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Case Report

A rare presentation of multiple sclerosis: Schilder's disease☆

Reda Taoussi, MD[a,], Hajar Khattab, MD[b], Youssef Sakhy, MD[a], Aïcha Merzem, MD[a], Hasna Belgadir, MD[a], Omar Amriss, MD[a], Nadia Moussali, MD[a], Naima EL Benna, MD[a]*

[a] *Imaging Department, Ibn Rochd University Hospital, 1 quartier des hôpitaux, Casablanca, Morocco*
[b] *Department of neurology, Ibn Rochd University Hospital, Casablanca, Morocco*

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ABSTRACT

Schilder's disease is a rare form of multiple sclerosis. It concerns mostly teenagers and young adults. The Clinical signs and symptoms might be atypical for early multiple sclerosis which often mimics intracranial neoplasm or abscess. Their course may be either progressive or relapsing and remitting, with a high sensitivity to steroids. The knowledge of this rare form of multiple sclerosis may help radiologists in assessing a precise diagnosis. We report the case of a young 22-year-old patient admitted to the emergency room with an array of headache, vomiting and frontal syndrome. Magnetic resonance imaging shows 2 bilateral demyelinating frontal areas. The patient was put under corticosteroids bolus with discreet improvement in her clinical condition. After 6 months of follow-up, we did not notice any real clinical improvement. Although Schilder's disease is considered to be a variant of Multiple Sclerosis, its clinical and imaging features behaves more like a demyelinating condition with its monophasic course with, however, serious clinical consequences if the treatment is delayed.

Introduction

First described in 1912 by Austrian physician, Paul Schilder, Schilder's disease is a rare form of Multiple Sclerosis (MS) evolving with a monophasic course. Mostly described in children, teenagers and young adults, the histology describes tumefactive demyelinating lesions within the bilateral centrum ovale of the cerebral hemispheres with sharply demarcated borders and almost complete sparing of axons [2]. The particularity of this case is the late clinical diagnosis.

Patient and observation

A 22-years-old female with no significant pathological background consulted for headaches, vomiting, frontal syndrome

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* Corresponding author.
E-mail address: Taoussi.md@gmail.com (R. Taoussi).

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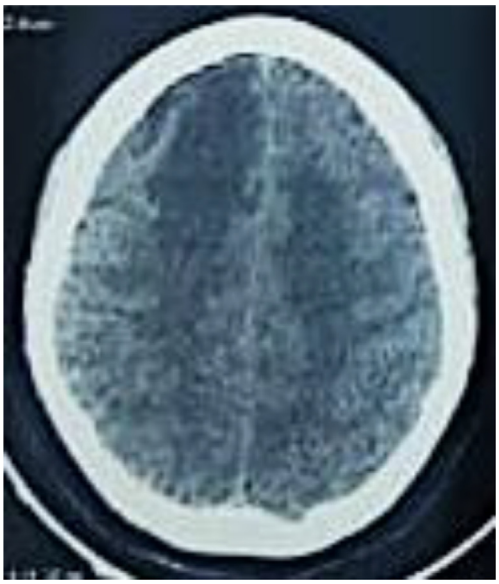

Fig. 1 – Axial Brain CT showing 2 large subcortical hypoattenuating lesions, without involvement of the cortex nor enhancement

and visual loss. She however didn't have any fever nor any patent infectious focus. A Brain computed tomography (CT) was performed showing bilateral frontal hypodensities, without any enhancement (Fig. 1). She was transferred in the infectious department with a diagnosis of meningo-encephalitis. She received 15 days of triaxone and acyclovir without any clinical improvement. A brain Magnetic resonance imaging (MRI) was then performed, showing 2 separate lesions in both frontal lobes, hypointense in T1 weighted sequence, hyperintensity in T2, FLAIR et DWI weighted sequences, a discontinuous ring enhancement and a mass effect on the midline (Fig. 2). The MRI pattern added to the clinical symptoms suggested the diagnosis of a demyelinating affection and in particular Schilder's disease. The patient was transferred to the neurological department where she received 5 days of high doses of corticosteroids.

The examination of the cerebro spinal fluid (CSF) revealed a normal cell count and biochemistry and the absence of oligoclonal bands, moreover, the viral and bacterial cultures were sterile. The few drops taken worsened the patient's neurological condition. At 6 months follow up, we noticed a significative clinical improvement with a near-total disappearance of the previously described bi-frontal lesions.

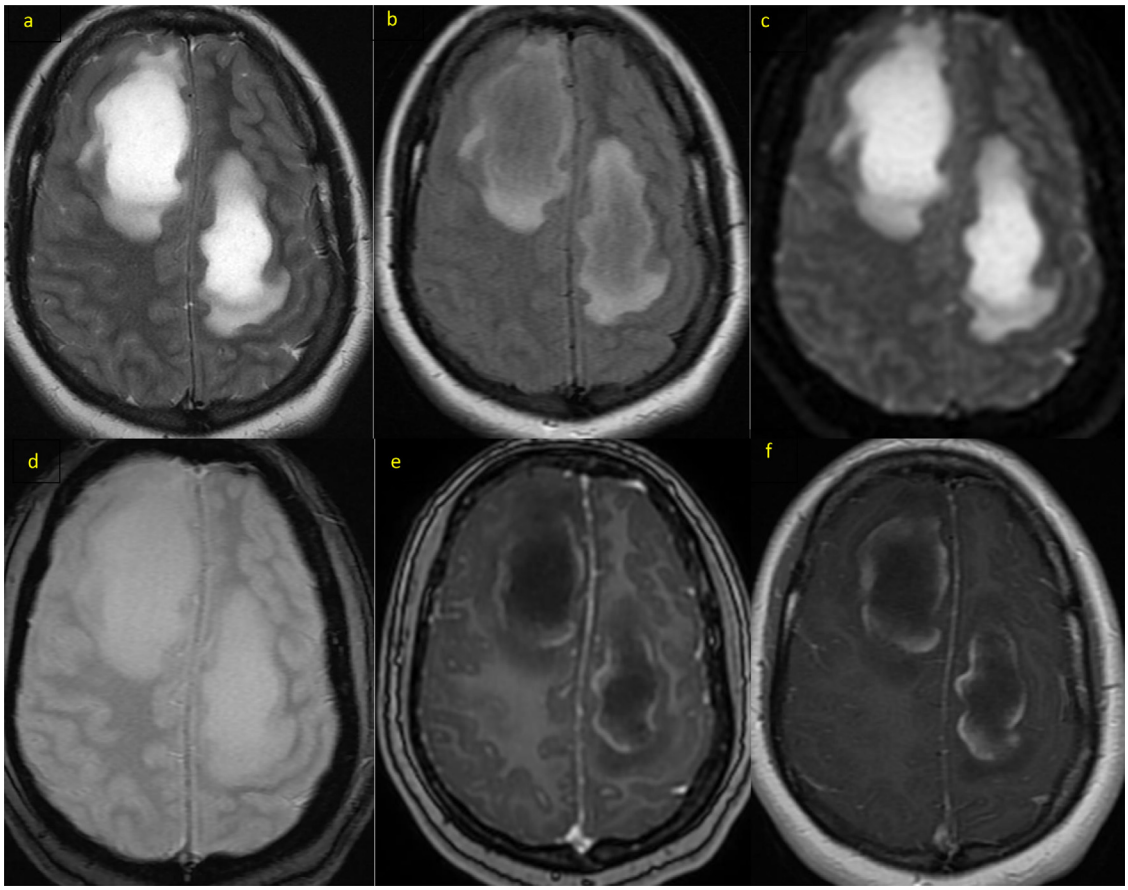

Fig. 2 – Axial Brain MRI before treatment: Large bilateral frontals lesions hyperintense on T2 (A), FLAIR (B) et DWI (C), without any loss of signal on T2* (D). It enhances with a discontinuous ring pattern in gradient T1 MPRAGE (E) and T1 FSE (F)

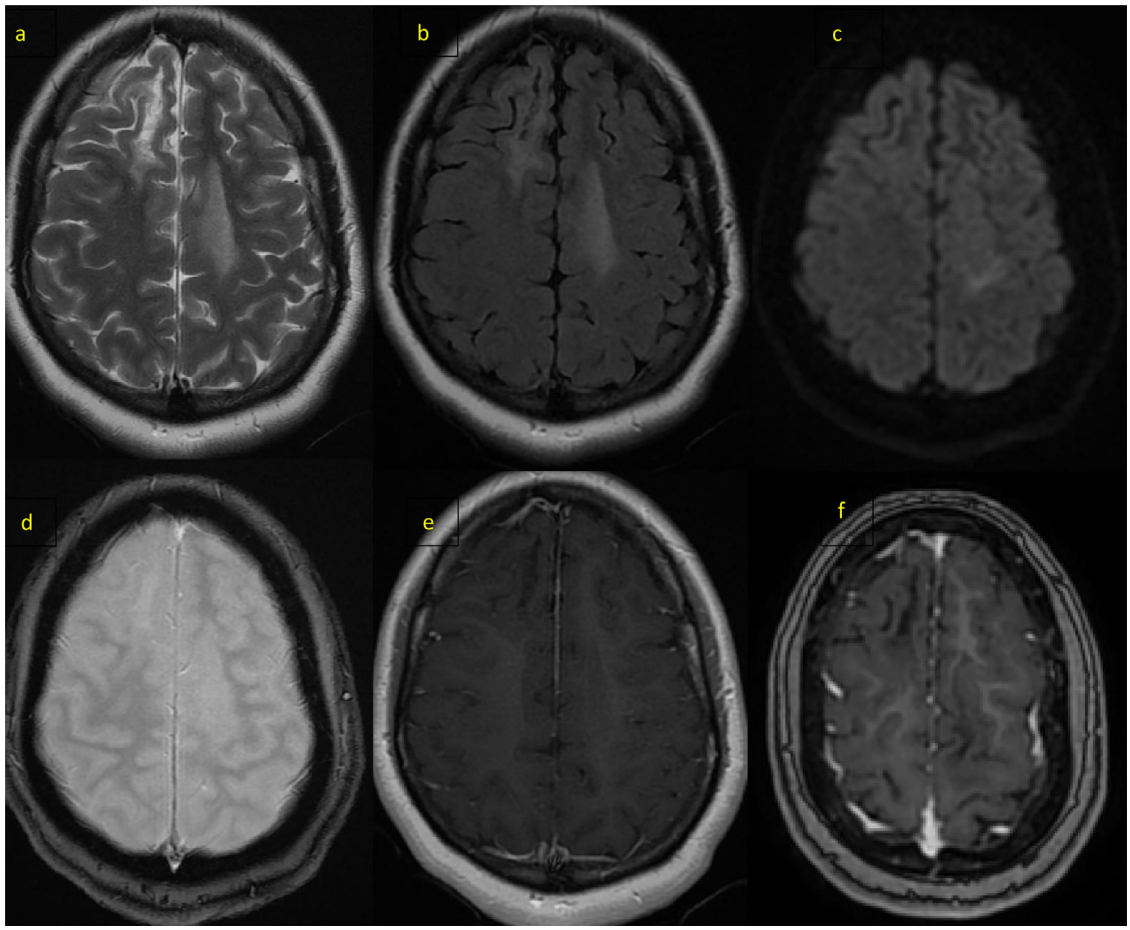

Fig. 3 – Six months follow-up axial Brain MRI after cortico-steroid treatment: Large bilateral frontals lesions hyperintense on T2 (A), FLAIR (B) et DWI (C), without any loss of signal on T2* (D). It enhances with a discontinuous ring pattern in gradient T1 MPRAGE (E) and fast-spin echo T1 FSE (F).

Discussion

Schilder disease, described as an encephalitis *periaxialis diffusa* for 3 cases in 1912, represents a rare monophasic form of MS with less than 10 cases reported in the literature that fulfil the original Poser diagnostic criteria [3]. It's a demyelinating affection mainly touching the children and young adults, affecting both genders. In 1986, Poser et al. suggested a list of criteria aiming to ease the radiological diagnosis and help for the early therapy of the patients [4]. Those criteria distinguish myelinoclastic diffuse sclerosis from adrenoleukodystrophy in which Charles Poser considered the former to be "Schilder's Disease." In fact, he was the 1 describing both affections in 1986 [5]. As we have seen in our case, the diagnostic's delay resulted in a bedridden state of the patient which could have been avoided. Those imaging criteria are:

- Clinical symptoms and signs often typical for the early course of MS.
- CSF normal or atypical for MS.
- Bilateral large areas of demyelination of cerebral white matter measuring at least 2 × 3 cm with a discontinuous ring enhancement commonly involving the frontal or parietal lobes. Those lesions may be bridged by abnormal signal in the corpus callosum [6]. A literature review by Dunn Pirio et al. in 2018 found a case of a 6-years-old patient who had unilateral hemispheric involvement at presentation, which is not as classically observed in Schilder's disease [5]. Typically, the cerebral cortex, cerebellum, brainstem, and spinal cord are relatively spared [7].
- No fever, no viral or mycoplasma infection or vaccination preceding the neurologic symptoms
- Normal adrenal function and serum concentration of very-long-chain fatty acids.
- Histological characteristics on brain biopsy are consistent with MS.

The clinical findings are not typical of a MS attack and no optic nerve or spinal cord involvement was described in the literature yet. The findings are usually an acute or subacute onset, disturbed neurologic symptoms, headache, vertigo, vomiting, seizure and diplopia [2]. Yet, mental deterioration and confusion may occur like for our patient [1]. Unlike adrenoleukodystrophy, it does not involve the peripheral nervous system, and patients have normal adrenal function and serum VLCFA content. The brain biopsy, if performed demon-

strated an inflammatory pathology with associated demyelination [5].

The clinical course may be either progressive or relapsing and remitting. Although recovery has been reported, the long term outcome is uncertain [1].

The mainstay of treatment is high-dose corticosteroids, although evidence-based data regarding their efficiency in children is obviously missing owing to the rarity of this disorder. In 2012 Kraus et al. proposed a treatment based on intravenous immunoglobulins (IVIg). It has been proposed to treat SD patients with IV Immunoglobulins prior to high-dose CS treatment, mainly due to the relative safety and limited side effects of high-dose and/or long-term CS treatment [8].

The main limit of this case was the lack of histological proof on brain biopsy, not done usually as very invasive.

Conclusion

While being a rare variant of Multiple Sclerosis, the clinical findings of Schilder's disease are not specific, thus the diagnosis may be delayed. The imaging criteria by MRI suggested by Poser et al. may help radiologists to orientate the diagnosis. Knowledge of this variant allows the establishment of early therapy as well as a better prognosis and rapid rehabilitation of neurological functions. Bolus corticosteroid therapy is the rule, although new avenues are being studied such as immunoglobulins in the future.

Authors' contributions

All the above authors contributed on the writing of this manuscript, the lecture of the imaging studies, or the care of the patient during his hospitalization.

Patient Consent

Written, informed consent for publication of their case was obtained from the patient.

Declaration of Competing Interest

The authors have no conflicts of interest to disclose.

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