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Primary Malignant Lymphoma of the Cranial Vault with Extra- and Intracranial Extension

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Bone involvement is a common finding in many types of lymphomas, particularly in advanced stages. However, cranial vault affliction has been regarded as an exceedingly rare presentation. Here, we report the case of a patient with cranial vault lymphoma who presented with a scalp mass. An 81-year-old woman presented with a gradually growing and non-painful frontal scalp mass that she noticed one month before admission. It was a flatly elevated, round mass measuring about 6×4×4 cm. Computed tomography and magnetic resonance imaging of the brain revealed a contrast-enhancing intracranial extradural mass at the counter-location of the scalp mass. The superior sagittal sinus was involved at the tumor site. Cerebral angiography showed that the tumor feeding vessels originated from the bilateral external carotid arteries. An operation was performed and the tumors were removed together with the involved bone. The pathologic diagnosis was malignant diffuse large B-cell type lymphoma. The patient was transferred to the Hemato-Oncology department for chemotherapy. Primary lymphoma of the cranial vault with scalp mass is very rare but it should be considered in the differential diagnosis of scalp masses. Although the results of reported cases are variable, the combination of surgery, radiation, and chemotherapy appears to offer favorable outcomes.

Key Words Primary lymphoma; Cranial vault; Scalp mass.

INTRODUCTION

Non-Hodgkin lymphomas represent only 3% to 4% of all neoplasms in the general population. Primary lymphoma of the bone (PLB) is uncommon, accounting for only 7% of all malignant bone tumors and less than 1% of all non-Hodgkin's lymphomas, and the majority of them involve the pelvis or limb bones [1-3]. Primary cranial vault involvement is rarely reported [4]. We report a case of a cranial vault lymphoma that was found as a frontal scalp mass in an elderly woman.

CASE REPORT

An 81-year-old woman presented with a flatly elevated mid-frontoparietal scalp mass that she noticed one month prior to admission. She was generally healthy and had no remarkable medical history besides taking antihypertensive medication for 20 years. The scalp mass was non-tender, rubbery, hard, non-movable, and its dimensions were 6×4×4 cm.

The patient was completely free from neurological signs and demonstrated no lymphadenopathy, hepatosplenomegaly, or Waldeyer's ring swelling. She had no history of head injury. All of the routine laboratory studies returned with normal results. A plain skull X-ray showed an irregular inner cortical bone margin under the scalp mass, and non-enhanced computed tomography (CT) scans of the brain showed moderately well-defined borders and some osteolytic margins on the inner table of the frontal bone with a permeative extra- and intracranial isodense mass (Fig. 1). T1-weighted image (T1WI) and T2-weighted image (T2WI) magnetic resonance imaging (MRI) of the brain showed an iso- to low signal intensity mass which was well enhanced with gadolininum-Diethylenetriamine pentaacetic acid. MR spectroscopy showed a choline peak in the mass. Digital subtraction angiography showed feeding arteries originating from the bilateral superficial temporal arteries. The tumor also invaded the superior sagittal sinus (Fig. 2).

An operation was performed to remove the extra- and intracranial tumor and the involved bone. The tumor was a well-localized, grayish white, soft hypovascular mass that was easily dissected from the surrounding tissue. The dura was diffusely

involved but easily separated from the underlying brain without cerebral involvement. Histologic study showed diffusely invading round nuclear immature cells. Immunohistofluorescence staining showed CD20 (+), CD79a (+), and Bcl-2 (+), suggesting a diffuse large B-cell lymphoma. Ki-67 staining was positive in over 90% of nuclei (Fig. 3). Positron emission tomography-CT, chest and abdomen-pelvic CT scans were performed to rule out systemic involvement. The study revealed no systemic involvement except a hypermetabolic lesion in the thyroid, which was diagnosed as a benign follicular

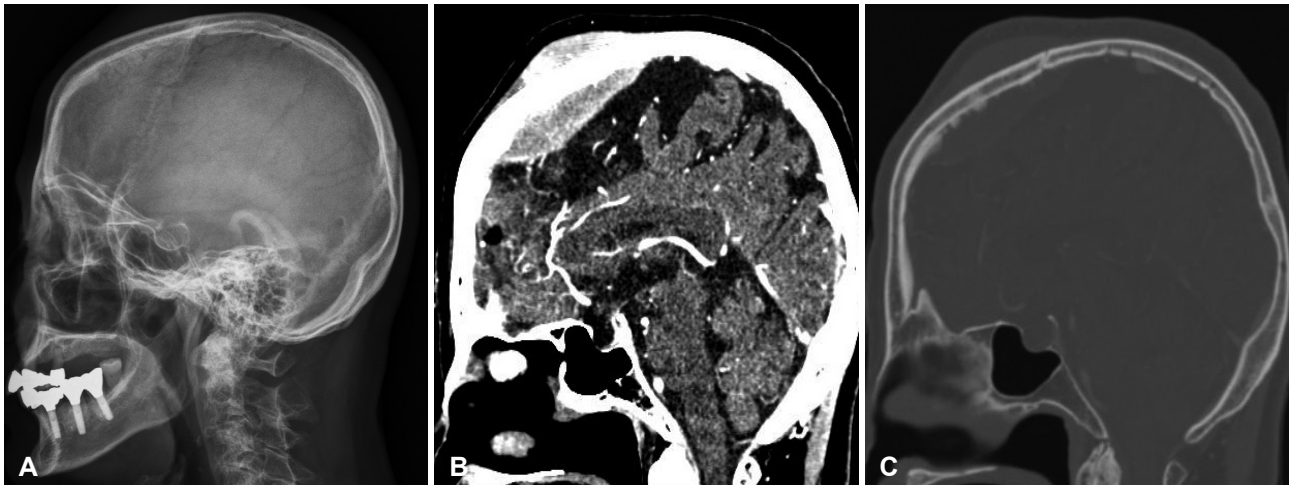

Fig. 1. Preoperative X-ray and computed tomography (CT) imaging. A: The skull X-ray shows an irregular inner cortical margin of the involved frontal bone. B: The sagittal view of the CT scan shows a permeative extra- and intracranial isodense mass. C: The bone window has moderately well-defined borders and shows some osteolysis of the inner table of the frontal bone.

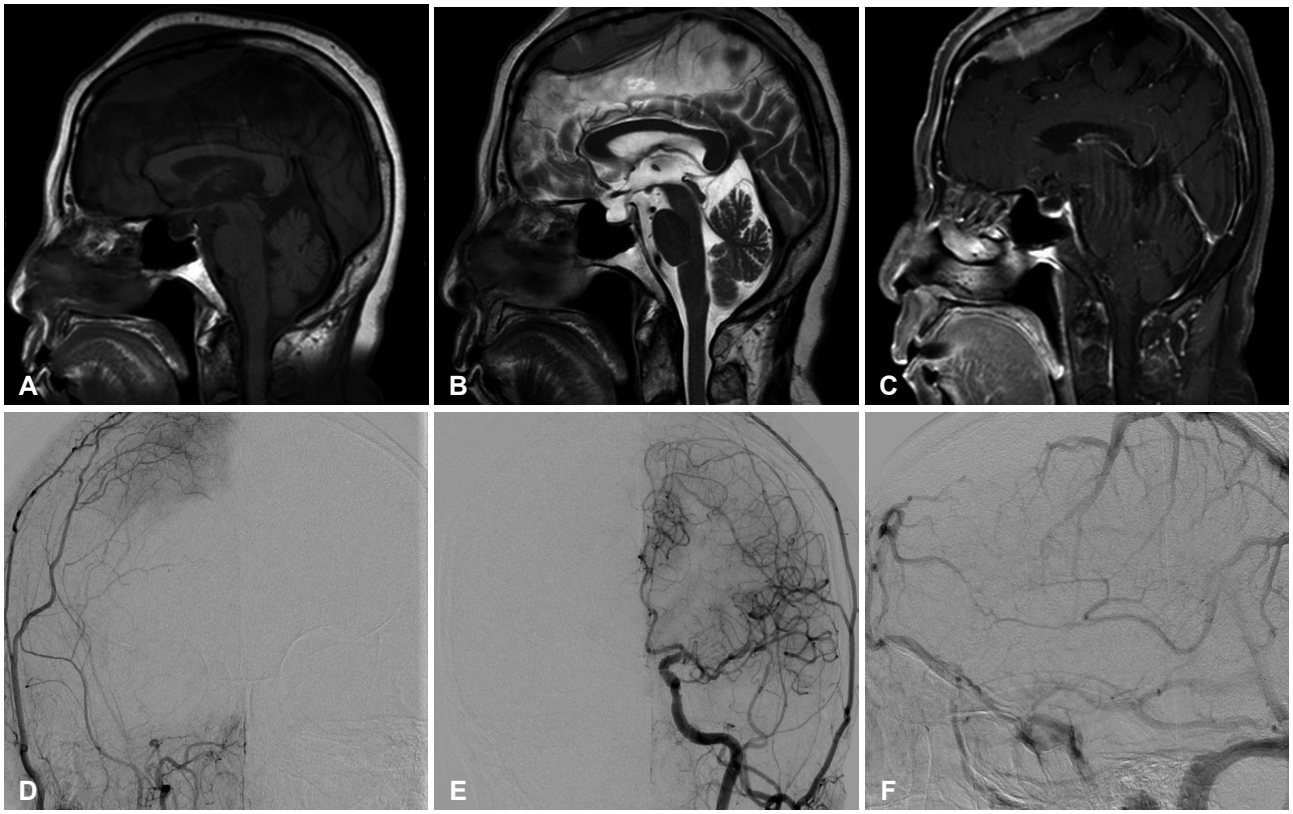

Fig. 2. Preoperative magnetic resonance imaging and digital subtraction angiography. A: The sagittal T1-weighted image shows an iso-signal intensity mass in the frontoparietal region. B: The sagittal T2-weighted image shows an iso-signal intensity mass in the same region. C: Diffuse contrast enhancement of the tumor and the dura. D: Feeding arteries from the right superficial temporal artery. E: Feeding arteries from the left superficial temporal artery. F: The superior sagittal sinus is occluded.

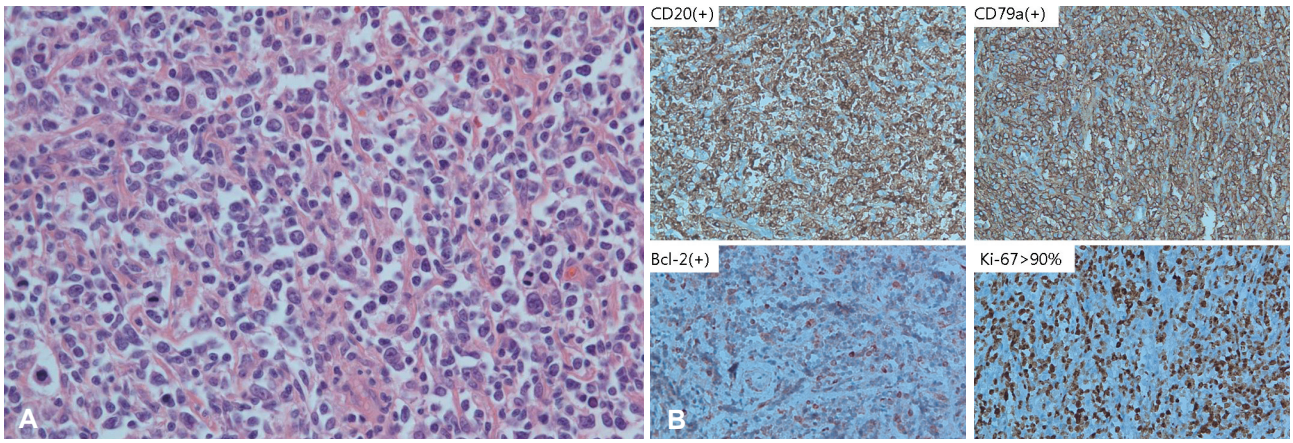

Fig. 3. Pathologic findings. The histologic features show diffusely invading round nuclear immature cells. A: Diffuse infiltration of round vesicular cells without an organoid pattern (HE stain, x100). B: The tumor cells stained strongly positively for CD20, CD79a, and Bcl-2. The proliferating index (Ki-67) was over 90% (immunohistochemical stain, x100).

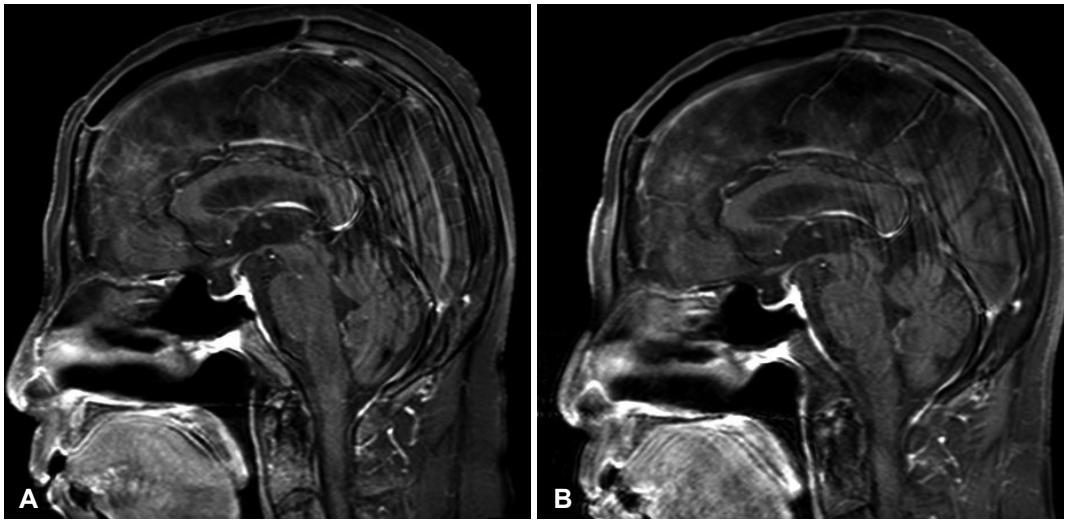

Fig. 4. Postoperative follow-up magnetic resonance imaging. A: The contrast-enhanced sagittal T1-weighted image taken 8 weeks after surgery shows good resection of the tumor. B: The contrast-enhanced sagittal T1-weigthed image taken 9 months after surgery shows that there is no tumor recurrence.

nodule.

The patient recovered well and was transferred to the Hemato-Oncology department, where she received three cycles of chemotherapy with CHOP-adriamycin, cyclophosphamide, vincristine, and prednisolone. Nine months after the operation, a follow-up MRI showed no tumor recurrence and the patient remained free from any noticeable symptoms (Fig. 4).

DISCUSSION

PLB is very rare and was first described by Oberling [5] in 1928. Parker and Jackson [6] classified PLB as a unique disease entity in 1939. The diagnostic criteria of PLB are 1) a primary focus in a single bone, 2) a positive histological diagnosis, 3) metastasis to only regional areas on presentation or the primary lesion preceding metastasis [2]. This patient had all of these diagnostic criteria. Causes of PLB have been suggested to be inflammation, trauma, or viral infections, but our patient had none of these [3].

The most common clinical manifestations of calvarial lymphomas are painless scalp masses [2,3,7-9], headaches [2,9], convulsions [1], or focal neurologic signs [1]. But these symptoms and signs are nonspecific, so further radiological and histological studies are needed for a conclusive diagnosis.

The MRI signal intensities are nonspecific and showed variable signal intensities on T1WI and T2WI, but most tumors show unenhanced isointensity and well-enhanced signal intensity [1,10]. Typical radiographic images show a large soft tissue component of the permeative growth or a moth-eaten pattern and less cortical destruction. However, this mimics the

appearance of other diseases such as Langerhan cell histiocytosis, osteomyelitis, leukemia, plasmacytoma, Ewing sarcoma, and metastatic cancer [1,7].

During the early stages of calvarial lymphoma, bony change is minimal but gradually extends outward and finally destroys the bone completely [4]. Our patient showed mild bony changes, suggesting that the lesion was in an early stage. The tumor grows rapidly and invades the dura and brain, so early diagnosis is important [4]. Chemotherapy and radiation therapy are the basic treatment modalities for primary calvarial lymphomas, but there are still no standard treatment protocols [4]. Surgery followed by local radiation is the preferred treatment for a single lesion of extranodal disease, but if the lesion is completely removed, radiological follow up without additional treatment may be another option [1]. Generally, surgery with chemotherapy and/or radiation therapy is recommended. In terms of chemotherapy, CHOP is often used [4]. If the patient's lymphoma has widespread dural involvement even after the tumor mass is removed, additional treatment may be provided by the Hemato-Oncology department.

Primary lymphoma of the cranial vault presenting as a scalp mass can easily be missed at the initial diagnosis because of its extreme rarity. However, because of its rapid growth and invasion to the brain, an early diagnosis and active treatment are very important. Although the results of reported cases are variable, the combination of surgery, radiation, and chemotherapy appears to offer favorable outcomes.

Conflicts of Interest

The authors have no financial conflicts of interest.

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