CASE REPORT

Extra-axial primary non-Hodgkin's CNS lymphoma mimicking meningioma, in a 5-year-old immunocompetent child: a rare entity

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SUMMARY

We describe a case of a 5-year-old immunocompetent girl who presented with features of raised intracranial pressure with left eye ptosis of 1-month duration. CT scan and MRI of the brain showed an extra-axial, intensely contrast enhancing lesion in the left temporoparieto-occipital region, consistent with meningioma. On open tissue biopsy and immunohistochemistry staining, a diagnosis of B cell non-Hodgkin's lymphoma was made. Six cycles of chemotherapy with cyclophosphamide, adriamycin, vincristine and prednisolone regimen were given and showed a good clinical outcome without any recurrence during follow-up of 5 months.

BACKGROUND

Primary central nervous system lymphoma accounts for about 2% of all brain tumours.¹ Non-Hodgkin's lymphoma in an immunocompetent paediatric patient has been rarely reported.^{2,3} We report a case of extra-axial non-Hodgkin's central nervous system (CNS) lymphoma in an immunocompetent paediatric patient.

CASE PRESENTATION

A 5-year-old immunocompetent girl presented with history of headache and vomiting of 1 month duration. On examination, there was no lymph node enlargement. Neurological examination showed bilateral grade-3 papilloedema and left-sided third and sixth cranial nerve paresis with no other neurological deficits.

INVESTIGATIONS

All routine laboratory investigations including viral markers and chest X-ray were normal. Non-contrast and contrast CT of the head revealed a large, well defined, dural based, hyperdense mass in the left temporoparieto-occipital region, with homogenous, intense contrast enhancement (figure 1A, B). MRI T1-weighted and T2-weighted (T1W1 and T2W1) images showed a hypointense to isointense mass with uniform enhancement on gadolinium administration (figure 1C–G). MR-venography of the brain showed attenuation of signals in the left transverse sinus and internal jugular vein, which was suggestive of compression of these venous structures by the mass. A radiological impression of supratentorial meningioma and the slight possibility of dural-based lymphoma

or germ cell tumour were kept in mind and the patient was prepared for surgery.

DIFFERENTIAL DIAGNOSIS

Meningioma, dural-based lymphoma and germ cell tumour were considered.

TREATMENT

A left temporoparieto-occipital craniotomy with gross total excision of the lesion was performed. Intraoperatively, the tumour appeared to be extra-axial, fleshy, moderately vascular and greyish-brown, with a well-defined plane. The surrounding parenchyma was compressed but uninvolved. Involvement of the left transverse sinus was noted. Postoperative CT showed complete excision of the lesion. Histopathological examination of the specimen showed a malignant lymphoid neoplasm against a mild inflammatory background. The individual tumour cells were larger than lymphocytes, and had a high nucleocytoplasmic ratio, and coarse chromatin and prominent nucleoli with scanty cytoplasm. Areas of mitosis were also evident at places (figure 2A, B). The proliferation fraction, as detected by Ki67, was high (60–70%). Immunohistochemistry was performed for confirmation and typing of the lymphoma; the tumour cells were strongly positive for CD45 leucocyte common antigen (figure 2C) along with CD20, CD4 and CD5 (figure 2D) and negative for cytokeratin and desmin. Based on immunohistochemistry, a diagnosis of B cell non-Hodgkin's lymphoma was established.

OUTCOME AND FOLLOW-UP

The patient was subsequently referred to the paediatric-oncology department, where a six cycle cyclophosphamide, adriamycin, vincristine and prednisolone (CHOP) regimen was given. She showed a good clinical outcome with adjuvant chemotherapy. There was no evidence of recurrence of the lesion at 5-month follow-up.

DISCUSSION

Non-Hodgkin lymphomas represent only about 4% of all lymphomas in the general population and occur more frequently in patients with AIDS.³ Direct involvement of the CNS occurs in 1–2% of patients with lymphoma. Common clinical manifestations of primary CNS lymphoma are in the form of raised intracranial pressure (headaches and vomiting), decrease in vision, fever, altered mental status, seizures and cranial nerve involvement or

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Figure 1 CT scan and MRI.

Figure 2 Histopathology and immunohistochemistry.

focal neurological deficit.^{3 4} Our patient presented with headache, vomiting and cranial nerve involvement. They have a male preponderance and a median age of presentation in the fifth or sixth decade in immunocompetent patients, but also occur in a much younger age group (mean age 37–43 years) in immunocompromised patients.^{5 6} Our patient was HIV negative and presented at a much younger age than patients in the literature. MRI findings in lymphoma are non-specific and show variable signal intensities on T1W1 and T2W1 images and ring enhancement in intra-axial lesions. Mostly these tumours are hypointense on T1W1 and hyperintense on T2W1, with intense enhancement after gadolinium administration and high ¹⁸F-fludeoxyglucose uptake on positron emission tomography scan.⁷ Differential diagnosis includes germ cell tumour, meningioma and haemangiopericytoma. The majority of primary CNS lymphomas are of B cell origin (as in our case). Chemotherapy (CT) and radiotherapy (RT) are the conventional treatment modalities for CNS lymphomas, but no standard protocol exists. The CHOP regimen was among the first combinations to produce complete response and long-term survival.

For patients with follicular and diffuse large B cell lymphoma, the addition of a monoclonal antibody against CD20 (rituximab) to CHOP is beneficial. The complete response rate with high-dose methotrexate-based chemotherapy plus RT was 61.1%, the median survival time was more than 26 months and the 2-year survival rate was 65.1% in a study by Yi *et al.*⁸ Prognosis for patients with such tumour is grim, with a median survival rate of <2 years with conventional CT/RT and with a 5-year survival of <5% despite CT/RT.⁴

Competing interests None declared.

Patient consent Obtained.

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Learning points

- Although extremely uncommon, extra-axial primary central nervous system (CNS) non-Hodgkin's lymphoma may occur in an immunocompetent paediatric patient.
- Radiological diagnosis of extra-axial CNS lymphoma is difficult and high index of clinical suspicion is warranted.

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