Ewing's sarcoma of the temporal bone: case report of a child and review of the literature

Abstract: Ewing's sarcoma is the second most common primary bone malignancy in children after osteosarcoma. It represents 4 to 9% of primary malignant bone tumors and affects the bones of the skull (maxillary, frontal, parietal, ethmoid, temporal bones) in only 1 to 4% of cases. Therefore, it rarely affects the head and neck. In this case report, we describe a case of a 13-year-old boy with left temporal Ewing sarcoma. When this patient presented with left temporal swelling and headache, imaging studies showed a mixed mass originating from the left temporal bone. During the biopsy and surgery (incomplete resection), the mass was invaded the facial nerve, which then required a nerve transplant. Postoperatively, he then underwent radiotherapy with induction and adjuvant chemotherapy. Although it is a rare tumor of the temporal bone, physicians should consider Ewing's sarcoma in the differential diagnosis of children and adolescents with facial nerve palsy. With a generally poor prognosis due to early metastasis to the lungs and other bones.

Keywords: Ewing's sarcoma skull, temporal bone, chemotherapy, radiotherapy.

INTRODUCTION:

Ewing's sarcoma is a primary bone cancer that primarily affects children and adolescents. It is one of a group of cancers known collectively as the Ewing sarcoma tumor family. It is the second most common bone cancer in children, but it is also relatively rare (Esiashvili, N. et al., 2008; & Horowitz, M.E. et al., 1992). It only accounts for 1% of all childhood cancers. Although it can occur at any age, it rarely occurs in adults over the age of thirty. Ewing's sarcoma accounts for 4% to 9% of primary malignant bone tumors (Esiashvili, N. et al., 2008; & Arndt, C.A., & Crist, W.M. 1999) and is most commonly seen in long bones or the pelvis. Primary Ewing sarcomas occurring in the skull are exceptionally rare. The prognosis for Ewing sarcoma is often poor due to early metastasis to the lungs or other bones. However, a review of articles suggests that metastases are much less common and long-term survival can be expected in Ewing's sarcoma occurring in the skull.

CASE REPORT:

A 13-year-old boy was doing well until 4 months before he was admitted when she noticed a small lump in the left temporal region. The mass thereafter quickly grew larger. He was in good health with no history of recent systemic symptoms or relevant medical history. Physical examination showed a firm, fixed hard mass, 4.5 × 4cm in size in the temporal region with apparently normal overlying skin. She did not have cervical lymphadenopathy, the general, systemic and neurologic exams were normal, as were the results of her blood test. Facial cerebral magnetic resonance imaging (MRI) showed a left temporal extra axial tumor process invading the locoregional dura and responsible for hydrocephalus and sickle cell involvement. Cerebro-thoraco-abdomino-pelvic computed tomography (CT) also showed an extra-axial tumor process centered on the left temporal bone, becoming massively calcified in connection with his Ewing's sarcoma (Figure 1).
Figure 1: axial cut of a cervico-facial scanner showed an extra-axial tumor process centered on the left temporal bone, becoming massively calcified in relation to his Ewing sarcoma.

With the presence of 2 parenchymal pulmonary micronodules upper left lobe and lower left lobe without any abnormality. The patient also underwent a bone scan which returned normal without secondary bone localization. For radical tumor removal, the soft tumor was separated from the temporalis muscle along an overlying thin membrane that appeared to be the pericranium. The tumor was dissected from the surface of the temporal bone and removed along with the deep layer of the temporalis muscle. The outer surface of the temporal bone was slightly irregular, suggesting tumor invasion from the outside into the bone. Histopathologic findings were consistent with Ewing's sarcoma, the tumor was composed of small round cells which were strongly positive for CD99, focused positive for s-100, and negative for cytokeratin (AE1 / AE3), desmin, myogenin, LCA and TdT. The tumor was positive for the EWS translocation -FLI-1. The margins of the room are tumorous. The tumor was primarily an extracranial mass that was attached to the exterior surface of the temporal bone with bone reaction on the surface, and with local invasion into the temporal bone. For the surgical revision is not possible. At 24 days after the operation, chemotherapy (vincristine, doxorubicin, and cyclophosphamide alternating with ifosfamide and etoposide [VDC / IE]) was administered. With local treatment with radiotherapy was administered after the third cycle of chemotherapy, at the total dose of 55Gy in 25fractions of 2.2Gy per fraction (Figure 2a, 2b, 2c, 2d). She completed her adjuvant chemotherapy 9 months after the operation and is currently in complete remission with good local control.
DISCUSSION:

Between 1973 and 2004, the annual incidence of Ewing’s sarcoma in the United States was 2.93 cases per million (Esiashvili, N. et al., 2008). Ewing’s sarcoma is more common in the second decade of life and involvement of the temporal bone is very rare. Ernest et al., reported a case of Ewing’s sarcoma in a seven-month-old white girl with her right leg. The mass was not mobile and seemed to come definitely from the tibia and not from the overlying soft tissues. No metastatic lesions were noted in the lung fields. She deteriorated rapidly and expired six months after her symptoms first appeared. Ewing’s sarcoma has been localized between
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the pericranial and the temporal bone. The main imaging and histopathology findings were: First, CT scan of the skull showed a sunray appearance like a periosteal reaction, without the presence of a bone defect. Second, the MRI showed two membranous structures that surrounded the tumor and were connected to the pericranium because the tumor was mainly located outside the temporal bone. Ewing's sarcoma is most often diagnosed in the second decade of life and is the second most common form of primary bone cancer in children (Horowitz, M.E. et al., 1992). In about half of the cases, the tumors were located at the extremities of the patients (Horowitz, M.E. et al., 1992). In contrast, primary skull involvement occurred in only 1–2% of Ewing sarcomas (Amdt, C.A., & Crist, W.M. (1999 ; & Paulussen, M. et al., 2001). Intracranial extension was present in the majority of primary Ewing sarcomas; Extra cranial extension was observed in only 8 cases of primary Ewing sarcoma.

Periosteal reaction is a rare x-ray appearance in primary Ewing's sarcoma involving the bone of the head and neck (Umredkar, A. et al., 2012). Cortical thickening was observed in only 8% of cases, while changes were recognized in 54% of cases. In contrast, pure lytic changes (58%) and honeycomb (21%) are more common in these patients; Among other patients with primary Ewing sarcoma, pure lytic and alveolar changes occurred in 18.6% and 5.8% of cases respectively; bone expansion, which occurs in 25% of head and neck cases, and cortical violation, which occurs in 50% of cases, are also much more common than in the general population (Siegal, G. P. et al., 1987). Of the 8 previous cases of primary Ewing's sarcoma involved extracranial extension, osteolytic destruction or cortical violation was observed in 7 cases; the remainder was a congenital case in which Ewing's sarcoma occurred in the frontonasal region (Umredkar, A. et al., 2012; Atyiah, F., & Haddad, F. 1966; Bricha, M. et al., 2007 ; Hara, N. et al., 1990 ; JAYARAM, G. et al., 1986 ; Lee, R. J. et al., 1988 ; Moschovi, M. et al., 2011; & Naidu, M. R. C. 1989).

Although a periosteal reaction was observed in our case in the form of osteolytic lesions, bone expansion and cortical violation were not observed on the x-ray or CT scan of the skull. The CT scan showed a strongly marked tumor on the surface of the temporal bone. Periosteal Ewing's sarcoma is a rare form of sarcoma that particularly affects the femur (Huang, X., & Saint-Jeannet, J. P. 2004), it is difficult to compare our case with previous cases of periosteal Ewing's sarcoma affecting long bones because there are structural differences between long bones and membrane bones, for example, the thickness of compact bone. Because the preoperative images and operative results indicated that the tumor was mainly localized in the subperiocranial region, we created a new classification of “Pericranial Ewing's Sarcoma”, and our case was diagnosed as Primary Pericranial Ewing's sarcoma. There is a previous report of a case of primary Ewing sarcoma attached to the skull in which the tumor was completely extra cranial (Naidu, M. R. C. 1989).

Vaccani et al., reported 70 cases of Ewing sarcoma between 1986 and 1996, of which five cases had Ewing sarcoma of the head and neck (7.1%) and the age of presentation ranged from 7.5 to 14 years. An enlarged mass in the mandible is the most common presentation, in none of these was the temporal bone involved. Three out of five patients have died of metastatic disease and two are alive without evidence of disease (Huang, X., & Saint-Jeannet, J. P. 2004). Kuzeyli et al., reported primary Ewing sarcoma of the temporal bone in a 10-year-old girl. She presented with a palpable mass on the right frontotemporal region and proptosis of the right eye, but six months later she died from pulmonary metastases (Brieha, M. et al., 2007). The histopathological features of Ewing's sarcoma show monotonous sheets of round, monomorphic cells with scant cytoplasm, round nuclei and inconspicuous nucleoli. Molecular genetic analysis of chromosomes the t(11;22) (q24; q12) translocation is pathognomonic. 13 Overexpression of CD99, a transmembrane protein encoded by the MIC-2 gene, is another adjuvant.3,13 Rhabdomyosarcoma, which is high on the differential like sarcoma

The most common base of the skull in the pediatric population is also a small round cell which can be easily confused with Ewing's sarcoma. In either case, an initial biopsy of a tumor infiltrating skull base is usually the most appropriate intervention because these tumors usually respond quickly and dramatically to chemotherapy. Local control ensured by surgery or radiotherapy. Treatment options for Ewing's sarcoma include induction chemotherapy for local control and adjuvant chemotherapy. Chemotherapy, using multimodal therapy, dramatically increased survival from less than 10% to over 50% (Huang, X., & Saint-Jeannet, J. P. 2004). Commonly used chemotherapeutic agents include vincristine, doxorubicin, cyclophosphamide, etoposide, and ifosfamide (Ayvöré, I. L. et al., 2005; & Paulussen, M. et al., 2001). Before chemotherapy was started, most patients died from metastatic disease within 2 years. Local control options include surgical excision and radiation therapy. Irradiation is the primary local control modality when tumors are difficult to access or when surgical excision would lead to morbidities. Unlike patients with Ewing's sarcoma affecting the pelvis or lower body, head injury patients often undergo urgent surgical treatment due to elevated intracranial pressure and impending neurological deficits; therefore, neoadjuvant radiation therapy or chemotherapy is not commonly used. Tumors appearing from the calvarium may lend themselves to complete resection surgery. However, complete excision of basal skull lesions is often impossible without significant morbidity. Our case shows no sign of recurrence or metastasis after eighteen months of follow-up.
CONCLUSION:

This case report of a 13-year-old boy is in addition to five previously reported cases of this rare form of Ewing’s sarcoma affecting the temporal bone. When bone is affected, the tumor presents a unique management due to its intimate relationship with structures critical neurovascular cells in this region. Given its rarity to affect the skull, particularly the temporal bone, our findings add to our overall understanding of treatment options and outcomes for this uncommon subset of patients with Ewing’s sarcoma.

REFERENCE