case report

Anaplastic large cell lymphoma presenting as a cerebellar mass

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Anaplastic large cell lymphoma (ALCL) is a T cell lymphoma occurring commonly in childhood and rarely in adults. Central nervous system involvement in ALCL is very rare and cerebellar involvement at presentation has never been described. We examine the case of a young adult who presented with a cerebellar mass. A 19-year-old boy presented with signs of raised intracranial tension, which, on imaging, revealed a right cerebellar mass. He underwent suboccipital craniotomy and partial excision of the tumor. However, the histopathology was inconclusive. He subsequently presented with cerebellar signs and repeat imaging showed recurrence of the cerebellar lesion. He underwent decompression and ventriculoperitoneal (VP) shunting. Histopathology was suggestive of ALK (anaplastic lymphoma kinase) positive anaplastic large cell lymphoma. The patient was started on chemotherapy. However, his neurological status deteriorated, his condition worsened, and he expired a month later.

KEYWORDS: ALCL; Cerebellum; Brain; NHL

INTRODUCTION

Anaplastic large cell lymphoma (ALCL) is a T cell lymphoma accounting for 2–8% of all non-Hodgkin's lymphoma and 20–30% of childhood lymphomas.[1] Although ALCL is primarily a nodal disease, extranodal involvement is not uncommon and usually involves skin, bone, soft tissue, lung, and liver. Central nervous system involvement in ALCL is very rare and cerebellar involvement at presentation has never been described before. We examine the case of a young adult who presented with a cerebellar mass.

CASE REPORT

A 19-year-old boy was evaluated in a neurosurgery center for headache and vomiting. A computerized tomogram (CT) scan of his brain showed a focal, mildly enhancing hypodense lesion measuring 2–1 × 1.3 × 1.5 cm in the right cerebellar hemisphere close to the midline abutting the falx cerebellum (Figs. 1A & B). Magnetic resonance Imaging (MRI) showed a 2.9 × 2.2 cm well-defined right cerebellar lesion, hypointense on T1 and hyperintense on T2 weighted images showing contrast enhancement below the tentorium with obstructive hydrocephalus (Figs. 2A–E). He underwent suboccipital craniotomy and partial excision of the tumor. The histopathology did not reveal any neoplastic tissue and the patient was kept on follow-up. He was apparently asymptomatic for about nine months after which he developed repeated episodes of bifrontal headache associated with vomiting. He had unsteadiness of gait which gradually increased, and he was unable to walk. There was no history of loss of consciousness, seizures, visual disturbances, weakness of limbs, bladder or bowel disturbance. He gave history of diminished vision in right eye since early childhood. On examination, he was conscious with normal higher mental

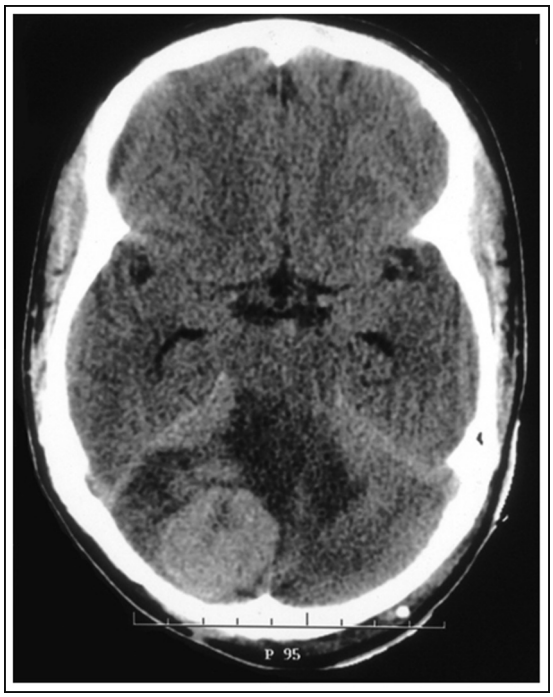

Figure 1A. CT brain plain axial image showing hyperdense lesion in the right cerebellum with surrounding edema.

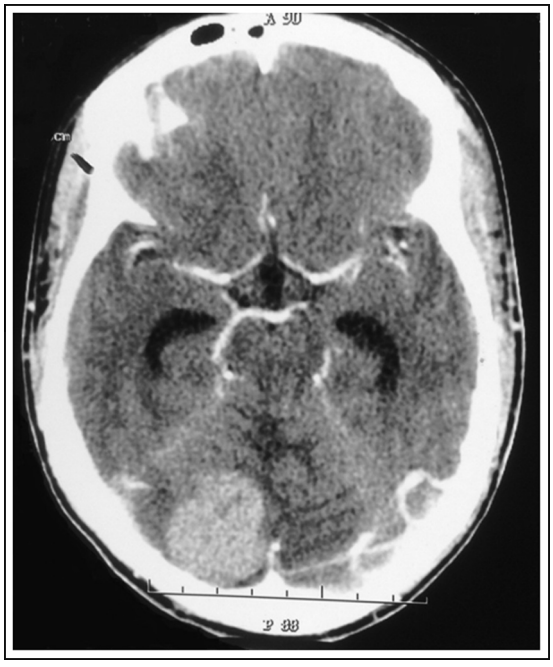

Figure 1B. CT Brain post contrast axial view showing the lesion in right cerebellar hemisphere.

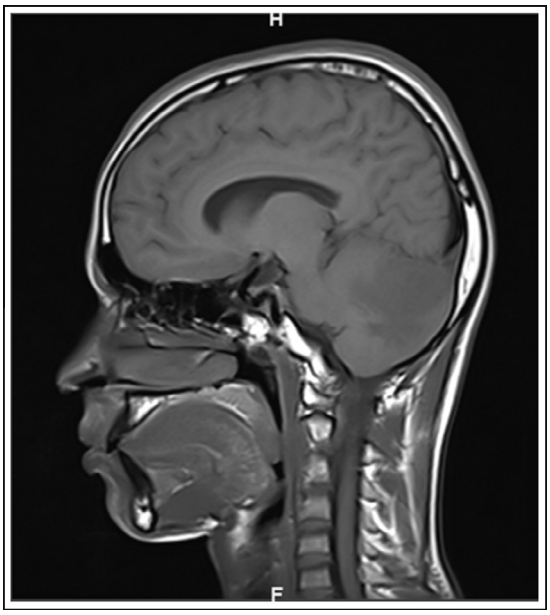

Figure 2A. MRI T1 sagittal view showing hypo intense lesion with surrounding edema in the cerebellum.

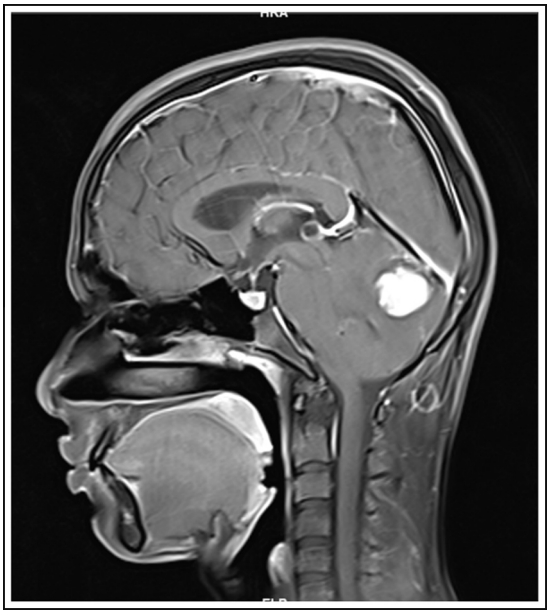

Figure 2B. MRI T1 post contrast sagittal view shows the same lesion with intense contrast enhancement.

functions. Pupils were equal and reactive, and extra-ocular movements were normal. Visual acuity was less on the right side. Nystagmus was present on the right side, and finger nose test was impaired on both sides. Fundi showed early papilledema. No other cranial nerve defects were present. Motor power was grade 4+ in all four limbs. CT brain showed recurrence of the right cerebellar lesion with surrounding edema and obstructive hydrocephalus. He underwent decompression and ventriculoperitoneal (VP) shunting. Post-operatively, the patient's condition deteriorated; he was ventilated, and he developed meningitis, which was managed with antibiotics.

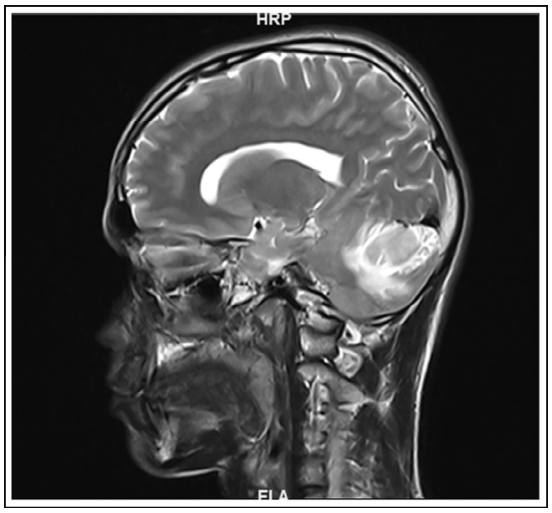

Figure 2C. MRI T2 weighted image sagittal view shows iso- to hyperintense lesion with surrounding edema.

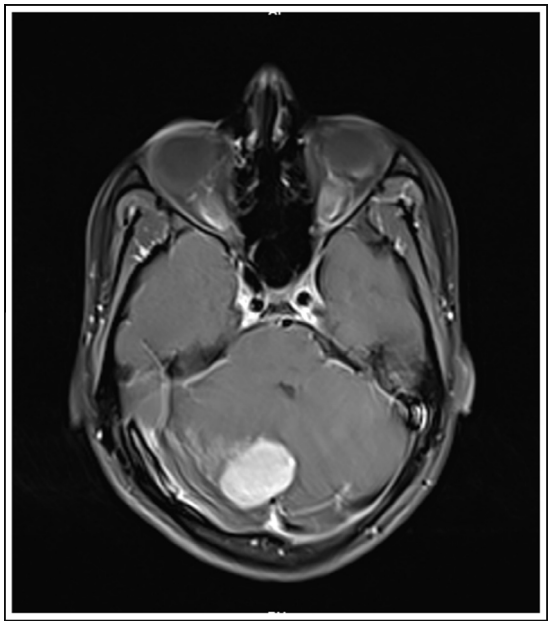

Figure 2D. MRI T1-contrat image axial view shows intense enhancement.

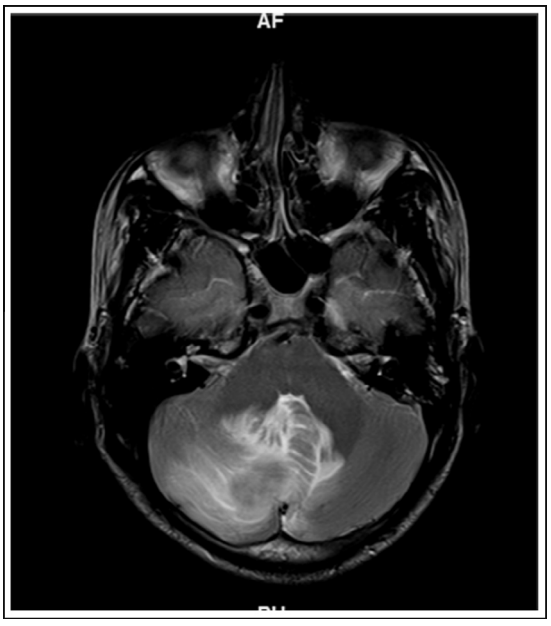

Figure 2E. MRI T2 axial image shows iso- to hyperintense lesion with surrounding edema.

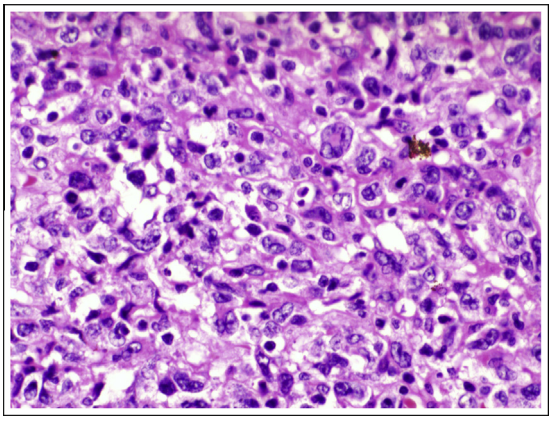

Figure 3. H&E × 400 showing sheets of pleomorphic tumor cells with classical doughnut cells.

Histopathology was suggestive of malignant lymphoma, and he was referred to us subsequently.

The patient's general condition was very poor. His hemoglobin was 12.5 gm%, total WBC 8900/mm^3, and platelet 352000/mm^3. His renal and liver functions were unremarkable and LDH was 1352. Ultrasonogram of abdomen showed very small, discrete, well-defined hypoechoic lesions in the para-aortic region. Bone marrow biopsy was normal. Histopathology was diagnostic of anaplastic large cell lymphoma (Fig. 3) which was positive for LCA, anaplastic lymphoma kinase (ALK) and CD30 (Figs. 4 & 5), and negative for CD5 and CD20 (Figs. 6 & 7). Skeletal survey and bone scan were normal. He was started on BFM90 ALCL protocol. He developed renal failure as part of tumor lysis syndrome, and he was subjected to hemodialysis. However, his neurological status deteriorated, his condition worsened, and he expired a month later.

DISCUSSION

ALCL, first described in 1985, represents a distinct category of large cell lymphoma defined by the strong expression of the cytokine receptor CD30 on all

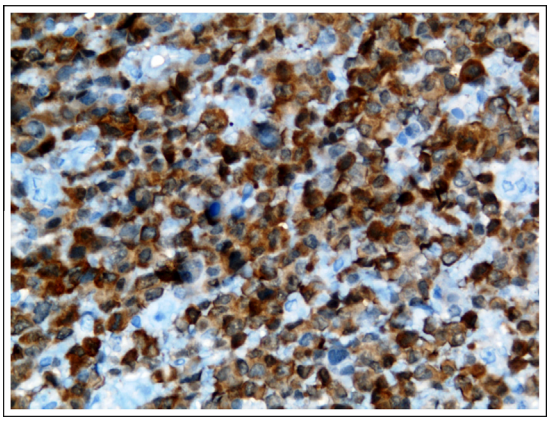

Figure 4. ALK × 400 Tumor cells are ALK positive.

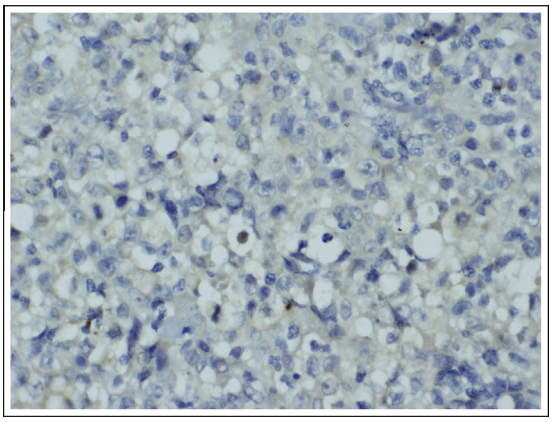

Figure 7. CD20 × 400 Tumor cells are negative for CD20.

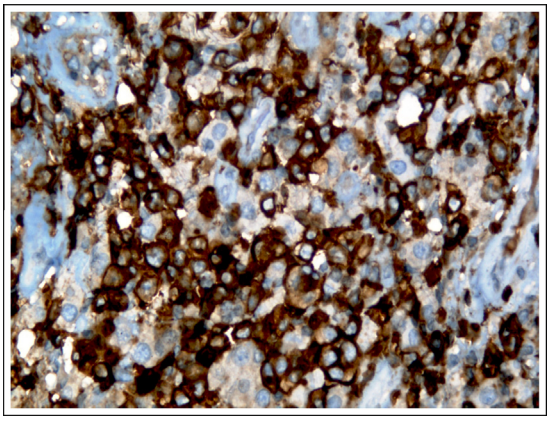

Figure 5. CD30 × 400 Tumor cells are CD30 positive.

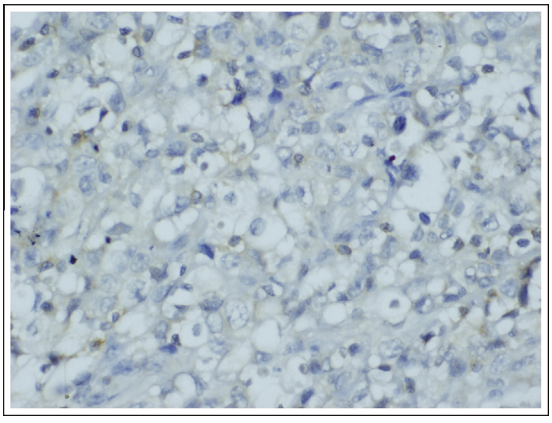

Figure 6. CD5 × 400 Tumor cells are negative for CD5.

neoplastic cells. ALK-positive ALCL is associated with a chromosomal abnormality, the t(2:5)(p23:q35) that fuses part of the nucleophosmin (NPM) gene on chromosome 5q35 to a portion of the ALK receptor tyrosine kinase gene on chromosome 2p23, resulting in expression of a chimeric NPM-ALK protein.[2] ALK+ ALCL is more commonly seen.

There are two forms of primary ALCLs: a primary systemic and primary cutaneous form. ALCL positive for the ALK protein frequently involves both lymph nodes and extranodal sites, which commonly include skin (21%), bone (17%), soft tissues (17%), lung (11%) and liver (8%) while involvement of the gut and central nervous system (CNS) is rare.[3]

Ponzoni reported the first case of primary brain CD30+, ALK+ ALCL with T cell phenotype in a 29-year-old male.[4] A 31-year-old male with ALK+ ALCL of leptomeninges, who was treated with high dose methotrexate and intrathecal chemotherapy, has also been described.[5] A 20-year-old male with primary central nervous system lymphoma (PCNS) ALCL was treated with chemotherapy and radiation, and survived for eight years.[6]

Cerebellar involvement in ALCL is very rare and has been described in only one patient, a 19-year-old female during the course of her treatment.[7] A cerebellar mass as a presenting manifestation of ALCL has never been described before and this is the first such case to be reported in the global medical literature. The most important prognostic factor in ALCL is ALK positivity, which is associated with a good prognosis. The overall five-year survival in ALK+ patients is 70% versus 49% in ALK cases.[8] Unfortunately, our patient died soon after starting chemotherapy.

DISCLOSURE

Authors do not have any conflict of interest or financial disclosure.

CONTRIBUTIONS OF AUTHORS

All authors have seen and approve the manuscript. G.N. – performed research, treated the patient, wrote the paper. S.K.P. – performed the research, provided radiology images. R.N. – provided pathology images. A.M. – provided pathology images.

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