgery revealed a smooth, grey-yellow lesion that had eroded the periosteum of the parietal bone; we were able to completely excise it. Histopathological examination showed irregular neoplastic cells with indistinct cytoplasmic borders covering the vascular projects (Fig. 2, A). Immunohistochemical analysis showed that these neoplastic cells were positive for vimentin and CD34 (Fig. 2, B) but negative for S-100, SMA and CK. Based on morphological examinations and immunophenotyping, we diagnosed the tumor as hemangiopericytoma. No metastases or signs of recurrence were detected within 9 months of the excision.

Discussion

Hemangiopericytoma is a rare mesenchymal tumor originating from pericytes, which are contractile cells surrounding capillaries [1]. The tumor is found most often in the pelvic area, proximal femur, vertebrae and humerus, but it can occur anywhere. Primary intraosseous localization is very rare. Hemangiopericytomas have been described in individuals of all ages, although more than 40% occur in individuals in their 50s and 60s [2]. The tumor is not associated with one sex over the other.

Clinically, hemangiopericytomas may present with pain or a mass. Pain in the parietal region was present in our case. The lesions grow slowly and may be present for twenty years before a diagnosis is made. Hemangiopericytomas have almost no distinguishing radiological characteristics. They may be lytic or cause focal sclerosis, or they may have a honeycomb or reticular pattern. They may also cause cortical erosion, which suggests the presence of a malignancy. CT scans and MRIs are not helpful in the diagnostic process, but they can help distinguish between benign and malignant lesions and help define the extent of the tumor. Angiography reveals spider-like radial branching vessels.

On gross examination, hemangiopericytomas may be well-circumscribed and appear grayish-white; they are much less hemorrhagic than endothelial tumors. The tumor cells are bapsophic spindle-shaped mononuclear cells that look like smooth muscle cells. The nuclei are round or oval and homogeneous. The cells have indistinct cytoplasmic borders. The tumor cells do not arise from endothelial cells even though they surround irregular vascular spaces. The branching vascular channels of varying sizes are often described as forming a ‘staghorn’ pattern. Silver stain highlights the reticulin sheath that surrounds each cell and confirms that the tumor cells are outside the vascular spaces [1]. The fact that the tumors cells do not stain positive for actin suggests that they may originate in cells other than pericytes.

The blood vessels are lined with normal endothelial cells, in contrast to malignant angiosarcoma, in which the vascular spaces are lined with malignant tumor cells. Under histological examination, several other tumors may show a similar vascular pattern and resemble hemangiopericytoma. Fibrous histiocytoma, synovial sarcoma and mesenchymal chondrosarcoma should be considered in the microscopic differential diagnosis. Fibrous histiocytoma shows a storiform or cartwheel pattern and a less prominent vascular network. Synovial sarcoma may show a biphasic cellular pattern and include fibrosarcoma-like areas. Mesenchymal chondrosarcoma cells are smaller than those of a hemangiopericytoma, and well-defined islands of cartilage are present [3,4].

Immunohistochemical analysis provides important informa-
tion that can distinguish hemangiopericytomas from other vascular neoplasms and mesenchymal tumors [5,6]. Antibodies against CD31, CD34, CD68, vimentin and cytokeratins are used for this analysis. The antibodies against CD34 and vimentin were used successfully in our study. These antibodies are characteristic of tumors with a mesenchymal origin and can identify neoplastic progenitor cells surrounding vascular spaces [5].

Hemangiopericytomas can be classified into one of three groups: benign, borderline or malignant. We examined the mitotic ratio (10 HPF<4), cellularity, pleomorphism, hemorrhaging and necrosis of the tumor to determine malignancy. There were 3-4 mitoses and mild pleomorphism. No metastasis or signs of recurrence were detected after 9 months [6,7].

The treatment of hemangiopericytoma of the bone is primarily surgical. Initial wide resection is mandatory if a cure is to be expected, and this procedure was completed in the present case. The role of chemotherapy in the treatment of the tumor is unknown. Radiation therapy has usually been reserved for unresectable and recurrent tumors or for palliative care [8].

Enzinger and Smith [1] found that more than two-thirds of the cases that eventually metastasize develop local recurrences first. The lungs are the most common site of metastasis. Several studies have shown that long-term follow-up of patients is necessary even after radical resection because recurrence or metastasis may be delayed by many years. We found no evidence of recurrence or metastasis in this case after 9 months.

**Conflict interest statement** The authors declare that they have no conflict of interest to the publication of this article.

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