Novel treatment (new drug/intervention; established drug/procedure in new situation)

Tumefactive demyelination-to cracks the nut without cracking the pot

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Summary

A 45-year-old female recently detected diabetic, admitted with difficulty in walking, irrelevant talk and urinary incontinence for 3 weeks and deviation of angle of mouth to left. Examination revealed pallor, delusional thoughts, right upper motor neuron facial weakness, ataxia. CT brain revealed bulky hyperdense lesion in corpus callosum and subsequently MRI of the brain revealed T2-hyperintense lesions involving genu and body of corpus callosum with restricted diffusion, and MR spectroscopy revealed reduced uptake of choline. Possibilities considered were infiltrative glioma of corpus callosum, demyelination and central nervous system lymphoma. As the patient and relatives were not willing to allow stereotactic biopsy, she was started on steroids. Her neurological deficits started improving and 1 month later repeat MRI brain showed a drastic reduction in the size of the lesion. Her neurological condition disappeared and is doing well.

BACKGROUND

Tumefactive demyelinating lesions (TDL) are defined as solitary demyelinating lesion with a dimension of >2 cm.¹ These lesions occupy a spectrum between multiple sclerosis and acute demyelinating encephalomyelitis.² It often presents atypically, including in the form of a focal weakness or seizure episodes, and occasionally presents as status epilepticus, intractable headaches or with features of raised intracranial tension.² There is a female predominance, with average age being 37, and usually does without history of similar episodes² or an antecedent viral infection or vaccination.¹ Differential diagnosis is that of central nervous system (CNS) tumours, including high-grade CNS glioma and lymphoma. As imaging findings are highly non-specific, the diagnosis of TDL needs biopsy and histological confirmation. It is hence prudent to give a course of steroids for those patients in whom the clinical profile and imageology do not absolutely favour CNS malignancy and those not willing for biopsy, and

should be reassessed after a period of 4–6 weeks with repeat imaging.

CASE PRESENTATION

A 45-year-old female recently detected diabetic, admitted with difficulty in walking, irrelevant talk and urinary incontinence for 3 weeks, and deviation of angle of mouth to left for 3 days, admitted to medical ward. On examination, she was found to be pale and nervous system examination revealed that she had delusions. Ocular movements were full and pupillary reaction was normal bilaterally. Right-sided upper motor neuron facial weakness was found. Lower cranial nerves were normal. Ataxia was present. The power of all four limbs was grade 5. Deep tendon reflexes were normal on both sides. Plantar reflex flexor on both sides. No sensory loss. Fundus was normal. All the other systems were within normal limits. Local examination showed a large ulcer on the back extending from root of scapula to sacrum in healing stage with healthy granulation.

Figure 1 (A) CT-brain (sagittal)-showing bulky hypo dense lesion involving genu and body of corpus callosum (white arrow). (B) CT-scan brain (coronal)-showing bulky hypodense lesion in the corpus callosum (white arrow).

Figure 2 (A) MRI brain T2 weighted images (coronal view) showing hyper intense lesion involving corpus callosum, with swelling of corpus callosum (white arrow). (B) MRI brain T1 weighted images (coronal view) showing hyper intense lesion involving corpus callosum with swelling of corpus callosum (white arrow).

INVESTIGATIONS

Her haemoglobin was 9 g%, total count 10 000/mm³, differential count polymorph 60%, lymphocyte 34%, eosinophil count 6%. The erythrocyte sedimentation rate was 60 millimetres at the end of the first hour. The peripheral smear was normal. Antinuclear antibody, ds-DNA, HIV, HbSag, Venereal Disease Research Laboratory test were negative and serum ferritin normal. Her fasting blood sugar was 120 mg/dl, urea 34 mg%, serum creatinine 1 mg%. Serum electrophoresis, α feto protein, carcino embryonic antigen and thyroid function tests were normal. Urine porphobilinogen was absent. Culture and sensitivity from the ulcer was negative. Lumbar puncture could not be done because of the large ulcer over the back. CT brain revealed bulky hyper dense lesion in corpus callosum (figure 1A,B) and subsequently MRI brain revealed T2-hyperintense (figure 2A–C) with restricted diffusion and MR spectroscopy revealing reduced uptake of choline. The lesion was hypoperfused. There was no evidence of micro haemorrhage. With this, the possibilities of infiltrative glioma of corpus callosum, demyelination and CNS lymphoma were present. A brain biopsy was planned but the patient and relatives were unwilling to undergo it.

DIFFERENTIAL DIAGNOSIS

1. Corpus callosal demyelination
2. Central nervous system lymphoma
3. Glioma

TREATMENT

During her in-hospital stay she was on regular insulin, which was adjusted according to her blood sugar and antibiotics with regular dressing of the wound. As the patient and relatives were not willing to undergo stereotactic biopsy, she was started on intravenous methyl prednisolone 1 gram daily for 5 days and was started on oral steroids, which were tapered off in a month.

OUTCOME AND FOLLOW-UP

Her neurological deficits started improving and 1 month later, repeat MRI brain showed drastic reduction in size of the lesion to merely a thin rim (figure 3A,B). She is under regular follow-up and doing well.

DISCUSSION

TDL are defined as solitary demyelinating lesion with a dimension of >2 cm.¹ These lesions occupy a spectrum

Figure 3 (A) MRI brain T2 weighted images (sagittal) view showing thinned out rostrum, genu and body of corpus callosum, (white arrow) and shows minimal hyperintensity (white arrow). (B) MRI brain T1 weighted images (coronal view) corpus callosum showing minimal hyperintensity (white arrow).

between the classical multiple sclerosis in which the demyelinating plaques are of smaller size and the lesions are distributed in space and time, and the acute demyelinating encephalomyelitis in which there is extensive white matter involvement, and there is usually history of an antecedent viral infection.² The clinical presentation of these lesions is often atypical and the usual presenting features are that of focal neurological deficit. Thus, the closest differential diagnosis for a TDL is that of CNS tumours, especially when the disease manifests as large demyelinating lesions within the cerebral hemisphere when the first differential will be a high-grade CNS glioma owing to the location and similar contrast enhancement.² TDL are amenable to treatment, with a very low recurrence rate having underscored the importance of an accurate and early diagnosis of these kinds of lesions. There is a female predominance for the lesion, with the average age of involved individuals being 37 years.² There will not be any history of similar episodes in the past.² The history of an antecedent viral infection or vaccination is uncommon.¹ The most common clinical presentations are a focal weakness, seizure episodes, or, rarely, features of raised intracranial tension.² Seizures progressing into status epilepticus are another common presentation. The patient may also complain of intractable headaches.¹ Examination usually reveals varying degrees of consciousness depending on the severity of mass effect, papilloedema and focal neurological deficits.

Evaluation of patients usually consists of an initial CT scan of head, plain and contrast. The CT appearance of the lesion is a hypodense lesion in the subcortical or periventricular region. The size of the lesions is usually >2 cm. There is minimal contrast enhancement of these lesions. The lesions as a rule show varying degrees of perilesional oedemas, but characteristically this is only minimal.^{1 3}

MRI characteristically shows hypointensities in T1 weighted images with hyperintensities in T2 weighted images. There is a peripheral ring-shaped contrast enhancement seen uniformly with gadolinium contrast. The ring is usually incomplete, opening towards the cortical grey matter.¹ The ring indicates the advancing area of active demyelination, which usually extends along the subcortical and periventricular white matter tracts. Though commonly

seen, the presence of this alone is not enough to conclusively make a diagnosis of TDL.³ Histopathological examination is an imperfect gold standard and the recognition of certain imaging features may facilitate the correct diagnosis.⁴ Imaging features of TDL are a large lesion with little mass effect and oedema,^{1 3} ring-like or open ring enhancement (due to the paucity of active inflammation towards the cortical grey matter),¹ central dilated veins on T2 echoplanar images on MR perfusion,⁵ decreased perfusion (average relative cerebral blood volume <1 in TDL, >2 for lymphomas and >6 for gliomas and rapid resolution of the lesions with steroid).⁵

CONCLUSION

It is prudent for those patients in whom the imageology and clinical profile are not absolutely in favour of a high-grade CNS malignancy, and if the patient and relatives, after fully understanding the risks involved, opt against a biopsy, to be given a course of steroids and be reassessed after a period of 4–6 weeks. If in these patients the repeat image shows a substantial reduction in the size of the lesion, they should be kept under follow-up with frequent imaging during the initial year until the recurrence of lymphoma has been ruled out.

Learning points

- ▶ In TDL, histopathological examination is an imperfect gold standard and the recognition of certain imaging features may facilitate the correct diagnosis.
- ▶ Dynamic contrast-enhanced T2-weighted MR imaging is a useful diagnostic tool in differentiating TDLs from intracranial neoplasms and may therefore obviate unnecessary surgical biopsy.
- ▶ Those patients in whom the imageology and clinical profile are not absolutely in favour of a high-grade CNS malignancy, and if the patient and relatives, after fully understanding the risks involved, opt against a biopsy, should be given a course of steroids and should be reassessed after a period of 4–6 weeks.

Competing interests None.

Patient consent Obtained.

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