ORIGINAL RESEARCH PAPER
VOLUME-6 | ISSUE-5 | MAY - 2017 | ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

TUMEFACTIVE DEMYELINATING LESION IN CHILDREN - SCHILDER'S DISEASE

Radiology

Chinmay Mehta Lecturer, Department of Radiodiagnosis, Grant Medical college and Sir JJ group of hospital, Mumbai- India.

Shilpa Domkundwar Head of Department of Radiodiagnosis, Grant Medical college and Sir JJ group of hospital, Mumbai- India.

ABSTRACT

Schilder's disease is a rare sporadic demyelinating disease of the brain. We report a 6year old boy with Schilder's disease who presented with progressive right hemiparesis. Brain magnetic resonance imaging (MRI) showed a single large tumefactive white matter lesion. Based on clinical features and pre and post treatment MRI findings, a diagnosis of Schilder's disease was made. The patient showed dramatic clinical improvement and significant regression of the lesion in response to high-dose pulse steroid.

KEYWORDS:

SCHILDER'S DISEASE, TUMEFACTIVE DEMYELINATION, DEMYELINATION IN CHILDREN, PEDIATRIC HEMIPARRESIS.

INTRODUCTION:

Schilder's disease, first described by Schilder in 1912, also known as myelinoclastic diffuse sclerosis, is a sporadic subacute demyelinating brain disease, usually affects children and results in formation of one or two bilateral large tumefactive plaques (1,2). The clinical presentation is atypical for pediatric multiple sclerosis, and symptoms are usually related to the pressure of a focal mass lesion. Distinguishing tumefactive lesions from other etiologies of intracranial space occupying lesions is essential to avoid inadvertent surgical or toxic chemotherapeutic intervention. Tumefactive demyelinating lesions (TDLs) are defined as large (usually >2 cm) demyelinating lesions mimicking brain tumors; they occur as solitary lesions or as a few separate lesions (3, 4). The magnetic resonance (MR) imaging features suggestive of TDL include large white matter lesions with little mass effect or vasogenic edema (3), incomplete or open-ring enhancement (5,6), vessel-like structures running through the center of lesions on dynamic T2*-weighted images, and low relative cerebral blood volume (7). MR imaging is the most sensitive imaging technique for depicting demyelinating disease. Here we contribute to the understanding of Schilder's disease by presenting the case of a child who presented with progressive right hemiparesis.

CASE REPORT:

A 6 yr old boy presented with gradual onset right upper limb weakness since 1 month. A year prior to the incident, he had similar complains which spontaneously disappeared and hence was not imaged.

Brain CT scan findings (Fig1): A large ill-defined hypo attenuating lesion in left frontal deep white matter and adjacent mild edema and resultant mild mass effect and minimal midline shift to the right. No significant contrast enhancement(Fig1).Differential on the CT scan were large abscess, low grade tumor or tumefactive demyelination and an MRI brain was performed.

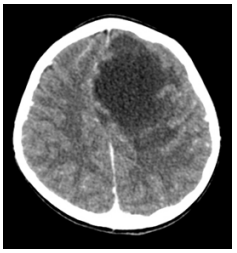

Figure 1: Unenhanced CT brain demonstrating a large illdefined hypoattenuating non enhancing lesion in left frontal deep white matter with mild perilesional edema.

Magnetic resonance imaging (MRI) brain findings(Fig 2-6): showed a large well defined lesion with T2 hyperintense (Fig 2) and T1 / FLAIR (Fig 3)hypointense signal and mild perilesional vasogenic edema in left centrum semiovale and frontal deep white matter. It showed an irregular open ring enhancement(Fig 4). No restriction on diffusion images. There was decreased perfusion on the perfusion weighted images(Fig 5). A demyelinating disease such as multiple sclerosis was suspected, but brain MRI was not typical for multiple sclerosis. Therefore, we diagnosed this patient with Schilder's disease while performing further evaluation. Oligoclonal bands in cerebrospinal fluid (CSF) were negative, and a spine MRI showed no abnormalities. The patient was treated with methyl prednisolone,intravenously for 10 days. Hemiparesis disappeared in a week, and the size of the brain lesion decreased on MRI(Fig 6). He was discharged after the second MRI on oral methyl predinisone, with significant resolution of symptoms.

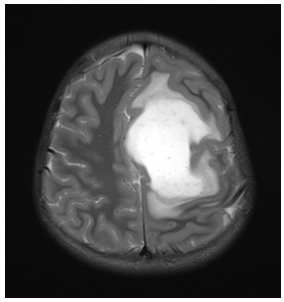

Fig 2- T2W MRI Brain showing well defined hyperintense lesion in left centrum semiovale and corona radiate with mild vasogenic edema.

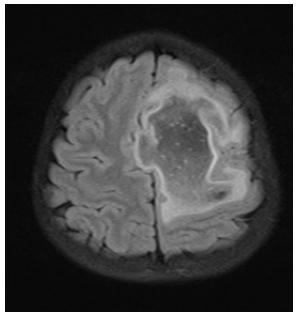

Fig 3-FLAIR MRI Brain showing well defined hypointense lesion with mild vasogenic edema in left frontal white matter.

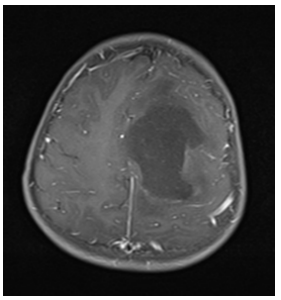

Fig 4-T1W Post contrast MRI Brain showing open ring low enhancement.

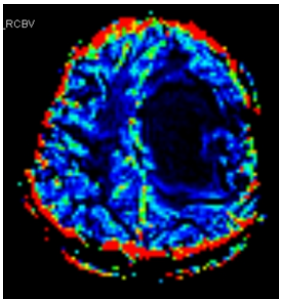

Fig 5-RCBV- PERFUSION MRI Brain showing low relative cerebral blood volume-hypoperfused lesion.

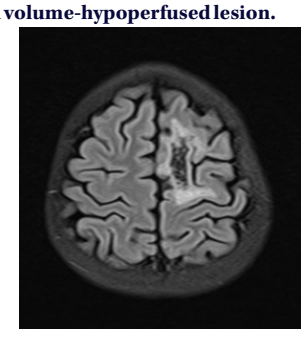

Fig 6-FLAIR – Post treatment showing significant reduction in the size of the lesion and associated perilesional edema.

DISCUSSION:

The diagnostic criteria of Schilder's disease proposed by Poser are as follows (8)- It is a subacute myelinoclastic disorder resulting in formation of one, or more commonly, two roughly symmetric bilateral plaques measuring at least 2×3 cm in two of three dimensions, involving the centrum semiovale. These must be the only lesions demonstrable by clinical, paraclinical, or imaging studies. There must be no peripheral nervous system lesion. Adrenal function and very long-chain fatty acids must be normal. The histopathology must be identical to multiple sclerosis. Our patient fulfills the Poser's criteria, except the absence of histopathology, as brain biopsy was not performed. As suggested in recent studies (9), improved MRI findings of biopsy-proven cases of Schilder's disease have enabled noninvasive diagnosis of Schilder's disease. In our patient, a good response to steroid pulse therapy was also supportive of the diagnosis. MRI in Schilder's disease show a hypointense lesion on T1 weighted images and a hyperintense lesion on T2 weighted images. Open ring contrast enhancement caused by an acute inflammatory process in the white matter is characteristic.(5,6) Other findings are the decreased signal on fluid attenuated inversion recovery (FLAIR) and an increase in the apparent diffusion coefficient (ADC). These radiologic features are also supportive of the diagnosis of Schilder's disease. Our patient had two historical evidences of hemiparesis without any encephalopathy. Differential of multiple sclerosis, low grade glioma and an abscess were considered. Multiple sclerosis was considered based on the 2010 revisions to the McDonald criteria of MS. He had two episodes of clinical attacks and one brain MRI lesion for dissemination in space. However, the single large tumefactive lesion was atypical for multiple sclerosis (Figure 2-4).Low grade glioma was another differential. However, hypodensity on CT scan (Fig1), incomplete rim enhancement, absence of a mass effect, lack of cortical involvement(Fig 2-5) and response on short course of steroid (Fig 6) favored tumefactive multiple sclerosis(10).Abscess was less likely in view of absent restriction on diffusion images and clinical picture.

CONCLUSION

Schilder's disease should be considered in children with acute or subacute neurological symptom, and large tumefactive lesion in MRI. We suggest that when dealing with a patient with atypical clinical and MRI presentation highly suspicious for Tumefactive MS, a short course of steroid therapy and follow up imaging may play a role in clarifying the diagnosis and leading to correct root of therapy. It is an uncommon diagnostic challenge. In such cases, the correct diagnosis is very worthy for eliminating biopsy, unnecessary radiotherapy, and execution of early treatment.

DISCLOSURE Conflict of interest: None

REFERENCES:

1. Schilder P. Zur Kenntnis der sogennanten diffusen Sklerose. Z Gesamate Neurol Psychiatr 1912; 10:1-60.
2. Poser CM, Goutiers F, Carpentier MA, Aicardi J. Schilder's myelinoclastic diffuse sclerosis. Pediatrics 1986; 77:107-12.
3. Dagher AP, Smirniotopoulos J. Tumefactive demyelinating lesions. Neuroradiology 1996; 38: 560–565. CrossRef, Medline.
4. Kepes JJ. Large focal tumor-like demyelinating lesions of the brain: intermediate entity between multiple sclerosis and acute disseminated encephalomyelitis? a study of 31 patients. Ann Neurol 1993; 33: 18–27.
5. Masdeu JC, Quinto C, Olivera C, Tenner M, Leslie D, Visintainer P. Open-ring imaging sign: highly specific for atypical brain demyelination. Neurology 2000; 54: 1427–1433. CrossRef, Medline
6. Masdeu JC, Moreira J, Trasi S, Visintainer P, Cavaliere R, Grundman M. The open ring: a new imaging sign in demyelinating disease. J Neuroimaging 1996; 6: 104–107. CrossRef, Medline
7. Cha S, Pierce S, Knopp EA, et al. Dynamic contrast-enhanced T2*-weighted MR imaging of tumefactive demyelinating lesions. AJNR Am J Neuroradiol 2001; 22: 1109–1116. Medline
8. Poser CM, Goutiers F, Carpentier MA, Aicardi J. Schilder's myelinoclastic diffuse sclerosis. Pediatrics 1986; 77:107-12.
9. Bacigaluppi S, Polonara G, Zavanone ML, Campanella R, Branca V, Gaini SM, Tredici G, Costa A. Schilder's disease: non-invasive diagnosis?: A case report and review. Neurol Sci 2009; 30:421-30.
10. S. K. Dae, G. N. Dong, H. K. Keon et al., "Distinguishing tumefactive demyelinating lesions from glioma or central nervous system lymphoma: added value of unenhanced CT compared with conventional contrast-enhanced MR imaging," Radiology, vol. 251, no. 2, pp. 467–475, 2009