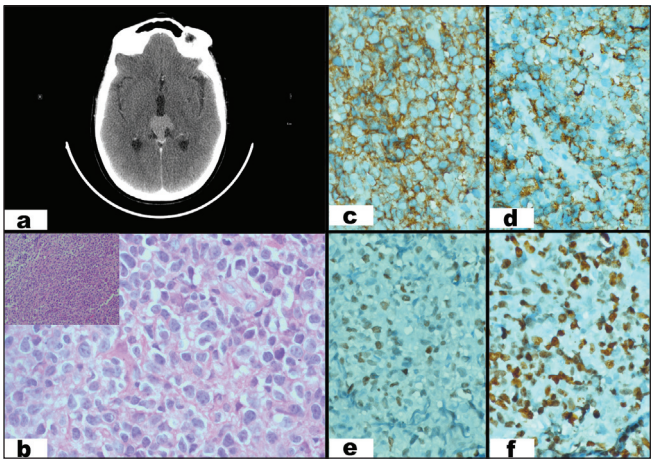

Figure 1: (a) Postcontrast axial computed tomography section of the brain shows a well-defined homogenously enhancing mass in the pineal region with no evidence of adjacent brain parenchymal infiltration. No cystic areas or calcifications seen. (b) Sheets of large tumor cells with scanty cytoplasm and round to oval nucleus with irregular nuclear membranes, fine chromatin and prominent nuclei (H and E, ×400). Inset showing the diffuse arrangement of tumor cells (H and E, ×200). (c-e) Tumor cells are positive for leucocyte common antigen, CD20, bcl-6. (f) Tumor cells show MIB 1 labeling index of around 70%

Primary diffuse large B-cell lymphoma of the central nervous system in pineal gland: Report of a rare case with review of literature

Editor,

Primary diffuse large B-cell lymphoma (DLBCL) of the central nervous system (CNS) in the pineal gland is an exceedingly rare occurrence. We present a case of a 58-year-old lady, who presented with behavioral and gait disturbances having a pineal gland mass and diagnosed as CNS DLBCL on histopathology.

The patient presented with a head ache, diplopia, behavioral changes, memory disturbances and unsteadiness of gait of 6 months duration. She was evaluated at an outside tertiary care center. Computed tomography scan (plain and contrast) showed a 1.6 cm × 1.3 cm lesion in the posterior third ventricle region causing a mass effect in the cerebral aqueduct with mild hydrocephalus [Figure 1a]. Radiologically, the picture was suggestive of a pineal mass possibly pineoblastoma with mild hydrocephalus. There she underwent third ventriculostomy and biopsy and diagnosed as pineoblastoma. She was referred to our center along with slides and blocks. Her general examination was with in normal limits. CNS examination revealed minimal ataxia and bilateral early papilledema. Pupils were bilaterally equal and reactive. No other cranial nerve deficits were present. Bulk and tone of muscle were normal in all the four limbs. Power was grade 5/5 in all the four limbs. Deep tendon reflexes were normal. No sensory deficits or cerebellar signs were present. Examinations of other systems were within normal limits. Routine laboratory investigations were within normal limits. Lactic dehydrogenase levels were normal. HIV antibody test was negative. Histopathology examination revealed a tiny fragment of cellular neoplasm composed of large round cells arranged in sheets. Individual cells were round with pale eosinophilic cytoplasm and round to the oval nucleus with irregular nuclear membranes, fine chromatin and prominent nucleoli [Figure 1b]. Immunohistochemistry showed the tumor cells to be positive for CD45, CD20 and bcl-6 [Figures 1c-e]. MIB 1 labeling index was around 70% [Figure 1f]. Other immunostains (synaptophysin, glial fibrillary acidic protein, and C-kit) were negative. Bone marrow and CSF study of the patient was normal. Thus, a diagnosis of CNS DLBCL of the pineal gland was made. After informed consent, the patient was started on Radiation Therapy Oncology Group (RTOG) protocol. She tolerated chemotherapy and is doing well.

Neoplasms of the pineal gland are exceedingly rare accounting for <1% of intracranial tumors in adults.[1] There are very few reports of pineal gland lymphomas.[2-4] CNS DLBCL in the pineal gland is not yet reported in the literature. Among pineal gland neoplasms, germ cell tumors are the most common followed in frequency by pineal parenchymal neoplasms, gliomas, meningiomas, and metastasis.[5] Our search of the literature revealed a case series of primary CNS lymphoma in which one of the sites mentioned is in the pineal gland.[6] However,

systemic lymphoma is not specifically excluded in that case. The majority of pineal gland tumors present at least by the third decade of life. Thus, other diagnosis should be entertained in pineal tumors occurring in older adults. Metastasis to pineal gland has been reported with low frequency.[7] Pineoblastomas can rarely occur in late adulthood.[8] Pineoblastomas are also composed of poorly differentiated discohesive cells with scanty cytoplasm and pleomorphic nuclei but unlike lymphomas they may exhibit nuclear molding, rosette formation and are negative for immunomarkers of lymphoma. Diagnostic work-up of a patient presenting with pineal gland mass includes serum tumor marker studies of germ cell tumors. Stereotactic biopsy of the pineal gland is recommended if the tumor marker levels are normal. Our patient was treated with RTOG protocol and is doing well.

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