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Autoimmune Encephalitis: A Case Report from the Neurology Department of the Doctor Joseph Guislain Neuropsychiatric Centre in Lubumbashi and Review of the Literature

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ABSTRACT

The term encephalitis refers to the inflammation of the parenchyma of the central nervous system, located in the cranial cavity. We report the case of a 17-year-old female patient with a history of tuberculous gonarthritis, who was admitted for convulsive seizures complicated by status epilepticus. Medical imaging did not reveal any brain lesions and biology revealed an elevated PCT. Through this case, we underline the risk of diagnostic errancy and thus of delay in the treatment of autoimmune encephalitis which, in 70% of cases, is effective. It also seemed important to us to insist on the fact that the absence of autoantibodies does not invalidate the diagnosis of autoimmune encephalitis, especially in developing countries where the technical platform to search for them is lacking both in terms of equipment and human resources. Finally, it should be noted that new autoantibodies are found almost every year by teams in developed countries. Thus, in front of any acute or sub-acute neurological picture not explained, it is necessary to think about autoimmune encephalitis and to start an immunomodulatory treatment, which guarantees the cure or the substantial improvement in most cases.

KEYWORDS

Encephalitis, Autoimmune, Encephalomyelitis, Demyelinating.

Introduction

The term encephalitis refers to inflammation of the central nervous system parenchyma located in the cranium [1]. The term acute disseminated encephalomyelitis is broader and takes into account the possible participation of the medulla in the pathological process, the primum movens of which may be unknown and the pathophysiological mechanisms imperfectly known at present [2]. Acute disseminated encephalomyelitis (ADEM) is defined as a multifocal inflammatory demyelinating disease mainly involving the white matter of the CNS. It is mediated by an autoimmune mechanism and usually develops following an infection or vaccination [3]. Many names are used to describe the same syndrome: post-infectious encephalitis, post-vaccinal encephalitis or perivenous encephalitis [4]. Several groups of autoimmune encephalitis are described depending on the nature of the auto-Ac. Some clinical presentations are specific to auto-Ac and the severity of symptoms varies according to the subtypes of encephalitis.

Recently discovered, anti-N-Methyl-D-Aspartate Receptor (NMDA-R) encephalitis appears to be the most common

autoimmune encephalitis [5]. It, therefore, seemed interesting to us to publish this case for which the presumptive diagnosis is relevant and the immunomodulatory treatment effective, even though no antibodies could be individualized in the current state of our technical platform.

Clinical Observation

We report the case of a young17-year-oldd female patient with a history of gonarthritis extra-pulmonary tuberculosis treated for 1 year with ATB (anti-tuberculosis antibiotics) who presented with myoclonus, attitude dystonia, instability in standing and walking, dysarthria and cognitive disorders (13/30 on the MOCA scale) after a state of epilepsy treated with clonazepam and levetiracétam, instability when standing and walking, dysarthria and cognitive disorders (13/30 on the MOCA scale). The neurological examination confirms the previous signs associated with predominantly axial hypotonia. In view of this picture suggestive of encephalitis, an MRI was normal as were the EEG and pelvic ultrasound, eliminating the possibility of an ovarian teratoma. Thoracic-abdominal CT did not reveal a possible neoplasm. Serum analysis did not reveal any autoantibodies. The standard biological workup showed a hyper leukocytosis with neutrophilic polynucleotides which normalized in a few days under broad spectrum antibiotics while the CRP remained normal; only the PCT was increased to 2.7. As the LP was traumatic, its results were not retained.

The diagnosis of encephalitis was then retained but the infectious proof could not be formally confirmed (inconclusive LP, normal MRI). The autoimmune cause was then retained despite the absence of autoantibodies; this element did not however invalidate the autoimmune cause.

Treatment with corticosteroids (Methylprednisolone IV 1 g per day for 5 days) led to a spectacular improvement in symptomatology but with a resumption of signs after a few weeks. Renewal of the treatment proved to be effective again, but we maintained treatment with hydrocortisone 200 mg in the morning for 5 days, then with decreasing doses of 20 mg every 5 days and currently with Methylprednisone 16 mg in the morning for 7 days with efficacy maintained even after a therapeutic window of 2 weeks, which supports the autoimmune hypothesis. We plan to renew the treatment in case of a resumption of the symptomatology while waiting to have a more durable result with the addition of an immunomodulator; Immunoglobulins and plasmapheresis not being available. We also raise the question of the appropriateness of maintenance treatment and its duration in certain cases.

Discussion

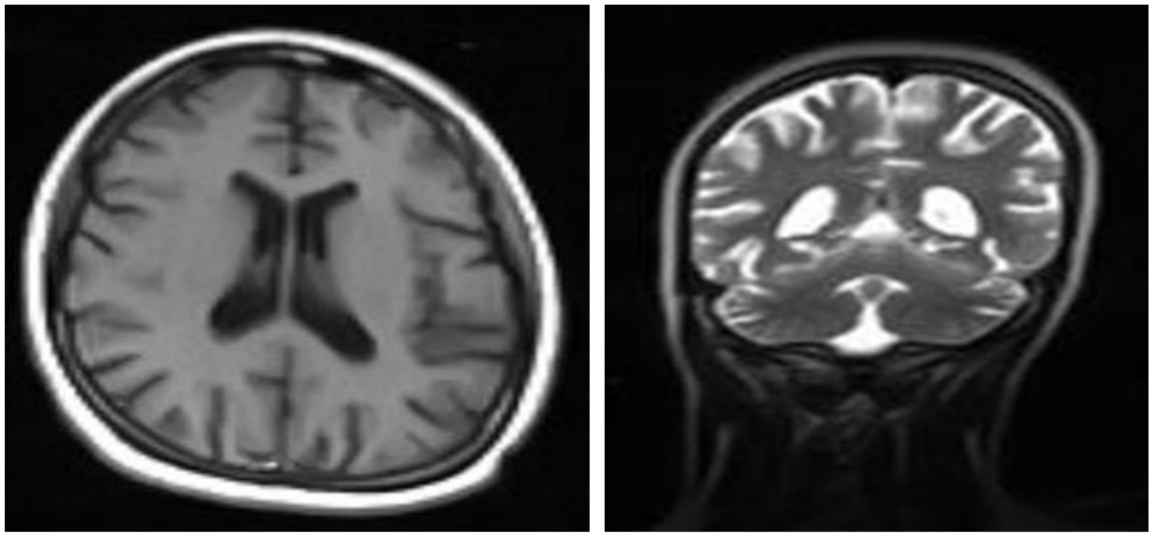
Our work consisted of a clinical and paraclinical study of a 17 -year-old female patient admitted to the Saint-Joseph neuropsychiatric centre in Lubumbashi for autoimmune encephalitis.

In 2005, Dalmau's team described a limbic encephalitis picture associated with an ovarian teratoma in 5 patients, raising suspicion of a paraneoplastic syndrome [6]. Two years later, the same team discovered the corresponding auto-Ab in 12 young women with ovarian teratoma [6]. It is worth noting that this discovery led to the identification and treatment of a significant number of patients with limbic encephalitis.

Epidemiologically, ADEM is more common in children than in adults [7]. Its incidence in children is estimated at 0.4/100,000 per year. No data are available for adults. A male preponderance has been reported in several paediatric cohorts with a female/male sex ratio of between 0.6 and 0.8. This trend is less clear in adults (the observed sex ratio is 1.3 to 1.7 [7].

Infection or vaccination are the most common triggers. These are found in about 75% of cases in children and in 45-50% of cases in adults

Historically, post-infectious encephalitis following exanthemata's rashes accounted for one-third of encephalitis from all causes [4]. The geographical distribution of ADEM in the world is not



Figures 1: Brain MRI axial section T1A sequence and coronal section T2 B sequence showing normal signal of all brain structures.

precisely known. It is thought to be more common in developing countries, where vaccination coverage is inadequate [9]. A recent epidemiological study in the United States [10] showed that the incidence and prevalence of infectious and autoimmune encephalitis are equivalent: 0.8/100,000 person-years and 13.7/100.

Autoimmune encephalitis can be triggered by the abnormal expression of an antigen by a tumour or by a viral infection, but in most cases, the origin of the appearance of the auto-Ac is unknown. In our patient, the paraclinical work-up (MRI, ECG, and biology) did not allow us to retain any other aetiology than the autoimmune cause, especially since the immunomodulatory treatment proved to be effective. It is also known that seronegativity does not invalidate the diagnosis of autoimmune encephalitis [11]. Moreover, the morbidity and mortality are much greater than the possible risk of treatment, the precocity of which guarantees a cure in 75% of cases. Finally, mortality is much higher in untreated patients [11]. The same study showed that rapid diagnosis and treatment with immunomodulator intravenous Immunoglobulins, high-dose corticosteroids and plasmapheresis are associated with good results. The same is true for the sequelae. Our case shows this.

A prospective study in France found 48% of patients with no evidence of autoimmunity [12]. This was also demonstrated by Picard and Mailles in England in a prospective multicentre study of the causes of encephalitis and the different clinical presentations published in October 2010 which reported 38% of patients for whom the cause was unknown and among the non-infectious causes in a sample of 42 patients, 9 or 21% expressed NMDA receptor antibodies and 7 expressed VGKC antibodies in their sera.

The most frequent aetiology is viral, in particular the herpes virus which is a diagnostic and therapeutic emergency and represents 19% of the causes of encephalitis [13]. Moreover, a recent prospective epidemiological study showed that encephalitis with anti-R-NMDA antibodies represented 4% of all causes of encephalitis, i.e. the fifth cause of encephalitis after herpes simplex virus infections, Varicella zoster virus (VZV), Mycobacterium tuberculosis Acute disseminated encephalomyelitis (ADEM) is the second most common autoimmune encephalitis [3].

The clinical presentation differs between children and adults. The clinical manifestations often occur in a febrile context and combine disorders of consciousness, focal deficits and/or convulsions [14]. All kinds of neurological syndromes have been described, from central involvement to psychiatric symptoms, seizures, cognitive disorders, motor disorders (deficits or abnormal movements), behavioral disorders, and peripheral involvement.

Encephalitis leads to serious neurological disorders which are fatal in 5% of cases. It is very important to make the diagnosis because early immunomodulatory treatment allows partial or even complete reversibility of the symptoms [6]. The diagnosis of these encephalitides is confirmed by the detection of an antibody directed against a central nervous system (CNS) antigen, either against an intracellular antigen (group I encephalitis) or against a membrane antigen (group II encephalitis), in particular, anti-N-methyl D-aspartate receptor (R-NMDA) antibodies [6].

The evolution is favourable in more than 75% of patients, provided that adequate treatment is put in place. If patients are not diagnosed, they deteriorate more or less slowly and the evolution can lead to disabling neurological sequelae or even death [10].

The therapeutic approach is based on immunomodulatory treatments. The most commonly used treatments are intravenous corticosteroids (CT), polyvalent Immunoglobulins (IVIG) and plasma exchange (PE). Most of the data on these treatments are derived from small patient series or isolated cases reported in the literature. To date, no randomized controlled clinical trials have been conducted in ADEM in children or adults [15].

Conclusion

Following this case of encephalitis in a young 17-year-old patient inaugurated by a status epilepticus, presenting with myoclonus, attitude dystonia, instability when standing and walking, instability when standing and walking, with neurological examination showing predominantly axial hypotonia and dysarthria suggesting cerebellar involvement and cognitive disorders. We emphasize the need for early diagnosis and appropriate treatment without prejudging the presence or absence of autoantibodies when the clinical picture is suggestive and other causes have been ruled out. The diversity of clinical pictures, the need to include in the workup of this disease the search for a neoplasm and the limits of the technical means available to us to search extensively for antibodies that may be incriminated in its occurrence in a context of developing countries where, moreover, the availability of drugs is a problem, remain the limits that we have encountered in the diagnosis and management of this case.

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