Decompressive aspiration in myelinoclastic diffuse sclerosis or Schilder disease

Case report

FARIDEH NEJAT, M.D., AND BEHZAD EFTEKHAR, M.D.

Department of Neurosurgery, Children’s Medical Center, and Department of Neurosurgery, Sina Hospital, Tehran University of Medical Science, Tehran, Iran

This 9-year-old girl with rapidly progressive cerebral demyelinating disease presented with hemiplegia and intracranial hypertension. Brain images revealed four lesions with mass effect in the subcortical white matter of both hemispheres. Demyelination was found on pathological studies of these lesions. The patient experienced some recovery with corticosteroid treatment but improved completely with decompressive aspiration of the largest lesion.

KEY WORDS • diffuse sclerosis • demyelinating disease • Schilder disease • corticosteroid therapy • aspiration

MYELINOCLASTIC diffuse sclerosis or Schilder disease is a rare acute or subacute demyelinating disorder, affecting mainly children. This disease seems to be immune mediated, identical to MS. There is no evidence of viral agents in biopsy and autopsy materials obtained in cases of Schilder disease. The cerebral lesions tend to be large and bilateral, although they are not necessarily symmetrical. In this report we describe the clinical and neuroimaging findings in one patient and our management protocol for this disease.

Case Report

History. This 9-year-old girl was admitted to the hospital for right hemiplegia, persistent vomiting, drowsiness, and headache; her illness was not accompanied by fever. Her medical history was unremarkable except for two episodes of generalized tonic–clonic seizures, one of which occurred approximately 3 years before and one several months before admission. Four weeks before admission, she experienced progressive right hemiparesis, vomiting, and headache. One week before admission, drowsiness, right hemiplegia, and persistent vomiting developed.

Examination. On admission, the patient was drowsy, afebrile, and her neck was supple. Results of the general examination were normal. The neurological examination revealed bilateral papilledema, right hemiplegia, upward plantar reflexes, and hyperreflexia. Results of routine blood tests, coagulation studies, and evaluation for vasculitis were completely normal. Brain CT scans revealed four large hypodense areas in the white matter of both hemispheres (frontal, parietal, and occipital lobes) with mild mass effect. Postcontrast brain CT scans demonstrated minimal marginal enhancement. Admission MR imaging demonstrated the same lesions, which were hypointense on T1-weighted MR images and hyperintense on T2-weighted MR images, with a midline shift from left to right and a discrepancy between the size of the lesions and the associated mass effects (Fig. 1). A lumbar puncture was not performed because of the midline shift and symptoms of intracranial hypertension.

Treatment. Laboratory tests for immunodeficiency and serum fatty acid levels yielded normal results. Treatment with 2 mg/kg/day dexamethasone was started intravenously, and a slight recovery occurred after several days. One week later local anesthesia was applied and a brain biopsy sample was obtained from the largest lesion in the left occipitoparietal region. The meninges were found to be normal. We removed 60 ml of clear fluid by aspiration from the cavity of the largest lesion. Cytological investigation of the fluid and pathological findings in the brain biopsy material were descriptive of a demyelinating lesion. Histological examination revealed extensive sharply demarcated areas with numerous macrophages and perivascular lymphocytic infiltration (Fig. 2).

Postoperative Course. Several hours after surgery, the patient had no neurological deficit and dramatic recovery occurred. She was discharged without neurological deficits and was given a tapered dose of dexamethasone. Three months later follow-up brain MR images demonstrated

Abbreviations used in this paper: CT = computerized tomography; MR = magnetic resonance; MS = multiple sclerosis.
marked resolution of the lesions (Fig. 3). After 2 years, the patient is well with no neurological deficit.

Discussion

Schilder disease usually affects children between the ages of 5 and 14 years. The most common manifestations are acute hemiplegia with headache, vomiting, behavioral deterioration, and ataxia. The clinical examination often reveals hemiplegia, aphasia, ataxia, and papilledema.2,5

As discussed by Poser, et al.,6 because the original description was published in 1912 by Schilder, several criteria have been applied for this condition. Poser, et al., established restrictive diagnostic criteria for true Schilder disease, as follows: 1) clinical symptoms and signs that are often atypical for the early course of MS; 2) cerebrospinal fluid normal or atypical for MS; 3) bilateral large areas of demyelination of cerebral white matter; 4) no fever, viral or mycoplasmal infection, or vaccination preceding the symptoms; and 5) normal serum concentration of very long-chain fatty acids.6

Computerized tomography and MR imaging demonstrate large cerebral white matter lesions with ill-defined margins, mild mass effect, variable edema, and variable enhancement at the margins.1,7 Cerebrospinal fluid may be normal or show a mild lymphocytosis or elevated protein and oligoclonal bands.12 The acute or subacute onset of focal neurological signs and intracranial hypertension initially indicate a space-occupying lesion, but CT scanning and MR imaging results exclude this diagnosis and point to a demyelinating process.6 An important test to be included in all cases of Schilder disease is the measurement of very long–chain free fatty acids. Patients with adrenoleukodystrophy have an elevation in the C26 fatty acids and an increase in the C26/C22 ratio and usually have signs of posterior white matter involvement on MR images.5

The diagnostic criteria established by Poser, et al.,6 were intended to end the confusion around the eponym “Schilder disease.” The term Schilder disease now refers to a condition marked by large hypodense lesions on CT scans at onset, demyelination in biopsy samples, and normal ratios of very long–chain free fatty acids excluding adrenoleukodystrophy. There is sometimes cystic degeneration in the area of demyelination in Schilder disease.5,6 Some authors believe that most cases will require biopsy sampling to exclude a neoplastic process.1

Our patient had cystic degeneration in the lesions, especially in the largest one in the left occipitoparietal area from which the biopsy sample was obtained. Within 24 hours after aspiration the patient attained complete recovery from hemiplegia, vomiting, and headache; this rapid recovery may be a consequence of decompression and decreased intracranial pressure. There are reports of large lesions with multiple permanent sequelae.4 We advise biopsy sampling and especially decompressive aspiration of the cystic lesions in suspected cases of Schilder disease to prevent sec-

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**Fig. 1.** Axial T₁-weighted MR images demonstrating multiple large hypointense lesions in cerebral white matter with probable cystic changes. Note the discrepancy between the size of the lesions and the midline shift.

**Fig. 2.** Photomicrograph showing pathological changes in the lesion; lymphocyte infiltration and numerous lipid-laden macrophages are seen. H & E, original magnification × 400.

**Fig. 3.** Follow-up T₁-weighted MR images demonstrating good recovery from the lesions in Fig. 1; no residual lesion or mass effect is seen.
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From this experience we conclude that decompressive aspiration of the largest lesions in this disease may provide an opportunity for recovery with the fewest sequelae.

References


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Address reprint requests to: Farideh Nejat, M.D., 47 Amiri Alley, South Hafez Street, Tehran, Iran 15875-1894. email: nejat@sina.tums.ac.ir.