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Rasmussen's Encephalitis of the Adult: A Case Report

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Abstract **Case Report**

Rasmussen syndrome (RS) is a chronic inflammatory disease of unknown etiology.it characterized by severe drug-resistant epilepsy, hemicorporeal neurologic deficit and progressive onset cerebral hemi-atrophy. Although it has long been considered a childhood disease, this pathology can appear in adolescence or adulthood. We report the observation of a patient with Rasmussen syndrome in adulthood, through which and the data of the literature we have discussed clinical, electro-radiological and therapeutic aspect of this serious pathology.

Keywords : Rasmussen's encephalitis; Adult; Seizure; MRI.

INTRODUCTION

Rasmussen's encephalitis is a chronic and serious inflammatory disease of the cerebral cortex whose etiology is still unknown. It basic description was given in 1958 by Theodore Rasmussen on the basis of four cases presenting a "chronic encephalitis with epilepsy syndrome" whose anatomopathological characteristics were those of a unilateral chronic encephalitis with progressive cerebral hemiatrophy on the pneumoencephalography [1]. This rare deaseas mainly affects young children. Late-onset forms are much rarer.

CLINICAL CASE

A 22-year-old woman, with no pathological history, admitted for a neurological symptomatology which has been evolving for two months, consisting of focal motor epileptic seizures, multi-daily, of the clonic type of the right foot with, sometimes, extension of these seizures to the whole right half body. Clinical examination on admission showed discreet right hemiparesis predominantly in the lower limb. The osteotendinous reflexes were sharp and diffuse with a Babinski sign on the right. There was a superficial ipsilateral hemi-hypoaesthesia. Higher functions have been retained the electroencephalogram (EEG) showed a slowing down of the background activity and interictal epileptic abnormalities with slow centro-parieto-frontal sharp waves on the left side. These abnormalities sometimes diffuse to the controlateral hemisphere (Figure 1). Encephalic magnetic resonance imaging (MRI) revealed two hypersignal lesions on the T2 and flair sequences, one frontal and one parietal left cortical-subcortical with no associated cortical atrophy (Figure 2-3). Brain MRI spectroscopy showed the presence of a lactate peak with a moderate decrease in NAA in favor of inflammatory lesions. The study of cerebrospinal fluid did not show an oligoclonal profile. The etiological assessment, including an inflammatory workup, viral and bacterial serologies in the blood, an immunological workup and an enzyme conversion test, was normal. In light of these results, the diagnosis of Rasmussen's encephalitis in the acute phase was retained according to the 2005 consensus criteria. She benefited from a bolus of methylprednisolone: 500 mg/day for 5 days followed by oral prednisone at a dose of 1 mg/kg per day, combined with three courses of intravenous human immunoglobulin (0.4g/kg per day for 5 days) 3 months apart. Thus, the seizures were progressively controlled in association with triple antiepileptic therapy (Carbamazepine, Topiramate and Clonazepam). The evolution was marked by the disappearance of focal epileptic seizures with persistence on the cerebral MRI of control a small residual frontal cortical lesion on the left hemisphere. This lesion appears as hypersignal only on the T2 sequence (Figure 4).

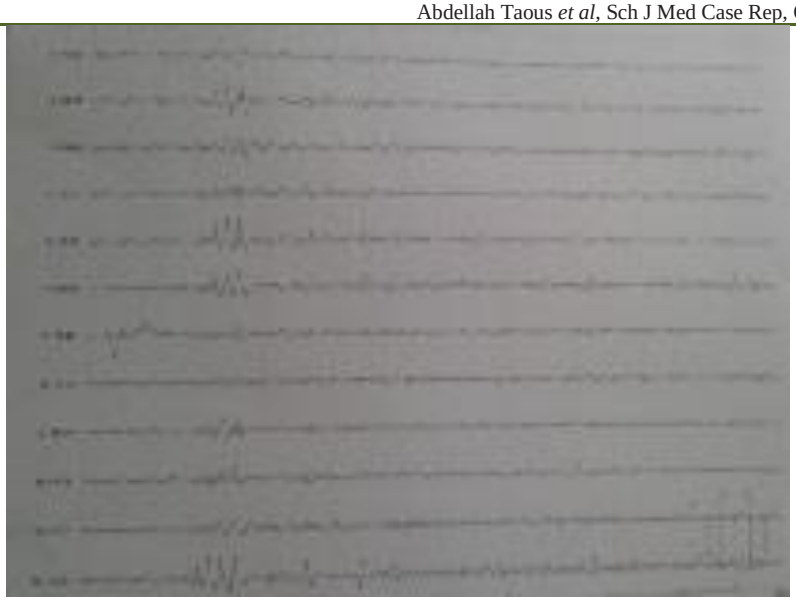

Figure 1: EEG showing slowed background activity and sharp waves in the left centro-fronto-parietal region with diffusion to the contralateral hemisphere

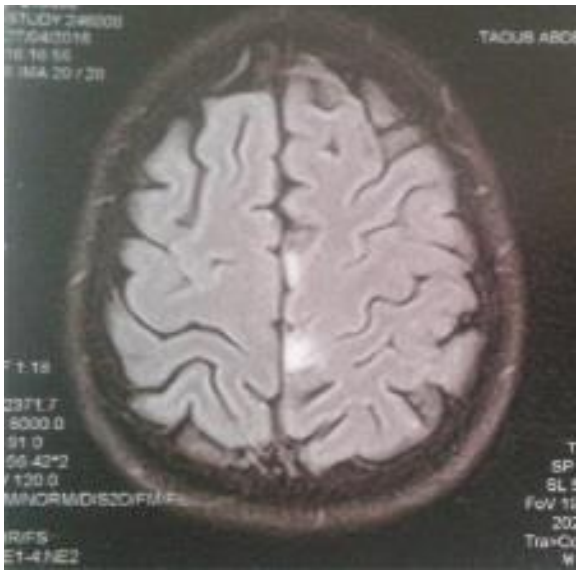

Figure 2: Cerebral MRI axial section in flair weighted sequence: two small left cortical lesions: one frontal para sagittal and the other parietal in hypersignal

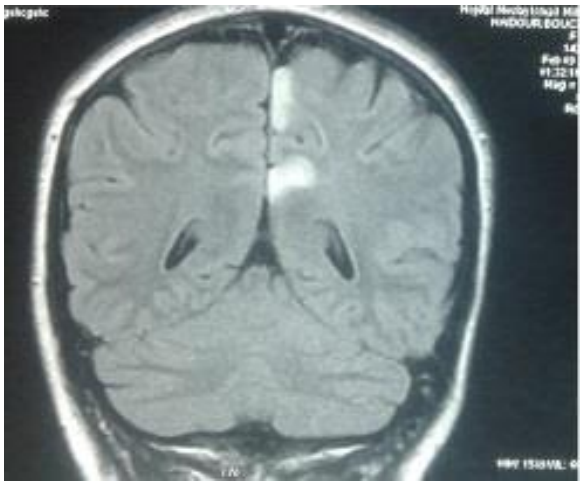

Figure 3 Cerebral MRI coronal section in T2- flair weighted sequence: two small left cortical lesions in hypersignal

Figure 4: Cerebral MRI axial section in T2-weighted sequence: left frontal cortico-subcortical lesion in hypersignal

DISCUSSION

Rasmussen's encephalitis is a severe epileptogenic and inflammatory encephalopathy. It is a rare entity with an estimated incidence of 2.4 cases per 107 persons per year [2]. It mainly affects children with a median age of onset of 6 years. Clinically, it is characterized by a drug-resistant unilateral focal epilepsy with progressive unilateral neurological deficit. In children, it evolves in three stages correlated with radiological and anatomopathological lesions [3-4]: a prodromal stage marked by hemiparesis or infrequent seizures, which can last several years. An acute stage marked by very frequent focal seizures emanating from a single hemisphere, most often focal motor seizures without alteration of consciousness with, in 50% of cases, a continuous partial epileptic picture very suggestive of the disease and a residual stage marked by a severe fixed deficit, most often motor and cognitive with persistence of drug-resistant epilepsy. Brain MRI is essential for diagnosis, it presents different aspects depending on the evolutionary stage. MRI may be normal or show a non specific widening of the ventricular system. In the acute stage, lesions are mainly observed in the cortical or subcortical regions, in particular the caudate nucleus. On the T1-weighted sequences, these lesions appear as hyposignal which are not enhanced after injection of gadolinium and as hypersignal on the T2 / FLAIR-weighted sequences. With progress, consecutive MRIs will show an extension of the hyper signal which will disappear, giving way to cortical atrophy [5]. Alongside these classic infantile forms of S R, late onset forms exist but are rarer and are distinguished by some specificities in their clinical presentations and evolutionary profiles [1]. Epidemiologically, the incidence of the late-onset forms remain low (37 cases reported in the literature). When it begins after the age of 13, the disease seems to preferentially affect women (sex ratio estimated at 3.3). The average age of the onset of the disease is 24 years (13 to 48 years). Clinically, reported cases in children begin with epilepsy often including partial polymorphic seizures, subsequently developing continuous partial epilepsy in more than half of cases over a varying period (ranging from 0 to 10 years). Classically, a motor deficit appears in the three years following the onset of epilepsy [6-7]. In adults, the clinical presentation appears even more polymorphic. Cases of continuous partial epilepsy account for only 56 p. 100 of the cases at the literature. The presence of a motor deficit is found in 75% of patients with a late onset of the disease, whereas it seems to be constant in the classic early onset forms. Visual seizures, reflecting possible occipital localization of lesions, appear to be more frequent in late-onset forms of RS [7]. On the other hand, atypical presentations can be noted, either with the presence of abnormal movements or with a symptomatology evocative of temporal lobe epilepsy. In terms of the evolution of the disease, the study of 37 cases reported in the literature supports a slower and less severe course in adults [1, 8, 9]. The average duration of neurological deterioration is estimated at 11.6 years, compared to 5.3 years in the Montreal Neurological Institute series that studied 48 early-onset SRs [7]. Electrical and radiological data may also be different in late onset forms. Typically, in early-onset RS, there is an EEG aspect associating a unilateral and progressive slowing of the basal rhythm and multifocal ictal and interictal discharges within a single hemisphere. It appears that in adults these EEG abnormalities can become bilateral [10, 11]. With regard to neuroradiological abnormalities, we note either an aspect identical to that encountered in early-onset forms [12] or focal atrophic aspects affecting the temporal lobe with possibilities of an aspect of hippocampal sclerosis. Anomalies of the basal ganglia, in particular atrophy of the head of the caudate nucleus, are found in both early and late-onset SRs and therefore do not represent a distinctive criterion. The exact etiology of Rasmussen's encephalitis is still unknown. Several pathophysiological hypotheses have been proposed to explain this disease, but the autoimmune hypothesis remains at present the most suspected, however, until now no specific autoantibodies have been detected [13].

Treatment of Rasmussen's encephalitis relies on anti-epileptic drugs, which are most often ineffective in suppressing seizures and deficit. Immunomodulatory and immunosuppressive treatments, prescribed in the acute phase, allow seizure control and slowing the progression of the deficit [6] or even disappearance of seizures in certain published cases of late onset [14] like in our case. As for surgery, it is indicated in children in the residual phase when the deficit becomes severe and fixed. For later onset cases, focal resections give good results in terms of seizure control [1]. However, until now, due to the lack of large-scale studies, no therapeutic strategy has been clearly defined.

CONCLUSION

Late-onset Rasmussen's encephalitis is a rare entity and must be known by neurologists. Its early diagnosis at the inflammatory stage is of great therapeutic interest as it allows the initiation of an immunomodulatory therapy that prevents the appearance of definitive brain lesions with lasting neurological sequelae and avoids resorting to surgery.

REFERENCES

1. Hart, Y. M., Andermann, F., Fish, D. R., Dubeau, F., Robitaille, Y., Rasmussen, T., ... & Scaravilli, F. (1997). Chronic encephalitis and epilepsy in adults and adolescents: a variant of Rasmussen's syndrome?. *Neurology*, *48*(2), 418-424.
2. Bien, C. G., Tiemeier, H., Sassen, R., Kuczaty, S., Urbach, H., von Lehe, M., ... & Kruse, B. (2013). Rasmussen encephalitis: incidence and course under randomized therapy with tacrolimus or intravenous immunoglobulins. *Epilepsia*, *54*(3), 543-550.
3. Oguni, H., Andermann, F., & Rasmussen, T. B. (1992). The syndrome of chronic encephalitis and epilepsy: a study based on the MNI series of 48 cases. *Advances in neurology*, *57*, 419-433.
4. Varadkar, S., Bien, C. G., Kruse, C. A., Jensen, F. E., Bauer, J., Pardo, C. A., ... & Cross, J. H. (2014). Rasmussen's encephalitis: clinical features, pathobiology, and treatment advances. *The Lancet Neurology*, *13*(2), 195-205.
5. Pradeep, K., Sinha, S., Saini, J., Mahadevan, A., Arivazhagan, A., Bharath, R. D., ... & Shankar, S. K. (2014). Evolution of MRI changes in Rasmussen's encephalitis. *Acta Neurologica Scandinavica*, *130*(4), 253-259.
6. Granata, T., Fusco, L., Gobbi, G., Freri, E., Ragona, F., Broggi, G., ... & Vigevano, F. (2003). Experience with immunomodulatory treatments in Rasmussen's encephalitis. *Neurology*, *61*(12), 1807-1810.
7. Oguni, H., Andermann, F., & Rasmussen, T. B. (1992). The syndrome of chronic encephalitis and epilepsy: a study based on the MNI series of 48 cases. *Advances in neurology*, *57*, 419-433.
8. Frucht, S. (2002). Dystonia, athetosis, and epilepsia partialis continua in a patient with late-onset Rasmussen's encephalitis. *Movement disorders: official journal of the Movement Disorder Society*, *17*(3), 609-612.
9. Villani, F., Spreafico, R., Farina, L., Giovagnoli, A. R., Bernasconi, P., Granata, T., & Avanzini, G. (2001). Positive response to immunomodulatory therapy in an adult patient with Rasmussen's encephalitis. *Neurology*, *56*(2), 248-250.
10. McLachlan, R. S., Girvin, J. P., Blume, W. T., & Reichman, H. (1993). Rasmussen's chronic encephalitis in adults. *Archives of neurology*, *50*(3), 269-274.
11. Krauss, G. L., Campbell, M. L., Roche, K. W., Huganir, R. L., & Niedermeyer, E. (1996). Chronic steroid-responsive encephalitis without autoantibodies to glutamate receptor GluR3. *Neurology*, *46*(1), 247-249.
12. Bien, C. G., Granata, T., Antozzi, C., Cross, J. H., Dulac, O., Kurthen, M., ... & Elger, C. E. (2005). Pathogenesis, diagnosis and treatment of Rasmussen encephalitis: a European consensus statement. *Brain*, *128*(3), 454-471.
13. Graus, F., Titulaer, M. J., Balu, R., Benseler, S., Bien, C. G., Cellucci, T., ... & Glaser, C. A. (2016). A clinical approach to diagnosis of autoimmune encephalitis. *The Lancet Neurology*, *15*(4), 391-404.
14. Sanfilippo, C., Giuliano, L., Fatuzzo, D., Sortino, G., Le Pira, F., Sofia, V., & Zappia, M. (2016). Late onset Rasmussen encephalitis: complete remission after one session of plasmapheresis. *European Journal of Neurology*, *23*(3), e15-e16.
15. Sanfilippo, C., Giuliano, L., Fatuzzo, D., Sortino, G., Le Pira, F., Sofia, V., & Zappia, M. (2016). Late onset Rasmussen encephalitis: complete remission after one session of plasmapheresis. *European Journal of Neurology*, *23*(3), e15-e16.