Primary Multifocal Skull Vault Lymphoma in an Acquired Immunodeficiency Syndrome Patient

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ABSTRACT

Primary skull vault non-Hodgkin's lymphoma with extra- and intra-cranial soft tissue components in an immunocompromised patient is extremely rare. There are only four cases reported in literature till date and in none were the lesions multifocal or had a discrete intraorbital component. We report such a case of multifocal primary skull vault lymphoma successfully managed in an acquired immunodeficiency syndrome patient. Distinguishing primary bone lymphoma from other skull vault lesions is important because of different management protocols.

Key words: Acquired immunodeficiency syndrome; human immunodeficiency virus; osteolytic skull lesions; skull lymphoma

Introduction

Non-Hodgkin's lymphoma (NHL) is one of the most common tumors and acquired immunodeficiency syndrome (AIDS) defining conditions in human immunodeficiency virus (HIV) positive patients.[1] About 20% of AIDS patients with multisystemic NHL have disease extension to the musculoskeletal system.[1] The presence of primary bone NHL in the absence of extraskeletal disease has also been reported in AIDS patients, affecting the lower extremities, spine, pelvis, and skull.[1] To the best of our knowledge and belief, among the four reported cases of primary NHL of the skull vault in AIDS patients, none of the cases has showed multifocal skull vault lesions and discrete orbital wall involvement. We describe such a case of an AIDS patient who was successfully managed by combined chemotherapy and radiotherapy.

Case Report

A 35-year-old married male patient presented to our Dermatology Department with gradually progressive, painless bilateral scalp swellings for last 1-month. Patient had a history of heterosexual contact with multiple sexual partners. Physical examination revealed firm, nontender, nonpulsatile scalp swellings with normal overlying skin. Laboratory findings showed a CD4+ cell count of 185/μl (normal: >400/μl) and positive HIV serology. Lumber puncture cytology was normal. Lateral radiograph of the skull revealed multiple variable sized geographical osteolytic lesions. Noncontrast computed tomography of the head [Figure 1] showed multifocal osteolytic skull vault lesions involving both inner and outer table of the skull with hyperdense extracranial as well as intracranial soft tissue components. A similar osteolyticlesion involving left zygomatic bone with extraconal intraorbital and infratemporal soft tissue components was also seen. Contrast-enhanced computed tomography (CECT) of the head showed enhancement of the soft tissue components [Figure 2]. Brain parenchyma appeared normal without any abnormal meningeal enhancement. Based on imaging and clinical history of HIV infection, diagnostic possibilities of aggressive lymphoma, metastasis and osteomyelitis were considered. CECT chest and abdomen were normal. Ultrasound neck was also normal. Aspiration cytology and biopsy of one of the swellings were done, and final diagnosis of diffuse large B cell lymphoma of immunoblastic variety was made based on cell morphology [Figure 3]. Immunohistochemistry revealed numerous CD20 positive cells. Bone marrow biopsy and peripheral smear were normal. The total leukocyte and platelets count were within normal range. Bone scan revealed

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	DOI: 10.4103/1115-3474.155746

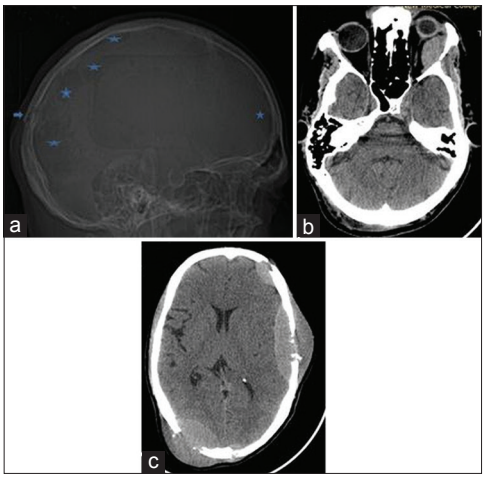

Figure 1: Noncontrast computed tomography head: A 35-year-old human immunodeficiency virus positive man with multiple osteolytic lesions (star) with focal cortical beach (arrow) in scanogram (a), hyperdense soft tissue component in left extraconal intraorbital region with lateral rectus muscle infiltration (b) and osteolytic lesions with intracranial and extracranial soft tissue components (c) in axial images

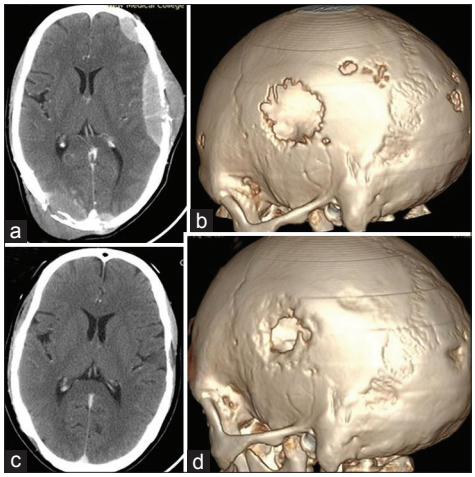

Figure 2: Contrast-enhanced computed tomography head: Pretreatment (a and b) and posttreatment (c and d) follow-up scans of the patient in brain window (a and c) and volume rendered images (b and d) at comparative levels showing healing of the skull vault lesions after the combined chemotherapy and radiotherapy

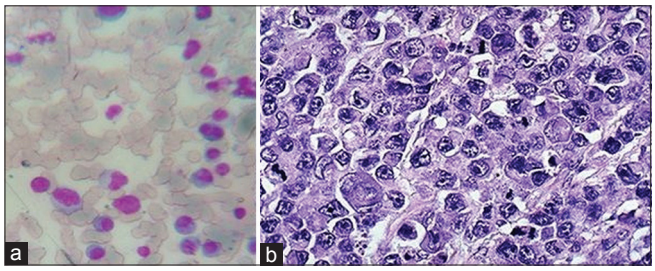

Figure 3: Aspiration cytology smear (a) and biopsy smear (b) showing large lymphoid cells with hyperchromatic nuclei (a - May Grunwald Giemsa, ×40; b - H and E, ×40)

multifocal increased radiotracer uptake in skull vault and left orbital regions. Patient was put on highly active antiretroviral therapy concurrent with cyclophosphamide, doxorubicin, vincristine, and prednisone regimen followed by cranial radiotherapy. After four cycles of chemotherapy follow-up, CECT scan of the head [Figure 2] showed healing of smaller osteolytic lesions and decreased size of the larger lesions with reduction in size of soft tissue component. Follow-up chest computed tomography (CT) and abdominal ultrasound were normal.

Discussion

After Kaposi's sarcoma, NHL is the second most common type of tumor in patients with HIV infection and it is one of the AIDS-defining illnesses occurring in the late stages of HIV infection with CD4 counts below 200 cells/μl.[1].Compared to the general population, HIV-positive patients have a 60 times increased incidence of NHL, majority of which are diffuse large B cell lymphomas.[1]

Secondary bone involvement is common in NHL, but primary osseous lymphoma is seen only in 3–4% of lymphoma patients with femur, tibia and pelvis being the most commonly affected sites.[2] Primary lymphomatous involvement of the cranial vault is rare, and most of the reported cases in literature are immunocompetent or of unknown immunological status.[3] Only a handful of cases is reported in AIDS patients.[3,4] Among these, only a single case showed multifocal osteolytic lesions with soft tissue component, and none of these case showed a discrete intraorbital component, a finding that was seen in our case. Primary bone lymphoma is classically defined as lymphoma presenting in an osseous site with no evidence of disease elsewhere for at least 6 months. Primary multifocal osseous lymphoma, a distinct subset of primary bone lymphoma, is defined as having lesions in multiple bones without lymph node or visceral involvement for 6 months after initial clinical diagnosis.[5] Most primary multifocal osseous lymphoma are described around knee joints with or without skull involvement.[6] In our patient, multifocal radiotracer uptake was confined only around the skull.

Most reported primary skull vault lymphoma in literature are solitary bone lesions with a mean age of the patient 60 years.[3] The symptoms and signs of skull lymphoma vary from a painless scalp lump to headache, seizures and focal neurologic deficits depending on the degree of bony destruction and brain infiltration.[4] Bone involvement can be in the form of a single osteolytic focus or multifocal/diffuse involvement with soft tissue component.[4,7] Characteristically, malignant lymphoma shows permeative growth pattern with predominant soft tissue component and little cortical destruction.[2]

The soft tissue component of skull lymphoma usually appears hyperdense compared to the cerebral cortex on plain CT and shows a strong postcontrast enhancement with or without underlying bone defect. Skull lymphoma with hyperostosis

mimicking meningioma has also been reported.[3,4] On magnetic resonance imaging, lymphoma appears iso to hypointense on T1-weighted and iso to hyperintense on T2-weighted sequences with a homogenous postcontrast enhancement.[3]

Metastasis is the most common malignant scalp lump[2] and it often presents with multifocal skull vault osteolytic lesions with soft tissue component, however, metastatic workup in our patient was negative. Another differential of multifocal osteolytic lesions of the skull vault especially in elderly people is multiple myeloma that shows lesions with well-defined margins resembling punch holes.[8]

Infective lesions like tuberculosis can cause calvarial osteolytic lesions with soft tissue, however, systemic symptoms or imaging manifestations often coexist.[4,8] Furthermore, a positive Mantoux test and raised erythrocyte sedimentation rate may give diagnostic clue for tuberculosis in up to 90% cases.[8]

Primary cutaneous follicular B-cell lymphoma (PCL) also appears to be a close differential as it mostly involves the head and neck region, however, compared to primary skull vault lymphoma, PCL shows an indolent course, less osteolysis, less soft tissue component and uncommon intra-cranial (extradural)/intraorbital extension.[9]

In HIV positive patients, Kaposi's sarcoma and bacillary angiomatosis can rarely involve skull vault, however, cutaneous lesions often coexist and in case of Kaposi's sarcoma a positive thallium scan in conjunction with a negative gallium scan is seen.[10]

A combined modality treatment with chemotherapy and radiotherapy is often used for localized bone lymphoma.[3] Most of the earlier reported cases of AIDS with primary skull vault lymphoma had an unfavorable prognosis.[4] Our patient showed a favorable response to the combined therapy and is currently alive 1-year after the initial diagnosis.

Conclusion

Although rare, lymphoma should be kept as an important differential diagnosis for multifocal osteolytic skull vault lesions, especially in middle-aged and elderly HIV positive patients.

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How to cite this article: Choudhary P, Kalwaniya S, Agarwal L, Saxena S. Primary multifocal skull vault lymphoma in an acquired immunodeficiency syndrome patient. West Afr J Radiol 2015;22:107-9.

Source of Support: Nil, **Conflict of Interest:** None declared.