CASE REPORT

Primary dural diffuse large B cell lymphoma mimicking parafalcine meningioma

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SUMMARY

A 58-year-old immunocompetent woman presented with complaints of recent onset recurrent generalised tonic-clonic seizures and left hemiparesis. On MRI a dural-based heterogeneously enhancing mass was seen along the falx cerebri mimicking parafalcine meningioma, causing brain parenchymal compression and vasogenic oedema in the right frontal and parietal lobes. The tumour was completely excised and on subsequent investigation it was diagnosed as diffuse large B cell type primary dural lymphoma. Chemotherapy with CHOP (cyclophosphamide, hydroxyl doxorubicin, vincristine and prednisone) regimen was started; however, the patient later died from episodes of acute pulmonary thromboembolism, aspiration pneumonia and septic shock.

BACKGROUND

Primary dural lymphoma is a rare disease with few cases reported in the literature. They are mostly low-grade marginal zone lymphomas and have a better prognosis than intraparenchymal primary CNS lymphoma (PCNSL). We report a case of diffuse large B cell type primary dural lymphoma, which presented as an extra-axial dural-based mass along the falx cerebri, mimicking parafalcine meningioma on MRI. Diffuse large B cell type primary dural lymphoma is an extremely rare entity with fewer than 10 cases reported in the literature.

CASE PRESENTATION

A 58-year-old immunocompetent woman presented with complaints of recurrent generalised tonic-clonic seizures for 15 days associated with weakness in the left upper and lower limbs. On examination she was conscious, orientated and her higher mental functions were intact. Power was decreased in the left upper and lower limbs (3/5) with extensor planter response in the left foot. On MRI brain (figure 1A–H), a large dural-based extra-axial mass was seen in the right parafalcine location with part of it extending across the midline below the free edge of the falx cerebri. The mass was iso to hypointense on T1-weighted imaging and mildly hyperintense on T2-weighted imaging/FLAIR. Post-contrast scans showed heterogeneous enhancement of the mass with non-enhancing areas in the centre. On susceptibility-weighted imaging, areas of blooming were noted in the central part of the mass suggestive of haemorrhage. Also, significant diffusion restriction was noted in the peripheral

part of the mass. The mass was compressing the body of the corpus callosum with probable infiltration of the medial right frontal lobe. There was extensive oedema in the adjacent right frontal and parietal lobes with presence of mild midline shift towards the left. On MR spectroscopy, lipid/lactate and choline peaks were present with decreased N-acetylaspartate.

Based on the imaging findings and clinical presentation, parafalcine meningioma was considered as the provisional diagnosis.

Surgical excision of the mass was performed. Intraoperatively, the tumour was adhered to the dura and there was an ill-defined plane of cleavage with the right middle frontal gyrus. The gross appearance was pinkish, firm, gritty, partly suckable and moderately vascular. The whole of the tumour was excised and middle frontal gyrus cortisectomy was performed. Postoperatively, the patient developed left-sided hemiplegia and was mostly bed-ridden. Limb physiotherapy and intermittent pneumatic compression were initiated in the early postoperative period.

Histopathological evaluation of the excised mass was performed. On H&E staining, sheets of medium to large size atypical lymphoid cells displaying round to oval nuclei with coarse chromatin and scant cytoplasm were seen in a fibrillar stroma (figure 2A). On immunohistochemistry, the cells were diffusely positive for leucocyte common antigen and CD20 but negative for CD3, glial fibrillar acid protein and epithelial membrane antigen (figure 2B–D). There were areas of necrosis and haemorrhage within the mass. Based on these findings, the diagnosis of high-grade diffuse large B cell lymphoma was confirmed.¹

The patient was referred to the haematology department for further management. Staging investigation was done to rule out systemic disease. Contrast-enhanced CT scan of neck, chest, abdomen and pelvis was normal. Ophthalmological examination, bone marrow biopsy and lumbar puncture for cerebrospinal fluid analysis did not show any abnormality. Based on the clinical evaluation, imaging and histopathological investigation, a final diagnosis of primary dural high-grade diffuse large B cell non-Hodgkin's lymphoma was made.

OUTCOME AND FOLLOW-UP

Subsequently the patient was started on chemotherapy with CHOP (cyclophosphamide, hydroxyl doxorubicin, vincristine and prednisone) regimen. On day 10 of the first session of chemotherapy the

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Figure 1 (A, B) Axial T2-weighted and FLAIR images showing a hyperintense extra-axial right parafalcine mass with extensive oedema in the right frontal and parietal lobes. (C) T2-weighted coronal image showing compressed corpus callosum and ill-defined cleavage plane between the mass and the right medial frontal lobe. (D) Susceptibility-weighted image showing multiple foci of blooming suggestive of haemorrhage within the mass. (E) Pre-contrast and (F) post-contrast T1-weighted images show heterogeneous enhancement of the mass. (G) Diffusion restriction in the peripheral part of the mass. (H) MR spectroscopy shows lipid-lactate and choline peaks with reduced N-acetylaspartate.

patient developed lower limb deep venous thrombosis followed by pulmonary thromboembolism and reported to emergency with complaints of dyspnoea, high-grade fever and loss of consciousness. She was started on anticoagulants, antibiotics and supportive treatment, but during the course of her stay she developed aspiration pneumonia, progressive respiratory distress and septic shock and could not be revived even after all possible efforts.

Figure 2 (A) Section showing a tumour in a sheet of medium to large sized atypical lymphoid cells in fibrillar stroma. The atypical cells display round to oval nuclei with coarse chromatin and scant cytoplasm (H&E, 200x). On immunohistochemistry, these tumour cells were diffusely positive for (B) leucocyte common antigen and (C) CD20 and focal positive for CD5 (D).

DISCUSSION

CNS involvement in lymphoma can be primary or more commonly secondary to systemic disease. PCNSL, including parenchymal and meningeal disease, accounts for less than 1% of all non-Hodgkin's lymphoma and approximately 2% of all intracranial tumours.^{2 3} A subset of PCNSL is primary dural lymphoma, which is rare with only a handful of cases reported in the literature. Histologically, most of these reported cases were of low-grade marginal zone lymphoma, which has a better prognosis than more aggressive intraparenchymal PCNSL.^{2 4-8} Diffuse large B cell lymphoma histology in primary dural lymphoma is extremely rare. A review of the available literature found fewer than 10 documented histopathologically proven cases of diffuse large B cell primary dural lymphoma.^{5 9} Iwamoto *et al*, after a review of the literature in 2006, compiled 56 previously reported cases of primary dural lymphoma, of which only two cases were of diffuse large B cell primary dural lymphoma.⁷ Nearly all the reported cases of diffuse large B cell primary dural lymphoma occurred in immunocompetent patients (including the current case), whereas intraparenchymal PCNSL is one of the most frequent brain tumours in immunocompromised patients.^{2 10} It is more common in women, in contrast to intraparenchymal PCNSL which is more common in men.^{5 10} The mean age at diagnosis is 51 years; however, in the literature there is one case report of this disease in a 14-year-old boy.⁹

Dura is devoid of lymphoid tissue, so the exact pathogenesis is not clear. Several theories exist, including chronic inflammatory conditions, autoimmune aetiologies or even chronic infections resulting in monoclonal proliferation of lymphoid cells.^{2 4 10}

Radiologically, the closest differential diagnosis is meningioma, and generally it is not possible to differentiate it on neuroimaging alone. In one case series, 14 out of 15 cases were diagnosed as meningioma on imaging, which later proved to be primary dural lymphoma.^{2 5 7 11} However, evidence of adjacent

structure invasion, diffusion restriction on MRI or extensive brain parenchymal vasogenic oedema is the clue to suspect a more aggressive pathology than meningioma.^{6,9} Clinically also, they share multiple features including being more common in women, similar age group and clinical presentation (headache, seizures, focal neurological deficits, meningeal signs and sometimes cranial nerve involvement).^{4,7,11,12} The presence of lipid/lactate and choline peaks with reduced N-acetylaspartate on MR spectroscopy (as in our case) favours primary dural lymphoma over meningioma and is associated with a poor prognosis.¹³ Nearly all previously reported cases of diffuse large B cell primary dural lymphoma were found to be associated with lytic bony involvement and vasogenic oedema of adjacent brain parenchyma; however, our case is unique in the sense that it involved falx cerebri away from calvarial bones and in addition to extensive brain parenchymal oedema; it also showed the presence of haemorrhage within the mass on histopathology, which was picked up beforehand on susceptibility-weighted MRI as areas of blooming in the central part of the mass. Other differential diagnoses include dural deposit secondary to systemic lymphoma, metastasis from breast or prostate carcinoma, granulomatous disorders and some rare dural tumours like gliosarcoma.¹³

Since this condition is rare, there is a paucity of sufficient data to devise the most appropriate treatment.^{2,7} Usually these cases are managed as extranodal Hodgkin's lymphoma affecting a single site. Previously reported cases of diffuse large B cell primary dural lymphoma have been treated with surgery, chemotherapy±radiotherapy.^{2,5,7,9} In one case reported by Angelo *et*

al, a good sustained response was achieved with chemotherapy alone.⁵ The prognosis is usually better than for intraparenchymal PCNSL.⁷ So far as the role of steroids in the treatment is concerned, PCNSL is known for its significant and rapid response to glucocorticoid administration. Moreover, the initial response to steroids has a positive prognostic effect on the overall response to chemoradiation in PCNSLs.¹⁴ Unlike in high-grade gliomas and metastases, steroid treatment leads to actual tumour destruction in addition to its effect on the associated vasogenic oedema in cases of lymphoma. The lympholytic effect of steroid is non-p53-dependent, unlike the innate apoptotic mechanism of the body, and this may produce a false negative result on histopathological evaluation.¹⁵ For this particular effect, steroids should be deferred until the biopsy confirmation is obtained in radiologically suspected cases of intracranial lymphomas. Although there are reports of sustained and complete remission of disease with steroids alone, most patients relapse eventually.¹⁵ However, because of their rapid action, steroids remain very useful in patients with raised intracranial pressure secondary to tumour mass effect and associated oedema.

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Learning points

- Even though primary dural lymphoma is rare, it must be considered in the differentials of dural masses, especially when the clinical and radiological presentation is non-classical, as the prognosis depends on early diagnosis and subsequent management.
- The closest differential is meningioma; however, the presence of lytic bony involvement, extensive brain parenchymal oedema, necrosis and haemorrhage within the mass suggest more aggressive pathology and one must exclude primary dural lymphoma and other uncommonly occurring dural-based lesions. Extensive clinical, radiological and biochemical work-up is imperative to diagnose these conditions before directly embarking on a neurosurgical procedure.
- Recent advanced modalities like functional MRI and diffusion tensor imaging fibre tractography delineate the eloquent cortex and corticospinal tracts, respectively, in relation to the lesion and help in proper surgical planning. Intraoperative squash smear cytology/frozen section biopsy should be obtained wherever possible as it may guide the surgeon towards less extensive excision of the tumour in these cases.
- Treatment includes surgery, chemotherapy±radiotherapy. Prevention of deep venous thrombosis in cases of malignant disease is of utmost importance, as it may result in pulmonary thromboembolism and subsequent increase in morbidity and mortality. In these high-risk cases, especially where the patient is not ambulatory, early institution of pharmacological prophylaxis should be considered as well as limb physiotherapy and intermittent pneumatic sequential compressions.

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