

Primary Cranial Vault Lymphoma: A Case Report

일차성 두개관 림프종: 증례 보고

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Cranial vault involvement in primary lymphoma is extremely rare in immunocompetent subjects. However, it should be considered as a differential diagnosis in the presence of a lesion involving all three compartments of the cranial vault, including the scalp, skull, and pachymeninges. We report a case of primary cranial vault lymphoma involving all three compartments of the cranial vault in an immunocompetent patient.

Index termsCranial Vault Tumor
Lymphoma

Received July 27, 2012; Accepted August 29, 2012

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INTRODUCTION

The incidence of central nervous system lymphoma has been increasing in both immunocompromised and immunocompetent subjects (1). Bony involvement is common in the advanced stage of many types of lymphoma (2). However, cranial vault involvement in primary lymphoma is rare.

CASE REPORT

A 72-year-old woman was admitted to the neurosurgery department for surgical excision of a palpable mass located at the right parietal region, which has persisted for over 1 year. A firm, non-pulsatile, non-tender mass attached to the skull with normal overlying skin was revealed. She showed no other signs or symptoms. Her medical history was unremarkable. She had no history of recent head trauma or human immunodeficiency virus. Neurologic and laboratory examinations were normal.

Anteroposterior view of the skull revealed a 4.3×3.4 cm-sized, ill-defined osteolytic lesion without a sclerotic rim (Fig. 1A). Brain and bone window images of the axial CT scan of the brain showed a 5.9×2.3 cm-sized, inhomogeneous intracranial and extracranial mass with mixed density, in addition to moth-

eaten bone destruction in the right parietal skull vault (Fig. 1B, C). Axial T1 weighted, T2 weighted and fluid attenuated inversion recovery images of the magnetic resonance imaging (MRI) of the brain showed a 5.6×2.3 cm-sized, relatively homogeneous isointense mass involving the scalp, skull, and pachymeninges at the right parietal region. Adjacent brain parenchyma showed normal appearance. The lesion also showed restricted diffusion on diffusion weighted images. After administration of gadolinium diethylenetriamine pentaacetic acid, the mass showed inhomogeneous enhancement (Fig. 1D-H). Therefore, the highly cellular mass was believed to originate from the diploic space and then extend to the inside and outside of the cranial vault.

The patient underwent gross total mass removal. Histopathology revealed diffuse proliferation of cells with loss of cohesiveness, suggesting hematologic malignancy or poorly differentiated carcinoma on hematoxylin-eosin stain (Fig. 1I). Immunohistochemical stains were positive for leukocyte common antigen (CD45), CD20 (Fig. 1J), Bcl-2, Bcl-6 and vimentin but were negative for CD10, CD30, CD56, epithelial membrane antigen, and cytokeratin. Proliferation marker Ki-67 was more than 80%. A pathological diagnosis was made of diffuse large B-cell lymphoma in the dura and skull vault.

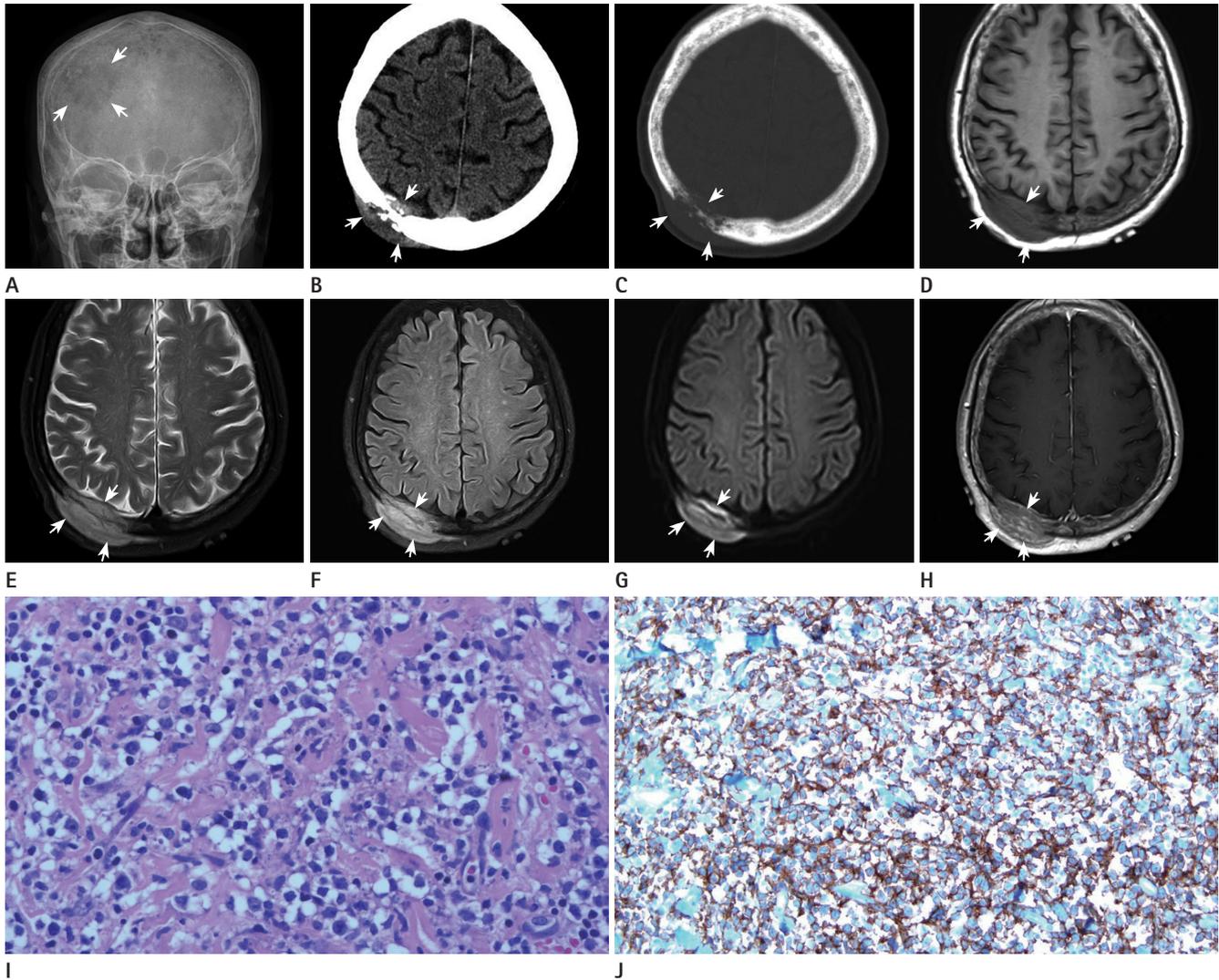


Fig. 1. A 72-year-old woman with a palpable skull mass.
A. Anteroposterior view of the skull reveals a 4.3 x 3.4 cm ill-defined osteolytic lesion without sclerotic rim (arrows).
B, C. Axial CT scan of the brain with brain (**B**) and bone window (**C**) images shows a 5.9 x 2.3 cm inhomogeneous intracranial and extracranial mass with mixed density, in addition to moth-eaten bone destruction in the right parietal skull vault (arrows).
D-H. Axial T1 weighted (**D**), T2 weighted (**E**) and FLAIR images (**F**) of the MRI of the brain show a 5.6 x 2.3 cm relatively homogeneous isointense mass involving the scalp, skull and pachymeninges at the right parietal region (arrows). Adjacent brain parenchyma shows normal appearance. The lesion also shows restricted diffusion on DWI (**G**) and inhomogeneous enhancement (**H**) on contrast enhanced image (arrows).
I, J. Photomicrographs of the surgical specimen show diffuse proliferation of cells with loss of cohesiveness in the hematoxylin-eosin stain (**I**, x 100) and strong staining of tumor cells for CD20 (**J**, x 100).
 Note.—DWI = diffusion-weighted imaging, FLAIR = fluid attenuated inversion recovery

CT scans of the abdomen and chest, and whole-body fluorodeoxyglucose positron emission tomography scans were negative for lymphadenopathy or hepatosplenomegaly. Bone marrow aspiration and biopsy from the posterior iliac crest revealed no involvement of malignant lymphoma.

Our case shows primary large B-cell lymphoma of the cranial vault without systemic involvement in an immunocompetent patient. The patient was treated with chemotherapy consisting

of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone. After 6 months follow-up, the patient is currently neurologically intact and shows no evidence of systemic dissemination.

DISCUSSION

Bony involvement is commonly seen with secondary non-

Hodgkin's lymphoma. However, the primary non-Hodgkin's lymphoma originating from bone is uncommon, comprising less than 1% of all non-Hodgkin's lymphomas (3); nonetheless, it is more common in patients with acquired immunodeficiency syndrome or traumatic head injury. A true primary malignant lymphoma of the bone is defined as a solitary mass lesion without any evidence of disease at other sites and no systemic dissemination found within 6 months of detection of the tumor (4). In addition, a primary lymphoma originating from the cranial vault of immunocompetent subjects is extremely rare.

According to most of the cases involving primary cranial vault lymphoma (PCVL), the disease occurs in all ages, but is much more common in older patients. PCVL shows no significant gender difference. The most common subtype of PCVL is a diffuse large B-cell lymphoma (5).

The initial symptoms and signs of lymphoma in the skull include painless scalp lump, headache due to bone destruction or tumor infiltration of meninges, seizures and focal neurological deficits resulting from infiltration of the cortex (6).

The most prominent characteristic of PCVL is scalp mass extending either intra- or extracranially, affecting all compartments of the cranial vault including the scalp, skull and pachymeninges (7). This review points to an identical imaging pattern in most of the lesions for all reported cases of PCVL, despite their different histological types (5). It is pathologically suggested that lymphoma cells infiltrate within the diploic spaces and extend along the emissary veins to infiltrate the soft tissues on either side of the bone (8). However, Ochiai et al. (9) reported a case of primary lymphoma of the dura mater and scalp without intervening skull bone invasion. In that report, they suggested it is attributable to the multi-centric nature of the malignant lymphoma. In some cases, destruction of the bone may not be seen because of the characteristic permeating growth pattern of lymphoma with a large soft tissue component and very little bone destruction (8).

The density on CT scans and signal intensity on MR images are nonspecific; the majority of reported cases of skull lymphomas have noted hyperdensity on unenhanced CT scans, isosignal intensity on unenhanced MRI and marked enhancement on postcontrast images. Subcortical edema and an indistinguishable border between brain and meninges in MRI, associated with focal neurologic signs or seizures, are predictive of

brain invasion (5).

The appearance of PCVL may mimic other differential diagnoses, such as metastatic carcinoma, osteomyelitis, meningioma, subdural hematoma, tuberculoma, Ewing's sarcoma and multiple myeloma (6). Carcinoma metastasis to the skull may show osteolytic bone lesions with extradural or intraparenchymal spread. Patients with osteomyelitis have lytic bone lesions and soft-tissue masses; however, the systemic signs are usually seen when soft tissue mass develops. Meningiomas usually come with associated hyperostosis, calcification, subdural extension, more prominent enhancement with dural tail sign and invasion of the superior sagittal sinus (6). Subdural hematoma may show variable and shifting signal intensity correlating with age of blood, with no abnormal marrow signal intensity. Tuberculoma may show a bony destruction often accompanying soft tissue opacity and sinus formation. Ewing's sarcoma occurs only rarely in the skull vault and patients are often much younger than those with PCVL (8). It is difficult to diagnose multiple myeloma with CT or MRI; laboratory data and pathologic features are therefore important in diagnosing this disease (10).

The optimum management of PCVL remains uncertain; however, surgical removal, followed by radiotherapy and chemotherapy can be recommended (1).

In conclusion, we report a surgically and pathologically proven case of PCVL. Radiologists should consider this PCVL as a differential diagnosis of a lesion involving all three compartments of the cranial vault including the scalp, skull, and pachymeninges.

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일차성 두개관 림프종: 증례 보고

윤소희 · 김명순 · 김영주

정상 면역인에서 일차성 두개관 림프종은 극히 드물다. 그러나 두피, 두개골, 경뇌막으로 이루어진 두개관의 세 층을 모두 침범한 병변이 있을 때 이 질환을 감별해야 한다. 저자들은 정상 면역 환자에서 두개관의 세 층을 모두 침범한 일차성 두개관 림프종을 경험하였기에 이를 보고하고자 한다.

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