LESSON OF THE MONTH

Bilateral enhancing thalamic lesions in a 10 year old boy: case report

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Abstract

A young boy presented with monoparesis of the left arm. MRI disclosed bilateral enhancing thalamic lesions. Biopsy results and subsequent clinical history were most compatible with postinfectious or acute disseminated encephalomyelitis. This represents one of the first cases of acute disseminated encephalomyelitis affecting the thalami, established by biopsy. This uncommon disease entity is reviewed and how it may affect the deep grey matter is described.

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Postinfectious or acute disseminated encephalomyelitis is an acute inflammatory demyelinating disease which can affect the brain or spinal cord. It typically occurs in children but can also affect adults. With advances in immunisation, it commonly follows a non-specific upper respiratory infection in which the aetiological agent is usually undetermined. Occasionally a history of viral illness cannot be obtained.¹ The clinical syndrome is variable but can range from focal neurological deficits to coma and death. The diagnosis is often confirmed by MRI, which shows multifocal white matter lesions corresponding to the abnormal neurological findings.¹⁻⁴ Grey matter lesions are uncommon.

We describe a 10 year old boy who presented with left arm weakness. MRI showed bilateral thalamic gadolinium enhancing lesions. Biopsy results and subsequent clinical follow up were most compatible with postinfectious encephalomyelitis.

Case report

This 10 year old, right handed boy presented with a 1 week history of weakness in the left arm. Although the parents were poor historians, they denied any history of fever, chills, headache, nausea and vomiting, or diarrhoea. Examination showed pyramidal type weakness and a pronator drift of the left arm. MRI showed bilateral gadolinium enhancing thalamic lesions (fig 1). Analyses of CSF including viral cultures were normal. Tissue antibody profile was negative. The patient eventually

Figure 1 T1 weighted gadolinium enhanced coronal images showing bilateral enhancing thalamic lesions.

underwent MRI guided stereotactic biopsy. The preoperative diagnosis was possible multifocal thalamic glioma. This showed an intense macrophage response and focal minimal perivascular cuffing with lymphocytes, plasma cells, and macrophages, most consistent with postinfectious encephalomyelitis (fig 2). Bacterial, tubercular, fungal, and viral cultures were negative. No treatment was instituted. At 3 months of follow up, the patient was neurologically normal and the MRI lesions had partially resolved although still enhanced with gadolinium. At 1 year of follow up, MRI (fig 3) disclosed partial resolution of the lesions with no enhancement with gadolinium and the patient remained neurologically normal. On further questioning, the only other history obtained was that the patient had had frequent episodes of otitis media.

Discussion

Postinfectious encephalomyelitis is an uncommon disease. In most patients it follows a viral illness but has been reported after bacterial

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Figure 2 Haematoxylin and eosin stained section showing perivenular lymphocytes (long arrow) and intense macrophage (open arrow) infiltration. ($\times 200$)

Figure 3 One year follow up T1 weighted gadolinium enhanced coronal image shows a corresponding area of encephalomalacia in the left thalamus that did not enhance and resolution of the right thalamic enhancement.

infection, immunisations, and drug and serum administration.⁵⁻⁷ It is known by various names including acute disseminated encephalomyelitis, acute perivascular myelinoclasia, and allergic or immune mediated encephalomyelitis.⁶

With the advance in immunisation, it usually follows non-specific upper respiratory infections in developed countries; in underdeveloped regions it is most commonly associated with measles. The frequency and severity of the

illness is variable. For example, rates after smallpox vaccination vary from 1:63 in the Dutch experience to 1:300 000 in the United States.⁶

The clinical findings are non-specific but the diagnosis is usually straightforward if the disease follows an exanthem or vaccination but may be confusing if a history of a preceding viral illness is not obtained. Typically, postinfectious encephalomyelitis is characterised by an abrupt onset of fever, obtundation, seizures, and focal neurological signs.⁶ Obtundation may progress to coma and up to 20% of patients die. It is thought of as a childhood illness but may occur in adults. Postinfectious disseminated encephalomyelitis is a diagnosis of exclusion. Other diseases that may present with similar findings are multiple sclerosis, multiple embolic infarctions, vasculitis, deep cerebral venous thrombosis, and multifocal tumour.^{4-5 8} The results of lumbar puncture are variable. Normal CSF has been reported although often there are increases in protein and cells (usually lymphocytes). Oligoclonal IgG bands may be present. Treatment has typically been supportive care or steroids. Many patients have shown rapid improvement with prednisone.

The results of both CT and MRI have been reported. CT is often normal although it may show non-specific changes in white matter. MRI is almost always abnormal; typically multifocal white matter lesions are seen.¹⁻³ Gadolinium enhancement may be seen in some of the lesions.⁹ Many of the lesions resolve on follow up scans.

The pathological findings of postinfectious encephalomyelitis typically show areas of perivascular demyelination with intense infiltration by lymphocytes and macrophages. Perivenular inflammation consisting of lymphocytes, macrophages, and plasma cells is a

common finding. The extent of inflammation correlates with the duration of the disease. As the lesions become older, the macrophages increase and lymphocytes decrease in number. Foci of fibrillary gliosis can also be seen in the adjacent brain tissue. These findings are similar to experimental allergic encephalomyelitis, an experimental model induced by exposing animals to a myelin antigen such as myelin basic protein.⁶⁻¹⁰ Although postinfectious encephalomyelitis has generally been considered a demyelinating disease, in a careful pathological study of 30 cases, perivenular demyelination was found in only six.¹¹ The most prominent histopathological features were lymphocytic infiltration of the meninges and perivascular infiltrates of the single or mixed cell types and perivascular necrosis, vasculitis, and glial nodules in the grey matter.

Although postinfectious encephalomyelitis typically involves the white matter, grey matter lesions have been reported in similar settings and presumed to be a form of postinfectious encephalomyelitis although usually not proved by biopsy. These lesions typically occur in children with acute onset of neurological dysfunction after a febrile illness. They have been reported in the basal ganglia, in the thalamus, and in cortical grey matter. Cusmai *et al* described two children with bilateral reversible selective thalamic involvement and acute severe neurological dysfunction with favourable outcomes.¹² Both followed a febrile illness and the authors considered that the pathophysiology was probably similar to that seen in postinfectious encephalomyelitis, although not established by biopsy. Nagai *et al* described four children with symmetric thalamic lesions seen on CT after influenzae A virus infection, two with Reye's syndrome and two without.¹³ All of these children had severe neurological deficits on presentation, some of which cleared and others that did not. Donovan and Lenn described two children with the acute onset of extrapyramidal signs in whom MRI showed bilateral basal ganglia involvement, which the authors considered were most compatible with postinfectious encephalomyelitis.¹⁴ Ohtaki *et al* described two children with post Japanese B encephalitis vaccination, who developed postinfectious encephalomyelitis with white matter lesions and bilateral thalamic lesions which responded to prednisone.¹⁵

We think that our patient represents a case of postinfectious encephalitis involving the thalami, who presented with a focal neurological deficit without evidence of systemic findings such as fever or disturbance in sensorium. MRI disclosed bilateral enhancing thalamic lesions somewhat similar to the lesions described by Cusmai *et al* in their report.¹² Gadolinium

enhancement was not done in their patients. They hypothesised that the disease was similar to postinfectious encephalomyelitis. Our patient's biopsy showed intense macrophage reaction and minimal perivenular inflammatory cell infiltration with negative cultures, most compatible with postinfectious encephalomyelitis. Although postinfectious encephalomyelitis has traditionally been thought to involve primarily the white matter, we think that a similar disease process probably occurs in the grey matter. This case report represents the only biopsy established case of postinfectious encephalomyelitis involving the thalami that we were able to find in our literature search. The clinical presentation is probably broad, from focal neurological deficits to coma. We think that this disease entity should be considered in the differential diagnosis of bilateral deep grey matter lesions occurring in children. If a clear history of preceding viral illness or immunisation can be obtained and other findings are negative, these children can probably be treated with steroids and biopsy should be reserved for those children in whom the diagnosis remains in doubt.

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