I Introduction

Ewing’s sarcoma is the second most common primary bone tumor presenting in childhood and adolescence. Ewing’s sarcoma is a malignant small that belongs to a family of round-cell neuroectodermally derived tumors. It can occur at osseous and extraskeletal sites. Its usual locations are diaphysis of long bones followed by pelvis, ribs, vertebrae, and rarely skull. Skull lesions are seen in 6 to 9% of cases.

In this article, we report a rare case of Ewing’s Sarcoma of the occipital bone presenting with non-communicating hydrocephalus. The tumor extended to intraparenchymal, which is quite rare in patients with Ewing’s sarcoma. The case presentation, imaging findings, operative approach, and postoperative treatment plan are discussed, with a literature summary.

CASE PRESENTATION

13-month-old female patient presented to the emergency department of our Academic General Hospital with a chief complaint of gradually progressively increasing head circumference. It was reported in the last 2 months after the surgical excision of the occipital lump by a general surgeon in the district hospital.

Initially, the size of the head was normal, but there was a lump behind it. After surgery, the head circumference began to increase. A detailed history from her parents revealed a behavior change; the patient was more fussy and could not sleep well. There is a history of seizure once a week ago lasting longer than 5 minutes, comprising body tonic posturing. The seizure was provoked by fever. The patient had no previous history of vomiting, fever, unconsciousness, or focal neurological deficit. The mother had a normal pregnancy and delivery, and she did not use any medication.

The patient was consulted to our department by the head and neck surgeon with extraskeletal Ewing sarcoma of the occipital region with status epilepticus and hydrocephalus. Physical examination showed a head circumference of 62 cm. The patient’s major fontanelle was tender and opened. CT scan with contrast administration revealed a bone defect in the occipital region, mass of the occipital lobe, and non-communicating hydrocephalus, as seen in Figure 1. There was mixed density lesion in the posterior fossa after contrast administration demonstrated an intraaxial tumor of the posterior fossa. The patient underwent a ventricular peritoneal (VP) shunt to relieve the intracranial pressure. The patient had an uneventful postoperative course with no reported complications. The patient was planned for elective surgery for craniotomy tumor excision. However, during the disease, the patient’s condition worsened due to sepsis and eventually died.

DISCUSSION

Ewing’s sarcoma of the cranial bones is rare for about 6-9% of cases. These are grouped under the primitive neuroectodermal tumor (PNET) family by the WHO classification, which had small round blue cells.
Most of Ewing’s sarcoma occurs in the long bones, pelvis, ribs and flat bones but rarely may have an extraskeletal origin named extraskeletal Ewing sarcoma. Ewing’s sarcoma mostly infiltrates the frontal, parietal, temporal bones and skull base, with the most common involvement being the parieto-occipital region. The most common symptoms associated with calvarial Ewing’s Sarcoma tend to develop due to the dural invasion, hydrocephalus or increased intracranial pressure. HEADACHES AND SCALP SWELLING ARE THE MOST COMMON SYMPTOMS, AND PAPILLEDEMA IS THE MOST COMMON SIGN.

The distinction between Ewing’s sarcoma and central PNET has therapeutic and prognostic importance. They are poorly circumscribed and can occur in any region of CNS but rarely metastases elsewhere. Involvement of cerebrospinal fluid is reported in 10–30% of cases. Invasion of the dura mater constitutes 80% of cases with destruction of the bone. Involvement of brain tissue in Ewing’s sarcoma is rare and carries a worse prognosis for this infrequent tumor.

FIGURE 1. Pre-operative CT scan with contrast administration showing a contrast-enhancing occipital mass with bone defect extending to the fourth ventricle. The ventricles appear dilated, showing a non-communicating hydrocephalus.

Ewing’s sarcoma is treated with a multidisciplinary approach, including surgery, chemotherapy, and radiotherapy. Primary Ewing’s skull tumors are considered to carry a better prognosis. However, in this case, Ewing’s tumor had evaded intraparenchymal, which carries a worse prognosis for this patient.

Awareness of unusual presentations of Ewing’s Sarcoma of the cranium is important for early diagnosis of the tumor, and there is a high clinical suspicion for this infrequent tumor.

CONCLUSION

Primary cranial Ewing’s sarcoma of the cranium is a rare occurrence. It is considered in the differential diagnosis in children with a tumor involving the skull with destruction of the bone. Involvement of brain tissue in Ewing’s sarcoma is rarely found and carries a poor prognosis. A prompt diagnosis and treatment are essential for long-term survival in these patients with Ewing’s sarcoma.

CONFLICTS OF INTEREST

None declared. The authors have no financial or other interest that should be known to readers related to this document.

ETHICAL CONSIDERATION

This manuscript data is from the medical records of patients at RSUD. Dr. Soetomo and the patients have already given the “informed consent” form for this case report.

FUNDING

This study is not funded by any external institutions, nor does it have any sponsors.

AUTHOR CONTRIBUTIONS

Galan Budi Prasetya contributed to the study’s conceptualization, data collection, writing, and editing. Wihasto Suryaningtyas and Muhammad Arifin Parenrengi contributed to reviewing, editing, and finalizing the study manuscript.

ACKNOWLEDGMENT

The authors would like to thank all the staff of the Department of Neurosurgery, Faculty of Medicine, Universitas Airlangga. We would also like to thank all who have contributed to and are involved in the process of this report.

REFERENCES

CASE REPORT


This work is licensed under a Creative Commons Attribution