Multiple epithelioid hemangioendothelioma of the skull in a child

A case report

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Abstract

Introduction: Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor characterized by neoplastic proliferation of epithelioid or histiocytoid epithelial cells. EHE of bone constitutes <1% of primary malignant bone tumor. EHE in the skull is an extremely rare case. Here, we report a case of multiple neoplasm of the skull in a 15-month-old boy who presented with gradual facial swelling for 2 months. On computed tomography (CT) scan, multiple irregular osteolytic lesions were seen on the right maxillary, sphenoid, left zygoma, and roof of the left orbit. Excisional surgery of the lesion was planned. Histopathological and immunohistochemical examination of excised specimen suggest it to be epithelioid hemangioendothelioma. Follow-up for 6 months showed no recurrence.

Conclusion: Epithelioid hemangioendothelioma is a locally aggressive tumor with metastatic potential. CT imaging could help in assessment of lesion, but final diagnosis is possible only with histopathology. Complete surgical resection at the early stage of the disease is the most effective treatment with better prognosis.

Abbreviations: CT = computed tomography, EHE = epithelioid hemangioendothelioma.

Keywords: computed tomography, epithelioid hemangioendothelioma, skull

1. Introduction

Epithelioid hemangioendothelioma (EHE) is a rare vascular neoplasm characterized by neoplastic proliferation of epithelioid or histiocytoid epithelial cells. Its clinical course shows features between hemangiomia and angiosarcoma.[1] EHE affects all age groups and has higher predilection in female than male. Although could present in multiple sites, it is commonly seen in soft tissue, skin, liver, pleura, peritoneum, and lymph nodes. EHE of bone constitutes <1% of primary malignant bone tumors.[2] EHE in the skull is extremely rare. Here we report a case of multiple EHE originating from the skull in a 15-month-old boy; the image findings and histopathological features were also discussed.

2. Case report

A 15-month-old boy was admitted to our hospital with 2-month history of gradually increasing facial swelling after he fell down from bed. The swelling was accompanied by pain, which on later days interferes with the normal eye opening. He then went to one local hospital. Evaluation of the swelling by CT scan showed multiple osteolytic lesions. He was then transferred to our hospital (The 1st Affiliated Hospital of Sun Yat-Sen University) for tertiary care. His medical history was not significant. On physical examination, left zygoma was slightly swollen with no skin redness or tenderness, no palpable lymph nodes detected.

Laboratory examination showed all parameters within normal limits. On CT, multiple irregular osteolytic lesions were seen on the right maxilla, sphenoid bone, left zygoma, and roof of the left orbit. The tumor showed irregular isodense (to soft tissue) lesion with ill-defined margin; destruction of outer bony cortex and lamina dura is also seen (Figure 1). Tumor stroma showed mild enhancement after contrast administration (Figure 1). Radiologically it was diagnosed as neoplastic lesion, but nature of tumor could not be determined.

The patient underwent excision surgery of the lesion. During the surgery, the left and right side of mucoperiosteum were separated to the zygomatic neoplasm and maxilla respectively. The specimen was then sent for pathological examination. Some of the cells were arranged in a tube-like structure. (Fig. 2) The specimen was then sent for pathological examination. Neoplastic lesion was seen. The cells were arranged in a tube-like structure. The specimen was then sent for pathological examination. Some of the cells were arranged in a tube-like structure. The specimen was then sent for pathological examination. Some of the cells were arranged in a tube-like structure. (Fig. 2) On immunohistochemistry, the tumor cells showed CD34(+), CD31(+), D2-40(−), Actin(−), HHF-35(−), Myogenin(−), MyoD1(−), S-100(−), Syn(−), Ki-67 10%(+) (Fig. 2). Finally the diagnosis was confirmed to be infant epithelioid hemangioendothelioma. Recovery was uneventful and the patient was discharged 10 days after the surgery. Six-month follow-up showed no local recurrence. This study was approved by the First Affiliated Hospital of Sun Yat-Sen University Institutional...
Review Board. Written consent for this case report was obtained from the patient.

3. Discussion

Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor characterized by neoplastic proliferation of epithelioid or histiocytoid epithelial cells. The causes of EHE are still a dilemma, but most believed it to be because of chromosomal mutation in gene involved with angiogenesis. Some associated with vascular dysplasia, trauma, oral contraceptives, abnormal estrogen, and progesterone levels is also reported. They are locally aggressive tumors and have potential for metastasis. EHE affects all age groups and has higher predilection in females than males. Although could present in multiple sites, it is commonly seen in soft tissue, skin, liver, pleura, peritoneum, and lymph nodes. Primary bone epithelioid hemangioendothelioma is rare and accounts for only 0.5% to 1.0% of primary malignant bone tumors. Clinically EHE could be asymptomatic or can present with local pain, tenderness, and swelling, leading confusion with infectious cause like pyogenic granuloma. Our case was presented with pain and swelling but lack of other inflammatory signs and normal blood report ruled out infectious cause.

CT scan can be very helpful in radiological diagnosis. The most common radiographic finding of bone EHE is an irregular osteolytic bone destruction with CT value of 15-50HU. The cystic or honeycomb configuration could be seen in the cortex and medulla. Multiple lesions within one bone or within multiple bones was reported. The presence of regional multifocal involvement could provide clue for tumor of vascular origin. As for skull EHE, the imaging report is extremely rare. The image manifestation of skull EHE resembles that of other bones. Our case showed multiple irregular osteolytic bone destruction with local cortical defect. Intracranial involvement was not observed, which could probably because of low histologic grade of our case. CT played a critical role in the evaluation of bone EHE. Despite all these radiological findings, definitive diagnosis of EHE is still based on histopathological analysis.

The characteristic pathological feature of EHE is round to slightly spindled eosinophilic neoplastic epithelioid cells. Tumor cells are arranged either as anastomosing cords, solid nests, or short strands and are embedded in a chondroid-like or hyalinized stroma. The tumor cells may show mild atypia, increased mitosis, and/or occasional necrosis. According to the morphology and differentiation of angioblast, EHE is classified into 3 stages, higher the grade worse the prognosis. EHE always exhibits immunopositivity for vimentin and endothelial markers, such as CD31 and CD34. In our case, immunohistochemistry of the tumor cells showed CD34(+), CD31(+), which was diagnosed as infant EHE finally.

Figure 1. CT images showed multiple irregular osteolytic bone destruction in the right maxillary, sphenoid bone, left zygoma, and roof of the left orbit, and destruction of cortical bone (white arrow) and lamina dura is seen (black arrow). Tumor stroma showed slight enhancement after contrast agent administration.

Figure 2. Histopathological examination of the skull epithelioid hemangioendothelioma (hematoxylin and eosin staining). Nested short spindle cells were found. Some of the cells were arranged in a tube-like structure.
En bloc resection is the commonly used modality in the treatment of EHE disease. Mass curettage was performed in our case and no recurrence was noted during 6 months’ follow-up.

In conclusion, epithelioid hemangioendothelioma of skull is a kind of low-grade malignant tumor of vascular origin. CT imaging could be helpful for assessing the lesion, but final diagnosis is possible only by histopathology. Complete surgical resection at the early stage of the disease is the most effective treatment with good prognosis.

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