Primary pericranial Ewing’s sarcoma on the temporal bone: A case report

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Abstract

Background: Primary Ewing’s sarcoma originating in the pericranium is an extremely rare disease entity.

Case Description: A 9-year-old female patient was admitted to our department due to a left temporal subcutaneous mass. The mass was localized under the left temporal muscle and attached to the surface of the temporal bone. Head computed tomography revealed a mass with bony spicule formation on the temporal bone, however, it did not show bone destruction or intracranial invasion. F-18 fluorodeoxyglucose positron emission tomography showed no lesions other than the mass on the temporal bone. Magnetic resonance imaging showed that the mass was located between the temporal bone and the pericranium. The mass was completely resected with the underlying temporal bone and the overlying deep layer of temporal muscle, and was diagnosed as primary Ewing’s sarcoma. Because the tumor was located in the subpericranium, we created a new classification, “pericranial Ewing’s sarcoma,” and diagnosed the present tumor as pericranial Ewing’s sarcoma.

Conclusion: We herein present an extremely rare case of primary pericranial Ewing’s sarcoma that developed on the temporal bone.

Key Words: Ewing’s sarcoma, pericranium, skull, temporal bone

INTRODUCTION

Ewing’s sarcoma, a small round cell tumor that was first described by James Ewing in 1921, is most commonly diagnosed in the second decade of life. Although it is the second most common form of primary bone cancer in childhood, the primary involvement of the calvaria is rare. Periosteal Ewing’s sarcoma is also rare and has been reported to affect the long bones. Here, we report a case of primary Ewing’s sarcoma originating in a pericranial location with mild invasion into the temporal bone.

CASE DESCRIPTION

A 9-year-old girl had been well until 2 months before admission when she noticed a small subcutaneous mass in the left temporal region. The mass thereafter rapidly became larger. A skull X-ray at the first hospital showed no bone defects or expansion; however, a skull X-ray with dynamic range compression modification showed a sunburst appearance on the left temporal bone [Figure 1a]. Computed tomography (CT) of the head also showed a sunburst appearance due to bony...
spicule formation in a soft mass that was attached to the surface of the left temporal bone, without osteolysis, bone expansion, or cortical destruction [Figure 1b]. She was referred to our hospital for further examinations and treatment. She was healthy, with no history of recent systemic symptoms or relevant medical history. A physical examination showed a firm, unmovable, mildly tender, and slightly elastic subcutaneous mass measuring 5 × 5 cm in size in the temporal region with apparently normal overlying skin. She had no cervical lymph node swelling. Her general, systemic, and neurological statuses were normal, as well as her blood examination results. 

F-18 fluorodeoxyglucose positron emission tomography/computed tomography (F-18 FDG PET/CT) detected a highly-labeled extracranial mass lesion on the nonlabeled temporal bone without metastasis [Figure 1c]. Magnetic resonance imaging (MRI) showed a mass lesion between the temporal muscle and the lamina externa of the temporal bone without intracranial extension. The mass lesion exhibited isointensity on T1-weighted imaging (T1WI) and T2-weighted imaging (T2WI) and was heterogeneously enhanced on the postcontrast T1WI [Figure 1d-f]. Postcontrast T1WI showed the enhancement of the outer and inner membranous structures that were connected to the pericranium [Figure 1e, f; arrows and arrowheads]. For the radical removal of the tumor, the soft tumor was separated from the temporal muscle along an overlying thin membrane that appeared to be the pericranium and which likely corresponded to the outer membrane on postcontrast T1WI. The tumor was dissected from the surface of the temporal bone and removed with the deep layer of the temporal muscle. An intraoperative histological examination confirmed the margin to be negative. The outer surface of the temporal bone was slightly irregular, suggesting invasion of the tumor from the outside into the bone. The temporal bone was also resected with a margin of 1 cm. Bone replacement material was used to fill the bone defect, and then cranioplasty was performed with absorbable plates and screws. The histopathological findings were consistent with Ewing’s sarcoma [Figure 2a]. The tumor was composed of small round cells that were strongly positive for CD99, focally positive for s-100, and negative for cytokeratin (AE1/AE3), desmin, myogenin, LCA, and TdT. The tumor was positive for EWS-FLI-1 translocation. The tumor was mainly an extracranial mass that was attached to the outer surface of the temporal bone with bone reaction on the surface, and with local invasion into the temporal bone [Figure 2b-d]. It infiltrated into some marrow cavities under very thin compact bone [Figure 2b-d]. Postoperative postcontrast T1WI showed no residual enhancement of the mass lesion. At 24 days after the operation, chemotherapy (vincristine, doxorubicin, and cyclophosphamide alternating with ifosfamide and etoposide [VDC/IE]) was administered. Local radiation therapy was administered after the third cycle of chemotherapy. She completed her course of chemotherapy at 9 months after the operation and is currently in complete remission.

Figure 1: Preoperative images. A skull X-ray with dynamic range compression (a), axial computed tomography of the skull (b), F-18 fluorodeoxyglucose positron emission tomography (c), axial T2-weighted imaging (d), and postcontrast T1-weighted imaging of the axial plane (e) and coronal plane (f). Arrows and arrowheads indicate the enhanced outer and inner membranous structures, respectively
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In the present case, Ewing’s sarcoma was located between the pericranium and the temporal bone. The main imaging and histopathological findings were as follows. First, the skull X-ray and CT showed a sunburst appearance as a periosteal reaction, without the presence of a bone defect. Second, MRI showed two enhanced membranous structures that surrounded the tumor and were connected to the pericranium. Third, although the tumor was mainly located outside the temporal bone, the histopathological analysis revealed that it had invaded some of the marrow cavities in the temporal bone. These findings suggested pericranial Ewing’s sarcoma with mild invasion into the membranous bone.

Ewing’s sarcoma, a small round cell tumor, is most often diagnosed in the second decade of life and is the second most common form of primary bone cancer in childhood.\[9\] Between 1973 and 2004, the annual incidence of Ewing’s sarcoma in the United States was 2.93 cases per million.\[6\] In approximately half of the cases, the tumors were located in the patients’ extremities.\[9\] In contrast, the primary involvement of the skull only occurred in 1–2% of Ewing’s sarcomas.\[1,17\] Intracranial extension was present in the majority of primary calvarial Ewing’s sarcomas; extracranial extension was only observed in 8 cases of primary calvarial Ewing’s sarcoma.

The periosteal reaction is a rare radiographic appearance in primary Ewing’s sarcoma cases that involve the bones of the head and neck.\[23\] Cortical thickening was observed in only 8% of the cases, whereas permeative changes were recognized in 54% of cases. In contrast, pure lytic changes (58%) and honeycombing (21%) are more common among these patients; among other primary Ewing’s sarcoma patients, pure lytic changes and honeycombing occurred in 18.6% and 5.8% of cases, respectively. Bone expansion, which occurs in 25% of the head and neck cases, and cortical violation, which occurs in 50% of the cases, are also much more frequent than in the general population.\[23\] Among the 8 previous cases of primary calvarial Ewing’s sarcoma that involved extracranial extension, osteolytic destruction or cortical violation was observed in 7 cases; the remaining case was a congenital case in which Ewing’s sarcoma occurred in the frontonasal area.\[2,5,8,11,13,15,16,25\] Although a sunburst periosteal reaction was observed in the present case, osteolytic lesions, honeycomb lesions, bone expansion, and cortical violation were not observed on the skull X-ray or CT [Figure 1a, b]. F-18 FDG PET/CT showed a strongly labeled tumor on the surface of the nonlabeled temporal bone [Figure 1c]. Postcontrast T1WI showed enhanced outer and inner membranous structures that formed the tumor boundaries and which were connected to the pericranium; the structures were considered to be the pericranium and a reactive attachment of the tumor to the surface of the temporal bone, respectively [Figure 1e, f]. A histopathological examination showed that there was little destruction of the underlying temporal bone with local infiltration of the tumor into the marrow cavities [Figure 2b–d]. These imaging and histopathological findings suggested that the tumor occurred at a subpericranial location.

Periosteal Ewing’s sarcoma is a rare form of Ewing’s sarcoma that particularly affects the femur.\[9\] Periosteal Ewing’s sarcoma is defined as a subperiosteal tumor without any medullary involvement. In the long bone, the medullary cavity (yellow bone marrow) is covered by hard and thick compact bone on which periosteal Ewing’s sarcoma may occur. In the present case, the temporal bone had marrow cavities in the compact bone immediately under the surface [Figure 2b, d] and mild invasion into the compact bone resulted in invasion into the marrow cavities [Figure 2c]. The present case was not considered to meet the criteria for the definition of periosteal Ewing’s sarcoma because the tumor involved the marrow cavities. However, it is difficult to compare the present case with previous cases of periosteal Ewing’s sarcoma affecting the long bones because there are structural differences between the long bones and the membrane bones, for example, the thickness of the compact bone (as mentioned above). Because the preoperative images and the operative findings indicated...
that the tumor was mainly located in the subperiosteal region, we created a new classification, “periosteal Ewing’s sarcoma,” and diagnosed the present case as primary periosteal Ewing’s sarcoma. There is a previous report of a case of primary calvarial Ewing’s sarcoma attached to the skull in which the tumor was completely extracranial.[10] Naidu reported that there was no evidence of any communication with the intracranial contents at the site of the attachment and only irregularity of the outer table of the skull. However, it is difficult to define that case as periosteal Ewing’s sarcoma because the CT and MRI findings were not provided. To the best of our knowledge, the present study represents the first report of periosteal Ewing’s sarcoma.

Then, what were the cells of origin in the present case? Ewing’s sarcoma expresses the EWS-FLI-1 fusion gene, which is generated by chromosomal translocation, whereas EWS-FLI-1 induces cell cycle arrest in normal murine and human fibroblasts because of EWS-FLI-1-mediated cytotoxicity.[14] This causes difficulty in inducing Ewing’s sarcoma, suggesting that the target cells of EWS-FLI-1 might be the cells of a narrow lineage and/or a limited differentiation stage.[24] The current view is that the disease arises from mesenchymal or neural crest-derived stem or progenitor cells.[12,19,24] The temporal bone arises from neural crest cells, which go through an epithelial-to-mesenchymal transition and migrate away from the neuroepithelium.[10] During intramembranous ossification, neural crest derived stem or progenitor cells will condense and differentiate into osteoprogenitor cells, which in turn give rise to osteoblasts, the mature bone-forming cells.[20] Undifferentiated progenitor cells remain at the bone periphery, forming the pericranium.[26] We, therefore, hypothesize that the cells of origin in the present case might be neural crest derived stem or progenitor cells in the pericranium. In the cranial bone marrow, there are also neural-crest derived stem cells, which might give rise to Ewing’s sarcoma in the cranial bone marrow.[22] The periosteal and meningeal dura are present on the inner surface of the cranial bone. Similar to the pericranium, the dura mater has been reported to induce osteogenesis and to have stem cells, which suggests that dura mater stem cells might give rise to intracranial Ewing’s sarcoma.[18,20]

**CONCLUSION**

We herein reported the first known case of primary pericranial Ewing’s sarcoma which occurred in a 9-year-old girl.

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**Conflicts of interest**

There are no conflicts of interest.

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