Primary Ewing's Sarcoma of the Cranium: Case Report

Kafatasının Primer Ewing Sarkomu: Olgu Sunumu

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Abstract: Primary Ewing's sarcoma of the cranium is quite uncomon. Pre or peroperative recognition of this disease is important as it leads planning the surgical strategy which effects the prognosis. We present a case of primary Ewing's sarcoma originating from mastoid bone and invading petrous bone, occipital bone, brain, dura, as well as tentorium, transverse sinus, skin and cervical muscles. Radiological and pathological findings were demonstrated.

Key words: Cranium, Ewing's sarcoma, magnetic resonance imaging, skull.

Özet: Kranyumu tutan primer Ewing sarkomu nadirdir. Hastalığın ameliyat öncesinde veya ameliyat sırasında tanınması cerrahi stratejinin planlanması ve prognoz açısından önemlidir. Bu makalede mastoid kemikten kaynaklanarak petröz ve oksipital kemikler ile beyin, dura, tentoryum, transvers sinüs, cilt ve boyun kaslarını tutan primer Ewing sarkomu olgusu sunulmuştur. Radyolojik ve patolojik bulgular gösterilmiştir.

Anahtar kelimeler: Kranyum, Ewing sarkomu, magnetik rezonans görüntüleme, kafatası.

INTRODUCTION

Ewing's sarcoma is a malignant primary bone tumor primarily seen in the long bones (4). Primary Ewing's sarcoma of the cranium is quite uncommon occuring in 1% of cases (2,3). We report on a case of primary Ewing's sarcoma that originated in the mastoid bone. The tumor invaded the skull base, and also extended to the cervical region.

CASE REPORT

This 15-year-old girl presented with the complaint of a rapidly growing painful mass in the right retroauricular area, as well as nuchal pain, nausea, and vomiting. Physical examination revealed a firm, 5x5x3 cm tender mass over the right mastoid bone. Neurological examination showed that the patient was ataxic with an abnormal gait. She had

right-sided dysmetria, and nystagmus was observed on right lateral gaze. Magnetic resonance (MR) imaging revealed a right extraaxial soft tissue mass involving the temporal and occipital bones. The lesion was displacing the right cerebellar hemisphere towards the midline, and extended intracerebrally in a transtentorial route. (Figure 1 a,b)

Digital subtraction angiography demonstrated anterior-superior displacement of the right middle cerebral arteries. There was no tumor staining on angiography. Right transverse sinus was not opacified secondary to the mass effect. (Figure 2)

The patient was operated on and the tumor was subtotally removed, leaving a small remnant adherent to the torcular herophili that was infiltrating the petrous bone. Profuse bleeding started when the tumor was detached from the right transverse sinus, and this was stopped by suturing the sinus next to the torcular herophili.

The region of scalp that had been invaded by the tumor was also resected, along with a 1cm margin of normal skin. Plastic surgery helped during the closure of the scalp defect by a rotational skin flap.

Histopathological examination revealed that the tumor cells contained scant cytoplasm and their nuclei were uniformly round or oval. The findings on periodic-acid-Schiff (PAS) staining was negative, as were immunochemical studies for leucocyte common antigen (LCA), desmin, glial fibrillary acidic protein (GFAP), CD 3 and CD 20. Some of the tumor cells stained positive for vimentin antibody. Based on these results the pathological diagnosis was Ewing's sarcoma (Figure 3).

Radiological and scintigraphic examinations confirmed that there were no other sites of involvement. The patient was discharged with no neurological deficits for adjunctive radiotheraphy and chemotheraphy.

DISCUSSION

Ewing's sarcoma is the second most common bone tumor primarily seen in the long bones. This neoplasm is most frequently seen in children, with 75 % arising in patients under 20 years. The male-to-female ratio is 1.6:1. Ewing's sarcoma is rarely encountered in the skull, and only 1% of cases originate from the cranial bones.In documented cases, the sites of origin have included the frontal, parietal sphenoid and petrous bones (3).

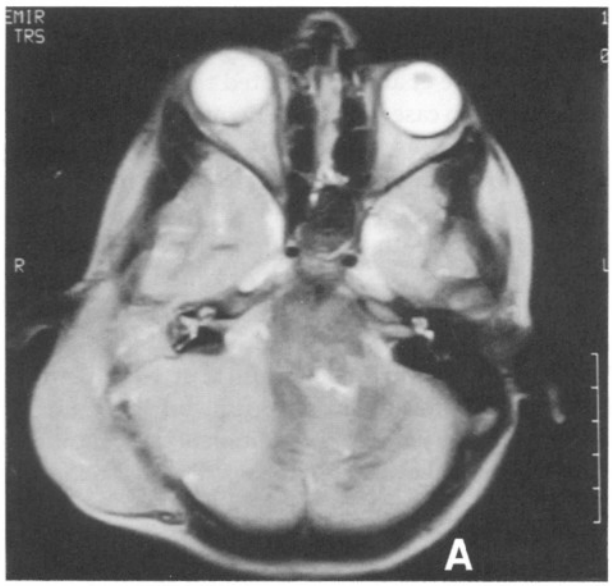

Figure 1a: A transverse T2-weighted MR image shows the high signal intensity of the extraaxial soft-tissue tumor, and reveals temporal and occipital bone involvement.

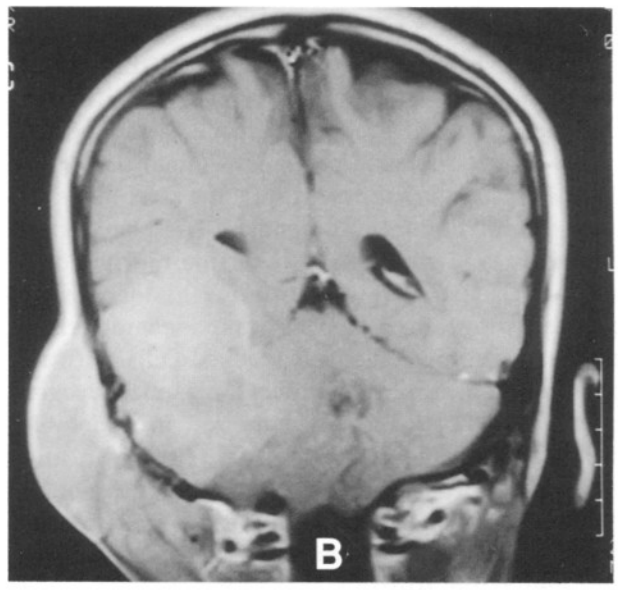

Figure 1b: A post-contrast coronal T1-weighted MR image demonstrates the enhancing extraaxial mass, which is producing a significant mass effect in the temporal and occipital lobes, and the right cerebellar hemisphere.

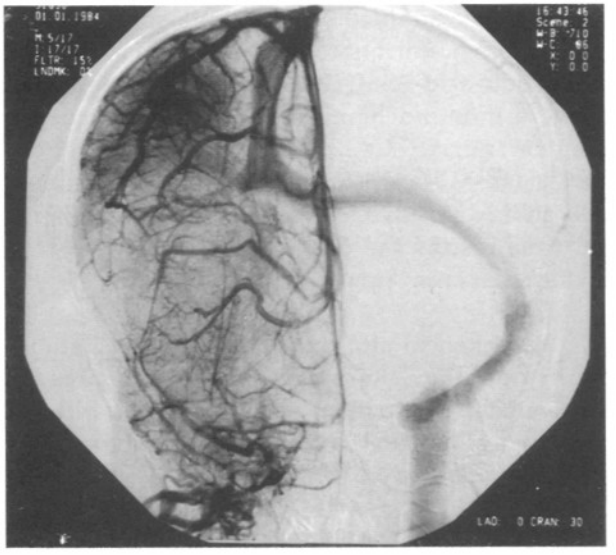

Figure 2: The subtraction print of a right internal carotid arteriogram demonstrates highly abnormal circulation with a large soft-tissue mass.

In published cases of primary Ewing's sarcoma of the cranium, the most common symptoms reported at the time of diagnosis have been local swelling and associated headache. Patients with the skull base as the primary site also showed proptosis and various types of cranial nerve palsy at initial presentation (3,5,9).

The list of clinical and radiological differential diagnoses for primary Ewing's sarcoma of the cranium includes osteosarcoma, lymphoma, meningioma, metastatic tumors, Langerhans' cell histiocytosis and rhabdomyosarcoma (7,8). Biopsy is essential for definitive diagnosis. The histological diagnosis is made by examining the morphology, chromatin structure and immunohistochemical profile of the tumor cells. This tumor is characterized by small, uniform, densely packed cells with no distinctive borders. Although cytoplasmic glycogen content was considered to be important in differential diagnosis, in specimens fixed in formalin and embedded in paraffin, the glycogen could not be demonstrated with PAS staining (6). In our case, positive immunohistochemical findings for vimentin and negative findings for desmin ruled out rhabdomyosarcoma, and negative results for of LCA, CD 3 and CD 20 ruled out malignant lymphoma.

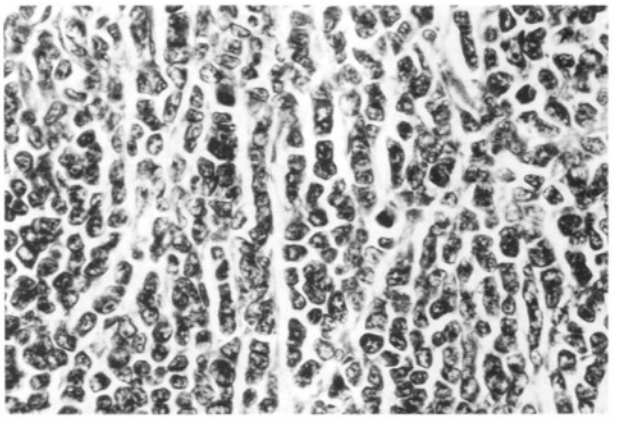

Figure 3: A slide shows the tumor tissue composed of uniform, small, round cells with mild mitotic activity (H&E x 230).

Early diagnosis and treatment prior to metastasis is essential for long-term survival in patients with Ewing's sarcoma (3). The disease is treated through a multidisciplinary approach that includes surgery, chemotherapy, and radiotherapy. When the tumor affects the cranial bone, surgical resection is particularly important. This has not only been shown to improve survival, but also may be needed to decompress vital neural structures, as was true in our case (3). Surgical resection also has been advocated to decrease the primary tumor mass in preparation for local radiation and chemotherapy (1,3).

Watanabe et al. documented that the survival rate is higher in cases of cranial Ewing's sarcoma than in patients with this neoplasm at other sites (9). Desai et. al reported high survival rates with the newer round cell chemotherapy (RCT)-II protocol (3). The main factor in treatment failure is the development of metastatic disease, the most common form being lung metastasis (1).

In conclusion, it is important to consider Ewing's sarcoma of the cranium both pre- and intraoperatively because recognition of this condition may lead to total excision which is reported to be an essential aspect for prognosis.

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The typical, but not diagnostic of Ewing's sarcoma, is the radiographic subperiosteal new bone formation creating "onion-skin layering" in long bones.