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 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. 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. 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. 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.
functions. Pupils were equal and reactive, and extraocular 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.
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/mm³, and platelet 352000/mm³. 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
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. 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.

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

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. 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. 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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