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CASE REPORT

Post-traumatic basal ganglia haemorrhage in a child with primary central nervous system lymphoma

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SUMMARY

Primary central nervous system lymphoma (PCNSL) is a rare tumour of childhood with 15–20 cases reported yearly in North America. We present a case of a 13-year-old boy diagnosed with PCNSL who presented more than one-and-a-half years post-treatment with high dose cytosine arabinoside and methotrexate with a right-sided basal ganglia haemorrhage on MRI following a concussion while playing organised football against medical advice. There was no evidence of an underlying vascular malformation or recurrent disease by MRI, cerebrospinal fluid analysis or positron emission tomography computed tomography (PET-CT). However, 6 months post-injury he presented with asymptomatic disease recurrence of the frontal lobe. Our case reports an unusual MRI pattern of post-traumatic injury in a child previously treated for PCNSL that would support a recommendation for the avoidance of contact sports in this population.

BACKGROUND

Primary central nervous system lymphoma (PCNSL) is a rare tumour of childhood with 15–20 cases reported yearly in North America.[1] Given the rarity of this disease in children, little is known regarding clinical features, treatment strategies and prognosis. Clinical presentation of PCNSL may include severe headaches, vomiting, papilloedema, facial nerve palsy, diplopia, dysarthria, ataxia, bulbar palsy, quadriparesis and obtundation.[2–6] Rarer signs and symptoms including seizures, acute blindness, proptosis, lower and upper-limb muscle weakness, nystagmus, paraesthesias, personality change, lethargy and somnolence have been reported.[3 5 7–10] Intracranial haemorrhage either at presentation or subsequent to therapy has not been reported.

We report a case of a 13-year-old boy diagnosed with a cerebellar PCNSL who underwent successful chemotherapy treatment presented with a traumatic basal ganglia haemorrhage following a concussion more than 1.5 years postdiagnosis associated with a new mild hemiparesis. To the best of our knowledge this is the first case of a traumatic basal ganglia haemorrhage in a child associated with PCNSL.

CASE PRESENTATION

A 13-year-old boy presented to our hospital with progressive headaches, imbalance and acute episode of vomiting and fever. Neurological examination revealed gaze-evoked nystagmus, bilateral dysmetria and a wide-based ataxic gait. MRI imaging (figure 1)

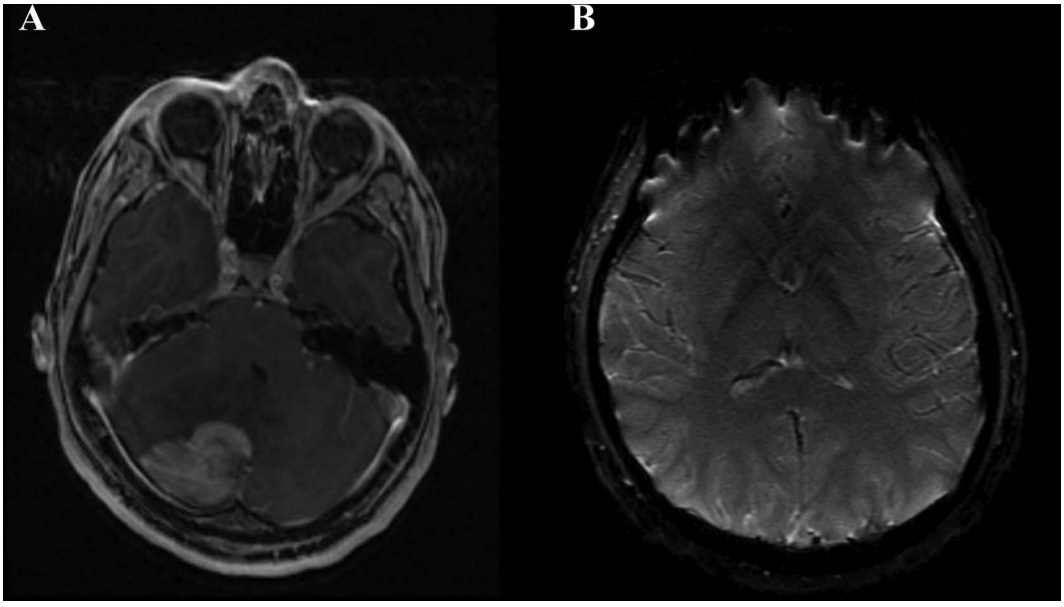

Figure 1 MRI characteristics of primary central nervous system lymphoma at presentation reveals contrast-enhancing cerebellar tumour on post gadolinium sequences (A) and no evidence of prior basal ganglia abnormalities on susceptibility-weighted sequences (B).

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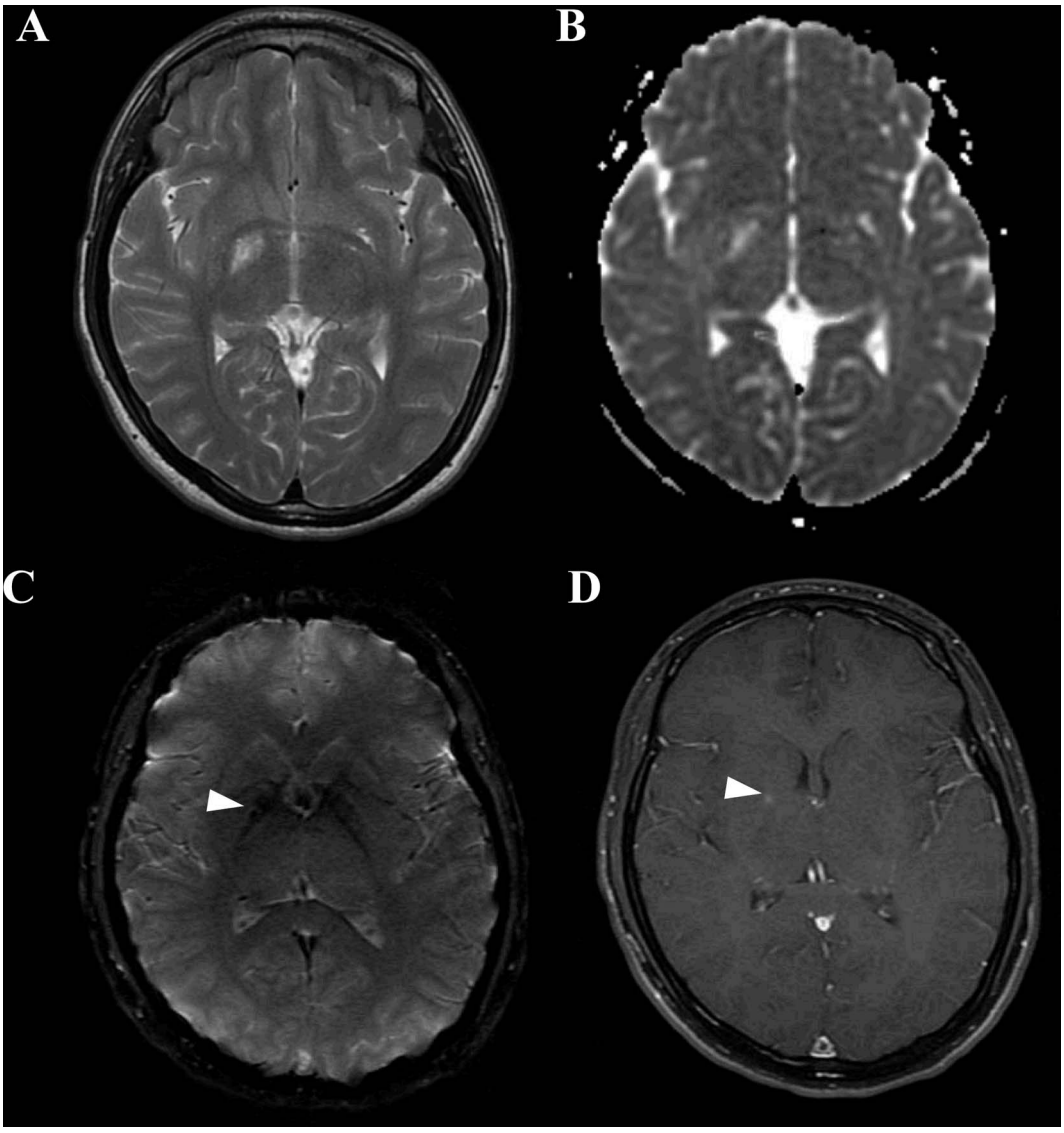

Figure 2 MRI features of right-sided lentiform nuclei haemorrhagic changes on T2 (A), apparent diffusion coefficient (B), susceptibility-weighted imaging (C) and postgadolinium (D) sequences consistent with a subacute haemorrhage.

showed a posterior fossa lesion with mass effect, partial effacement of the fourth ventricle and right greater than left inferior cerebellar tonsillar herniation. A biopsy was performed which revealed B cell lymphoma of the cerebellum. Paraffin-imbedded pathology along with immunostaining was positive for CD45 and CD20, and negative for ALK1. CT of the chest, abdomen and pelvis showed no evidence of a mass, suggesting the central nervous system (CNS) as the primary site. MRI of the spine ruled out any spinal dissemination. Cerebrospinal fluid (CSF) analysis, bilateral bone marrow aspirations and ophthalmologic examination were all negative for any signs of extracranial neoplasm. An immune workup consisting of quantitative immune globulins, T cell subsets and CH50 was unrevealing as was testing for HIV and Epstein-Barr virus. The patient was treated with four cycles of high-dose MTX and Ara C with no evaluable disease post-therapy. At 18 months post-treatment follow-up neuroimaging demonstrated abnormal signal within the right globus pallidus in association with new mild left hemiparesis on examination (figure 2). During the follow-up visit the patient admitted to sustaining a concussion without loss of consciousness while playing organised football a few weeks prior to the MRI study against medical advice.

INVESTIGATIONS

The lesion was followed with repeat MRIs that showed a gradual decrease in the size of the lesion over the course of the next 6 months. CSF cytology, MRI spectroscopy, MR perfusion and PET-CT were all negative for any evidence of recurrent neoplasm.

DIFFERENTIAL DIAGNOSIS

The radiological differential diagnosis of the basal ganglia lesion included late subacute/chronic haemorrhage or recurrent PCNSL. The cerebellum, which was the location of the original disease, demonstrated stable appearance with volume loss and mild gliosis without significant change.

OUTCOME AND FOLLOW-UP

The lesion continued to decrease in size on follow-up imaging. The patient's left-sided hemiparesis resolved; however,

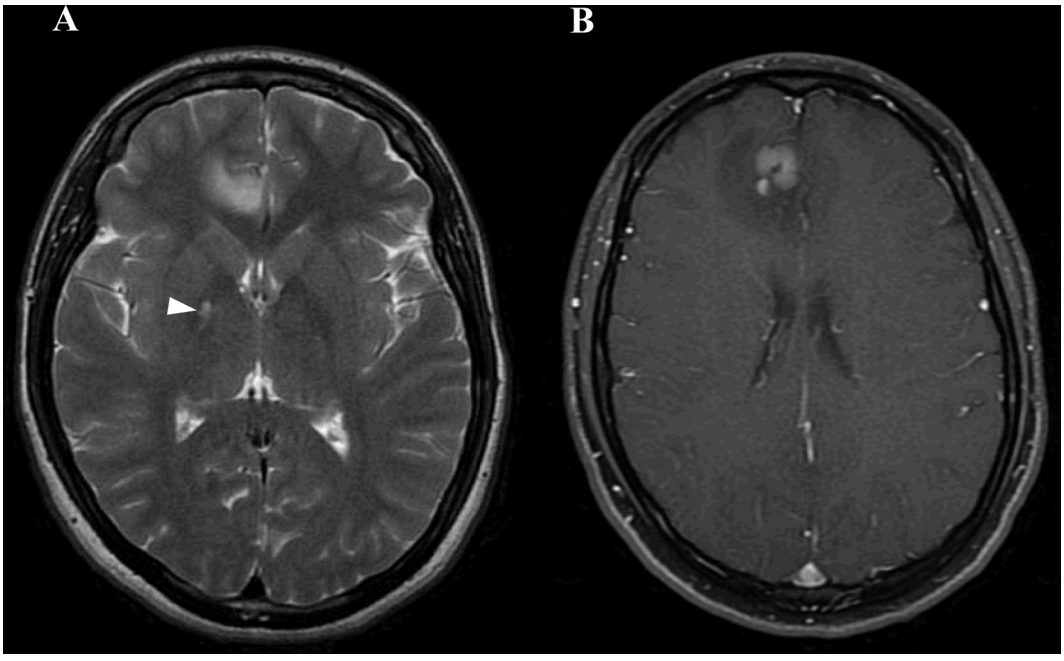

Figure 3 MRI findings at primary central nervous system lymphoma recurrence demonstrates a T2 hyperintense frontal recurrence (A) with postgadolinium enhancement (B) and improvement of the T2 haemorrhagic changes (arrowhead).

approximately 6 months after discovery of the right basal ganglia lesion, the patient started to develop increased frequency of headaches along with several episodes of vomiting. MRI was obtained and a new right medial frontal cortical and subcortical mass was discovered (figure 3). Imaging of the spine was negative for any new masses. The patient underwent biopsy of the frontal lobe lesion, which confirmed a diagnosis of PCNSL.

DISCUSSION

Traumatic basal ganglia haemorrhages (TBGH) in adults are uncommon with incidence ranging between 2% and 3% in closed head injuries.[11] However, in paediatrics their true incidence has yet to be determined. Kurwale *et al*[12] analysed 21 paediatric cases of TBGH and from their cohort found 11 cases isolated to the basal ganglia with the remainder occurring in association with haemorrhages in other locations. Symptoms that have been reported with TBGH include hemiparesis, dystonia and tremors.[12] Patients with PCNSL may present or develop different symptoms during the course of the disease. However, PCNSL in association with haemorrhage is quite rare in the adult population and until now has yet to be reported in a paediatric patient.[13 14] Only three cases of PCNSL in the adult population presenting with haemorrhage have been reported.[14–16] One case was reported in a patient with HIV and the other two were in immunocompetent patients. None of the three were associated with traumatic head injury nor related to prior chemotherapy treatment.[14–16] While we cannot definitely establish an association between TBGH with PCNSL in our patient, it is possible that either disease, prior therapy or a combination may have placed our patient at an increased risk of injury and is worthy of further study. Our reported finding would support a recommendation for the avoidance of contact sports in children diagnosed with PCNSL.

Learning points

- Primary central nervous system lymphoma (PCNSL) should be included in the differential diagnosis of T2 hyperintense, contrast-enhancing tumours in both supra and infratentorial locations.
- Traumatic basal ganglia haemorrhages in the paediatric population are rare and deserve an in-depth workup to exclude underlying pathology.
- Presenting features of traumatic basal ganglia haemorrhages may include hemiparesis, dystonia or tremors.

Contributors All authors contributed equally to the conception and writing of the manuscript. All authors have reviewed the case report and agree to its content prior to submission.

Competing interests None.

Patient consent Obtained.

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REFERENCES

1 Kadan-Lottic NS, Skluzacek MC, Gurney JG. Decreasing incidence rates of primary central nervous system lymphoma. *Cancer* 2002;95:193–202.
2 Abla O, Naqvi A, Ye C, *et al*. Leptomeningeal precursor B-cell lymphoblastic lymphoma in a child with minimal bone marrow involvement. *J Pediatr Hematol Oncol* 2004;26:469–72.
3 Abla O, Sandlund JT, Sung L, *et al*. A case series of pediatric primary central nervous system lymphoma: favorable outcome without cranial irradiation. *Pediatr Blood Cancer* 2006;47:880–5.
4 Taga T, Sakaue Y, Anzai Y, *et al*. Pediatric primary leptomeningeal lymphoma treated without cranial radiotherapy. *Pediatr Blood Cancer* 2007;4:477–8.
5 Uhlenberg B, Reich S, Varnholt V, *et al*. Atypical manifestation of childhood primary cerebral lymphoma restricted to the leptomeninges. *Eur J Pediatr* 2005;164:653–4.

6 Wynn R, Arkwright PD, Haque T, *et al*. Treatment of Epstein-Barr-virus-associated primary CNS B-cell lymphoma with allogeneic T-cell immunotherapy and stem-cell tranplantation. *Lancet Oncol* 2005;6:344–6.
7 Baleydier F, Galambrun C, Manel AM, *et al*. Primary lymphoma of the pituitary stalk in an immunocompetent 9-year-old child. *Med Pediatr Oncol* 2001;36:392–5.
8 Felice MS, Zubizarreta PA, Rossi JG, *et al*. Diagnosis and successful treatment of childhood primary leptomeningeal lymphoma. *Med Pediatr Oncol* 2000;34:361–3.
9 Shehu BB. Primary central nervous system Burkitt's lymphoma presenting with proptosis. *Ann Trop Pediatr* 2003;23:319–20.
10 Schulman H, Hertzanu Y, Maor E, *et al*. Primary lymphoma of brain in childhood. *Pediatr Radiol* 1991;21:434–5.
11 McPherson P, Teasdale E, Dharkar S, *et al*. The significance of traumatic hematoma in the region of basal ganglia. *J Neurol Neurosurg Psychiatry* 1986;49:29–34.
12 Kurwale NS, Gupta DK, Mahapatra AK. Outcome of pediatric patients with traumatic basal ganglia hematoma: analysis of 21 cases. *Pediatr Neurosurg* 2010;46:267–71.
13 Coulon A, Lafitte F, Hoang-Xuan K, *et al*. Radiographic findings in 37 cases of primary CNS lymphoma in immunocompetent patients. *Eur Radiol* 2002;2:329–40.
14 Kim IY, Jung S, Jung TY, *et al*. Primary central nervous system lymphoma presenting as an acute massive intracerebral hemorrhage: case report with immunohistochemical study. *Surg Neurol* 2008;70:308–11.
15 Fukui MB, Livstone BJ, Meltzer CC, *et al*. Hemorrhagic presentation of untreated primary CNS lymphoma in a patient of AIDS. *AJR Am J Roentgenol* 1998;170:1114–15.
16 Rubenstein J, Fishbein N, Aldape K, *et al*. Hemorrhage and VEGF expression in a case of primary CNS lymphoma. *J Neurooncol* 2002;58:53–6.