

MULTIFOCAL PRIMARY LYMPHOMA OF THE CRANIAL VAULT IN A NON-IMMUNOCOMPROMISED ADOLESCENT

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Primary non-Hodgkin lymphoma (NHL) of the skull is extremely rare. The authors report a case in a 19-year-old boy who presented with a progressively increasing scalp mass on background of generalized headache of 6-months duration. Imaging showed multifocal skull vault lesion with intra- and extra-cranial soft tissue masses, causing permeative destruction of underlying skull bone. Further investigation failed to identify any other evidence of systemic lymphoma. Histopathology examination of superficial scalp mass showed low grade non-Hodgkin B cell lymphoma. The precise anatomical location and multifocal nature of the neoplasm occurring in young nonimmunocompromised individual makes this case clinically and radiologically unique.

Key-word: Primary cranial vault – Non-Hodgkin lymphoma – Nonimmunocompromised.

Non Hodgkin lymphoma (NHL) is an uncommon neoplasm with an incidence of only 3% to 4% in general population. It occurs more frequently in patients with acquired immunodeficiency syndrome (AIDS). Primary involvement of the central nervous system (CNS) occurs only in 1% to 2% of patients with lymphoma (1). NHL originating primarily in bones is rare, occurring only in 3-4% of patients with lymphomas (1), with a predilection for the long bones of the upper and lower extremities, the pelvis and the spine. NHL presenting with primary involvement of cranial vault and dura mater is an unusual event and have rarely been reported in literature. Out of 16 cases reported, 7 cases were associated with systemic disease on presentation (2). In the other 9 cases, the disease was restricted to the cranial vault and these cases were therefore described as primary cranial vault lymphomas. Only 3 out of 9 cases were non-HIV positive. We report an unusual case of primary cranial vault lymphoma with multifocal involvement that occurs in a nonimmunocompromised adolescent boy.

Case report

A 19-year-old boy presented with generalized headache and progressively increasing swellings on the left side of scalp which had been developing over a period of 6-months. His medical history was otherwise unremarkable. There was no history of head trauma.

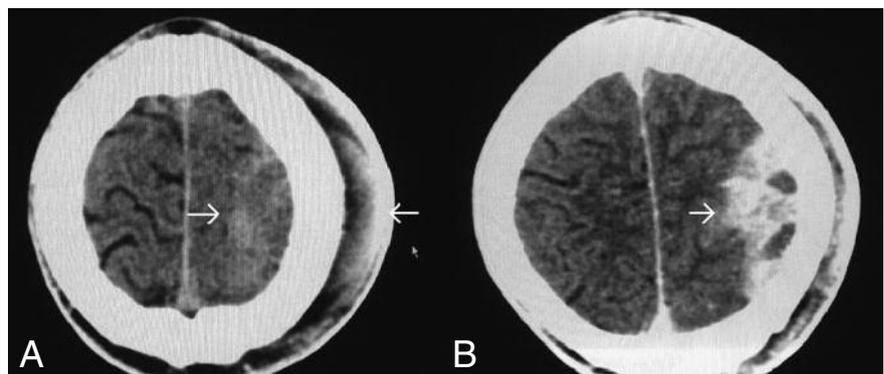


Fig. 1. — A. Non contrast enhanced CT image shows extra-cranial mass lesion with intra-cranial extradural extension in left rolandic region. B. Post contrast enhanced image shows heterogeneous enhancement of mass lesion.

On initial examination the patient was alert and oriented; there were no focal neurological deficits. Physical examination revealed a 6x6 cm firm smooth, non tender swelling in left parietal region. There was no local rise of temperature and overlying skin was normal. There was no lymphadenopathy, hepatomegaly or splenomegaly. Results of hematological and blood chemistry tests were within normal limits. X ray chest was normal. Patient was HIV infection negative.

Patient underwent computed tomography (CT) scan of the brain which confirmed the presence of the extra-cranial mass and showed its intra-cranial extradural extension in left fronto-parietal (rolandic) region. The extradural lesion exerted mass effect on the underlying brain in the form of displacement of gray-white

matter interface without any evidence of cerebral edema (Fig. 1). Lesion was slightly hyperdense than brain parenchyma on pre contrast study. After contrast administration the lesion was heterogeneously enhancing and bone window revealed permeative destruction of both inner and outer table (Fig. 2B). In addition CT scan also revealed another intracranial extradural lesion overlying right occipito-cerebellar region (Fig. 2B), however the underlying bone was normal. The superficial scalp mass was biopsied and diagnosis of low grade non-Hodgkin's B-cell lymphoma was made (Fig. 3A). Further work-up with CT examination of neck, thorax, abdomen and pelvis did not reveal presence of lymphoma at any other site. Ga-scintigraphy was negative. Whole-body fluorodeoxyglucose positron emission tomography (FDG-PET), bone marrow biopsy and cerebrospinal fluid examination also did not showed any other lesion.

Patient was treated with cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP)

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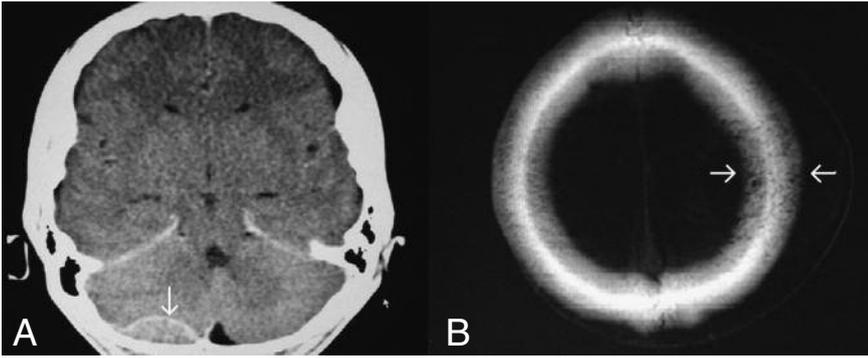


Fig. 2. — A. Post contrast CT image shows mass lesion in right occipito-cerebellar region. B. CT with bone window setting shows permeative destruction of left parietal bone without osteolysis.

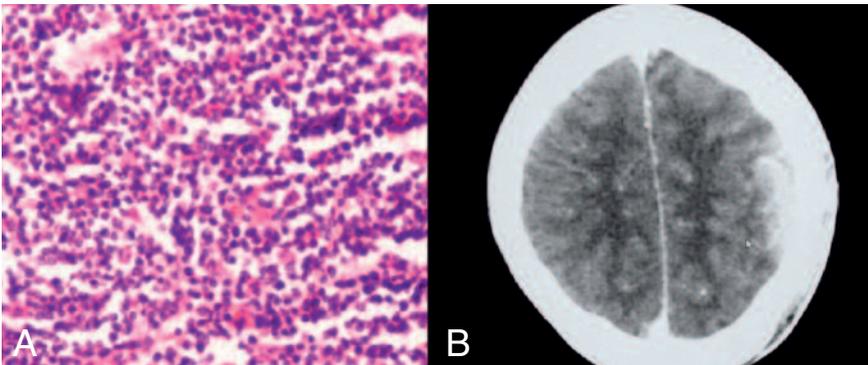


Fig. 3. — A. Microscopy of histopathological specimen. Hematoxylin and eosin stained section shows small cells with round to oval small nuclei and clumped chromatin with no nucleoli consistent with low grade lymphoma (original magnification $\times 400$). B. Post contrast CT image after two cycle of chemotherapy reveals significant reduction in size of lesion.

regimen. Superficial scalp swelling in left fronto-parietal region responded well and CT revealed resolution of intracranial lesion (Fig. 3B). Patient remains symptom free in 2-year follow up period.

Discussion

A variety of cases of primary CNS lymphomas has been reported in medical literature earlier. But only a few have characteristics similar to the one we have reported. The precise anatomical location and multifocal nature of the neoplasm occurring in nonimmunocompromised adolescent boy makes this case clinically and radiologically unique.

Bony involvement is commonly seen with secondary NHL. However, primary bone NHL is extremely rare with reported incidence of only 3-4% (1). By definition primary bone lymphoma is a solitary mass lesion with no evidence of disease at other sites and no systemic dissemination

within 6 months of the detection of tumor (3). Clinical presentation of NHL primarily involving the skull vault is unusual. However in recent times incidence of CNS lymphoma has increased in both immunocompromised and immunocompetent individuals. Lymphomas of the cranial vault usually involve the pericranium, meninges, and subcutaneous tissues. Histologically all reported lymphomas of the cranial vault are of B-cell origin. Most of cases are reported in middle aged and elderly women. Its occurrence in an immunocompetent adolescent boy as reported by us is uncommon.

Computed tomography (CT) features of primary bone NHL are non specific. Characteristic features are a permeative growth pattern with very little cortical destruction (4). Periosteal reaction is occasionally seen. Although lymphomas usually reveal homogenous appearance, most reported cases of the cranial vault lymphoma have demonstrated

heterogeneous hyperdensity (5). This finding is substantiated in our case and it may represent a characteristic feature of lymphomas of cranial vault. An associated large soft-tissue component is again very common with extension of mass in the extradural compartment and galeal compartment. Post contrast studies show mild contrast enhancement. Calcification is usually absent (5). Magnetic resonance (MR) imaging provides a better anatomic delineation in the evaluation of the soft tissue component and extradural extension of the lesion however it is also non specific. In our case MR imaging was not done because of financial constraint which is a commonly encountered problem in developing countries like us.

Considering the age of patient with CT finding of soft tissue mass and permeative pattern of bone destruction, Ewing's sarcoma is a likely differential diagnosis which is also one of the small round cell tumors. However it commonly involves long bone with femur being the most common site. Flat bones are commonly involved but pelvis and ribs are much more common sites than skull which is rarely involved. Patient usually presents with localized pain and swelling with systemic symptoms that simulate infection.

Primary malignant lymphoma of the skull vault may also mimic metastases or less frequently osteomyelitis, solitary fibrous tumor and granulocytic sarcoma (1, 6, 7). A metastatic carcinoma to the skull may spread to extradural compartment and brain parenchyma, however it usually demonstrates massive osteolysis. Patient with chronic osteomyelitis also demonstrate lytic bone lesions but systemic signs of infection are usually evident before scalp swelling is evident (4). Granulocytic sarcomas (chloromas) are rare and seen most commonly in children with acute myelogenous lymphoma. They have a predilection for the epidural space and skull and reveals osteolysis (7). Solitary fibrous tumor is an uncommon spindle cell tumor that usually occurs in the pleura. Extra pleural sites including meninges are rarely involved. Imaging features are nonspecific.

Suggestive radiological features along with Clinical characteristic such as the patient's age and the tumor site are important in diagnosing these lesions. Nevertheless definitive diagnosis of such rare lesion requires biopsy with immuno-

histochemistry. Treatment for extranodal Hodgkin lymphoma usually includes adriamycin containing regimen like CHOP protocol, followed by radiation therapy. Alternatively chemotherapy without subsequent radiation therapy and radiation therapy alone can be used in carefully selected cases (8). Clinical stage is the most important prognostic variable in predicting overall survival.

Conclusion

Although it is extremely uncommon, Primary malignant NHL of the skull vault should be considered in differential diagnosis of scalp masses. This case illustrates that this differential diagnosis should be considered not only with discrete and focal skull lesions in typical setting of immunocompromised middle age individual, but also with diffuse and multifocal vault infiltration in unfavorable clinical scenario like nonimmunocompromised individual and young age. Differentiation is important because their natural history and treatment outcome are

much different from other more common diagnoses in the skull. A thorough search for systemic disease is an essential part of the management. Characteristic CT features of primary malignant NHL of cranial vault are a permeative growth pattern, a large soft tissue component, and very little cortical destruction. Biopsy of the superficial scalp mass was sufficient to obtain a diagnosis, avoiding the risks associated with open intracranial biopsy.

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