Skull Metastasis of Ewing's Sarcoma
—Three Case Reports—

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Abstract

Three cases of skull metastasis of Ewing's sarcoma were treated. The metastatic lesion was located at the midline of the skull above the superior sagittal sinus in all cases. Surgery was performed in two patients with solitary skull lesions involving short segments of the superior sagittal sinus without remarkable systemic metastasis, resulting in good outcome. The third patient had extensive, multiple tumors involving the superior sagittal sinus which could not be excised, and died due to intracranial hypertension. The surgical indication for skull metastasis of Ewing's sarcoma depends on the location and length of the involved superior sagittal sinus, and general condition.

Key words: Ewing's sarcoma, metastasis, midline, skull, superior sagittal sinus

Introduction

Ewing's sarcoma is an uncommon primary malignant bone neoplasm usually occurring in children and young adults, and most often originating in the long bones of the lower extremities.) Metastasis frequently spreads to the lungs and the skeletal system. However, the incidence of secondary central nervous system involvement has increased because of increased effectiveness of adjuvant chemotherapy and irradiation. Meningeal invasion or spinal cord compression are the best known forms of central nervous system involvement. The frequency of metastatic involvement of the skull is unclear. We present three cases of Ewing's sarcoma with midline skull metastasis.

Case Reports

Case 1: A 12-year-old female underwent resection of a Ewing's sarcoma in her mandibula followed by six courses of chemotherapy 1 year previously. She noticed parietal bulging 3 months before admission. Neurological examination revealed no findings. The parietal tumor was 4 cm in diameter, elastic hard with tenderness.

Computed tomography showed an isodense lesion, which was inhomogeneously enhanced by contrast medium, extending from the epidural space to the scalp in the midline of the parietal region with mild skull thinning (Fig. 1 left). Cerebral angiography showed compression of the superior sagittal sinus beneath the tumor (Fig. 1 right). Craniotomy disclosed the tumor involving the skull and adhering to the dura mater above the superior sagittal sinus. The tumor was totally extirpated except for the dural attachment because the superior sagittal sinus was patent. Histological examination of the surgical specimens showed that the neoplastic cells were small and round, and arranged in an alveolar pattern, partially forming rosettes. Periodic acid-Schiff (PAS)-positive granules were seen in the cells. The histology was consistent with Ewing's sarcoma, and similar to specimens from the initial mandibular lesion.

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of the frontal region (Fig. 2 left). Cerebral angiography showed a tumor stain fed by the superficial temporal and meningeal arteries and obstruction of the anterior part of the superior sagittal sinus beneath the tumor (Fig. 2 right).

The tumor was removed surgically with ligation and cutting of the superior sagittal sinus adhering to the tumor. Histological examination of the surgical specimens showed uniform small round tumor cells forming solid nests, involving the periosteum. PAS-positive material was found within the sparse cytoplasm. The histology was identical to the specimens from the right rib lesion. The diagnosis was skull metastasis of Ewing’s sarcoma.

Postoperative systemic chemotherapy was performed because bone scintigraphy detected a hot spot in his right scapula. Now he is healthy and no tumor recurrence has occurred in the 1-year postsurgery follow up.

Case 3: A 16-year-old male underwent surgery for a Ewing’s sarcoma in his left proximal femur in April 1996, followed by systemic chemotherapy. However, bone scintigraphy revealed metastases to multiple ribs and parietal skull. Paraplegia due to thoracic intraspinal metastasis and parietal bulging appeared in August 1996, so local irradiation was performed.

MR imaging revealed multiple metastases along the midline of the skull, extending from the frontal to parietal regions and involving the epidural space over the superior sagittal sinus and the scalp (Fig. 3).

Surgical intervention was not carried out because the skull metastases were too large to excise the skull of the frontal region (Fig. 2 left). Cerebral angiography showed a tumor stain fed by the superficial temporal and meningeal arteries and obstruction of the anterior part of the superior sagittal sinus beneath the tumor (Fig. 2 right).

The tumor was removed surgically with ligation and cutting of the superior sagittal sinus adhering to the tumor. Histological examination of the surgical specimens showed uniform small round tumor cells forming solid nests, involving the periosteum. PAS-positive material was found within the sparse cytoplasm. The histology was identical to the specimens from the right rib lesion. The diagnosis was skull metastasis of Ewing’s sarcoma.

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Case 2: A 17-year-old male underwent resection of a Ewing’s sarcoma in the right 10th rib 4 years previously, followed by chemotherapy (vincristine, Adriamycin, cyclophosphamide, and actinomycin D) per month for 1 year. He noticed frontal bulging 6 months before admission. Neurological examination revealed no remarkable findings. The tumor was 3 cm in diameter, elastic hard with tenderness.

Magnetic resonance (MR) imaging showed an isointense mass, which was irregularly enhanced by contrast medium, extending from the epidural space to the scalp through the skull defect in the midline of the frontal region (Fig. 2 left). Cerebral angiography showed a tumor stain fed by the superficial temporal and meningeal arteries and obstruction of the anterior part of the superior sagittal sinus beneath the tumor (Fig. 2 right).

The tumor was removed surgically with ligation and cutting of the superior sagittal sinus adhering to the tumor. Histological examination of the surgical specimens showed uniform small round tumor cells forming solid nests, involving the periosteum. PAS-positive material was found within the sparse cytoplasm. The histology was identical to the specimens from the right rib lesion. The diagnosis was skull metastasis of Ewing’s sarcoma.

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Ewing's sarcoma was too serious. He died on August 29, 1996, due to intracranial hypertension. Autopsy disclosed severe carcinomatous meningitis caused by Ewing's sarcoma from spinal invasion and multisystemic metastases including the midline of the skull and invading the superior sagittal sinus.

**Discussion**

Ewing's sarcoma is a highly malignant bone tumor, accounting for approximately 10% of all primary bone tumors. Metastasis occurs in up to 85% of patients within 2 years of recognition. The frequency of metastases to various organs is as follows: Lungs 85%, bones 69%, pleura 46%, lymph nodes 46%, dura and/or meninges 27%, and central nervous system 12%. The central nervous system may be involved in 10% to 35% of patients.

Skull metastasis of Ewing's sarcoma is not rare compared to primary Ewing's sarcoma of the skull, but the actual frequency is unknown. We treated six patients with Ewing's sarcoma from 1981 to 1996, and three patients presented here had skull metastasis.

The location of skull metastasis of Ewing's sarcoma is also not clear. All our three cases occurred on the midline of the skull, just above the superior sagittal sinus. Another metastatic deposit from a Ewing's sarcoma occluded the occipital superior sagittal sinus, but others of skull metastasis have also been described. The tendency to involve the midline may be because the midline of the skull is the watershed area of the external carotid artery, resulting in harboring of metastatic tumor cells, but this implies that metastatic skull tumors should frequently occur in the midline, but this is not so. Alternatively, dural metastasis may mimic skull metastasis. Arachnoid granules are distributed beside the sagittal sinus along the midline. Therefore, metastasis in the arachnoid granules would grow in the midline and involve the sinus and epidural space. Proof of this hypothesis requires direct evidence from further cases.

The outcome for patients with Ewing's sarcoma was poor, with 5-year survival of less than 10%. Since the introduction of aggressive chemotherapy and radiation, the 5-year survival rates have increased to 50%. Therefore, management of the metastatic lesions may be as important as that of the primary focus. Solitary skull metastasis should be removed totally, followed by systemic chemotherapy, resulting in the good outcome of Cases 1 and 2. The indication of surgical intervention for skull metastasis depends on the possibility of excision or preservation of the superior sagittal sinus. MR imaging and cerebral angiography provide useful information concerning dural sinus involvement by the tumor. If the tumor involves the longer segment of the superior sagittal sinus as in our Case 3, the tumor will be hard to remove without sinus reconstruction. Therefore, early detection of skull metastasis is essential, and if possible, aggressive treatment should be considered for the metastatic lesion, depending on other systemic metastases and the general condition.

**References**


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