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Case Report

High-Grade Diffuse Large B-Cell Lymphoma of the Dura with Skull and Scalp Involvement with Simultaneous Sternum Involvement

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Abstract

Diffuse large B-cell lymphoma (DBCL) is the largest subgroup of non-Hodgkin's lymphomas. Primary dural lymphoma, primary skull vault lymphoma, and primary sternum lymphoma are the rare lymphomas. We present the case of a 69-year-old patient with scalp, skull, and dura involvement that accompanying sternum involvement. It should be kept in mind that in the differential diagnosis of high-grade diffuse large B-cell lymphoma in patients presenting with a mass in the skull or sternum.

Keywords: *B-cell lymphoma, non-Hodgkin's lymphoma, skull, sternum*

Introduction

Diffuse large B-cell lymphoma (DBCL) is the largest subgroup of non-Hodgkin's lymphomas (NHLs). The incidence is approximately 4–17/100,000/year.[1] It is a little more common in males, and the median age is around 60 (14–98) years. The incidence of DBCL with a single extranodal involvement is approximately 71%, whereas multiple extranodal involvements are 28%–29%. Patients with the extranodal disease are usually older and have lower performance status.[2] Primary dural lymphoma constitutes approximately only for 0.1% of all NHL.[3] Primary skull vault lymphoma is another rare entity and accounts for only 0.2% of lymphoma cases.[4] Primary sternum involvement is very rare in DBCL like dural and skull vault involvement.[5]

We present the case of a 69-year-old patient with scalp, skull, and dura involvement that accompanying sternum involvement.

Case Report

A 69-year-old female patient presented to the emergency department with a severe headache and painful swelling in the right temporoparietal area. She also had a painless lump on her sternum. The lesion in the scalp had been growing during the past 4 months, and the lesion on the sternum had been present for the past 2 months. She denied any head or thorax trauma history. Her medical history was unremarkable except for warfarin therapy due to atrial fibrillation and hypertension. Her neurological examination was intact, and the infection markers were negative. The patient was referred to the neurosurgery outpatient clinic. Magnetic resonance imaging (MRI) revealed a lesion in the right temporoparietal area which caused local bone erosion. Together with the bone erosion, dural and scalp involvement were seen [Figure 1].

Thorax and abdominal computed tomography was planned for a probable primary lesion, but there was not a suggestive focus except bone erosion on sternum just beneath the painless lump. Surgery was performed for diagnostic purposes for the skull pathology. The pathologic bone part was removed following the scalp incision. The dural area was also removed because the dura was also affected, and then, duraplasty was performed. The removed lesion was sent for histopathological examination. Since the diagnosis was not definite during the surgery, cranioplasty was planned after the confirmation of the diagnosis and treatment strategy. The pathology showed widespread strong positivity with CD20, bcl-2, bcl-6, and CD10, moderate positivity on the ground with CD5 and CD3 observed

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with a small number of T-lymphocytes. It was negative for CD56, thyroid transcription factor-1 and pancytokeratin were negative, and Ki-67 proliferation index was around 80% [Figure 2]. The specimen was diagnosed with high-grade diffuse large B-cell lymphoma [Figure 3]. The cerebrospinal fluid sample was not taken from the patient. Since the patient had no previous history of lymphoma, the patient was not considered bone marrow involvement before the histopathologic results. The patient was referred to a center with the oncology department. To the best of our knowledge, the patient is still under the treatment of the referenced oncology clinic. . The patient consent form was obtained.

Discussion

The primary sites for extranodal DBCL include the stomach (22.4%), intestine (16.0%), nose and sinuses (8.9%), testis (8.4%), skin (7.9%), thyroid (6.9%), central nervous system (6.4%), breast (5.7%), bone (3.4%), salivary gland (2.7%), oral cavity, kidney, lung, orbit, and other miscellaneous organs, respectively.[6] The authors reported the incidence of 2 or more sites in extranodal DBCL as 28.1% at admission.[6]

Primary bone involvement in extranodal-DBCL is seen in about 3.5% of patients. This involvement generally affects the femur, tibia, pelvis, spine, mandible, and scapula. To diagnose true primary malignant bone lymphoma, the solitary mass lesion should be free of any trace of associated disease and systemic dissemination during 6 months following the diagnosis.[4] However, our patient had dura and scalp involvements in addition to bone involvement. Therefore, our case cannot be classified as a true primary malignant bone lymphoma.

Immunocompromised or trauma patients constitute the typical population of primary vault lymphomas.[7] However, primary NHL of the skull is rarely seen in these specialized populations. Our patient did not have an immunocompromised condition and a history of previous head trauma.

The expected admission symptoms for lymphomas of the skull are painless scalp lump, headache due to bone destruction or tumor infiltration of meninges, seizures, and neurologic deficits due to cerebral cortex involvement. One of the suggested progression mechanisms for lymphomas of the skull is infiltration of the spaces within the diploe and extension through the veins and soft tissues and covering the bone all over. On the other hand, the dura has been considered as a durable against lymphoma infiltration of the cerebral cortex.[4] The presenting symptoms of our case were painful swelling and diffuse headache in the right parietal-temporal region. She had no other symptoms such as focal neurological deficit or seizure with an intact neurological examination. The skull lesions of NHL expected to appear as isointense or hypointense in both T1- and T2-weighted imaging on MRI.[8] We also observed similar features in our patient's images. During the surgery, the periosteum, scalp, and dura involvements were

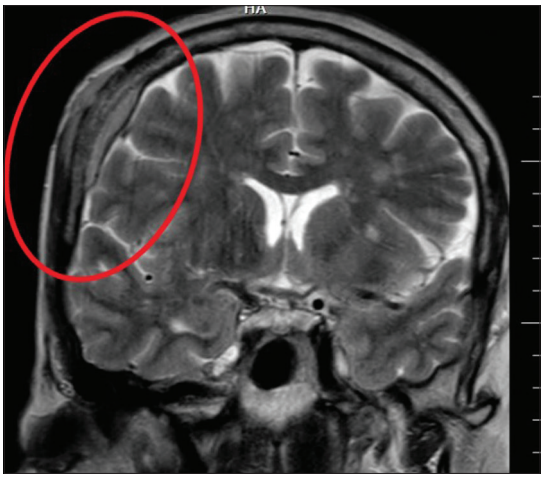

Figure 1: The cranial magnetic resonance imaging of the patient showing a lesion in the right temporoparietal area with bone erosion

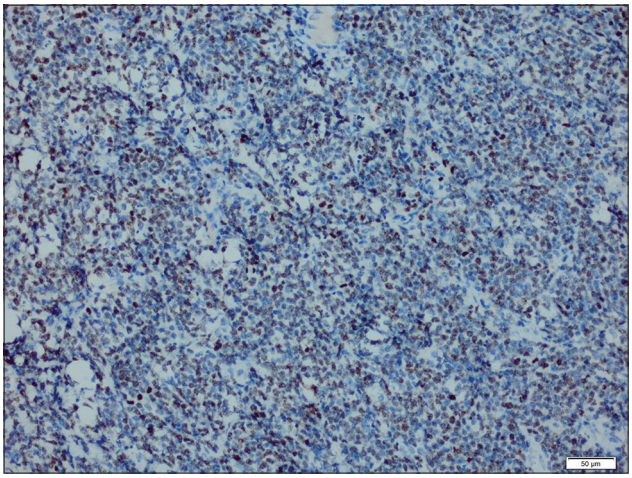

Figure 2: The pathological specimen showing Ki-67 proliferation index was around 80%

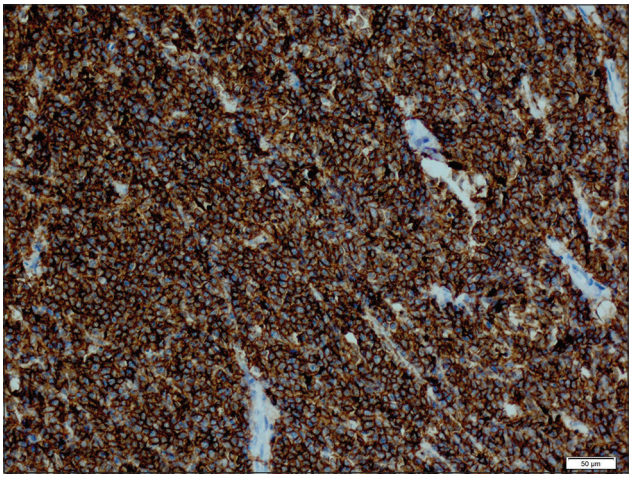

Figure 3: The pathological specimen showing widespread strong positivity with CD20

observed. The dura, which was thought to be invaded, was removed during the operation, and the pathological result revealed the presence of invasion as well in the dura.

There is no established treatment approach for malignant lymphoma of the skull vault. However, radiotherapy and chemotherapy after surgery have been advised.[9] After the cranial decompression, the present case was referred to the oncology clinic.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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