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Cerebellar Syndrome Revealing Carcinoma of The Cavum

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Abstract **Case Report**

Subacute cerebellar degeneration is one of the classic paraneoplastic neurological syndromes. It may be isolated or is only part of a larger nervous system involvement. We report the observation of a 45-year-old patient with no pathological history who presents an isolated cerebellar syndrome that has progressed for 3 months and has gradually worsened. Cerebral MRI found, in addition to cerebellar atrophy, thickening of the lining of the cavum which led to an ENT examination with biopsy of the cavum allowing confirmation of the undifferentiated corcinoma of the UCNT type. The patient was referred to the oncology department for management. Depending on the clinical and immunological criteria, different sub types of paraneoplastic cerebellar ataxias can be defined. These syndromes correspond to an autoimmune process originally directed against the tumor. They often have a devastating and irreversible course because they are accompanied very quickly by neuronal degeneration. Two therapeutic axes have been proposed: immunosuppressive therapy and tumor treatment. The prognosis of these syndromes can only be improved with the fastest possible diagnosis and treatment of cancer.

Keywords: Cerebellar Revealing Carcinoma Cavum.

INTRODUCTION

Paraneoplastic neurological syndromes have always fascinated clinicians due to the lack of understanding of how a tumor, often very small or only discovered at autopsy, could cause such devastating effects on the nervous system from a distance without it invade [1]. What is more, these syndromes turned out to have such a predictive value for the existence of an occult cancer that the diagnosis of one of them, in a given patient, justified implementing all the means necessary to the discovery of the tumor.

CLINICAL OBSERVATION

A 54-year-old patient was hospitalized for a balancé and gait disorder that had progressed for three months in a context of apyrexia and general condition maintenance. His history of illness did not reveal any recent pathological history, with moreover an absence of medication or ingestion of toxicants. There is no dry syndrome. The patient was conscious and afebrile with a stable hemodynamic state.

Neurological examination showed an ataxic and unsteady gait, an increase in the polygon of support, normal deep sensitivity, hypotonia with pendular reflexes and coordination disturbances, producing an isolated statokinetic cerebellar syndrome. The remainder of the physical examination did not show any abnormality. Magnetic resonance imaging (MRI) of the brain showed in addition to cerebellar atrophy, thickening of the lining of the cavum.

The otolaryngology examination with nasofibroscopy found a suspicious aspect of the lining of the cavum. The biopsy and histological study of the mucosa confirmed the malignant nature of the lesion: undifferentiated corcinoma of the UCNT type.

The cerebrospinal fluid study did not show any abnormalities, including testing for anti Yo and anti Hu autoantibodies.

Testing for these autoantibodies in serum was negative. The patient was referred to an oncology department for management and was not seen again.

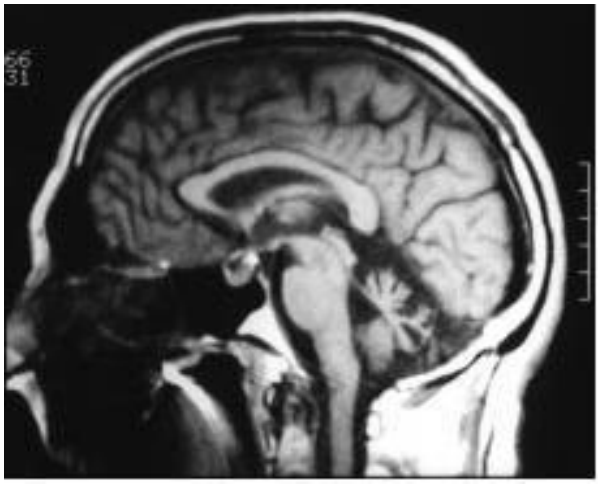

Fig1: Brain MRI in sagittal section showing cerebellar atrophy

DISCUSSION

Paraneoplastic cerebellar ataxias are one of the most common clinical paraneoplastic syndromes [5]. It was probably described by Brouwer in 1919 in a 60-year-old woman with pelvic sarcoma. This syndrome was not finally clearly established until 1982 [3]. Neuropathologic features of paraneoplastic cerebellar ataxia include elective disappearance of Purkinje cells from the cerebellum, and inflammatory perivascular infiltrates [8]. Sometimes we can also observe a decrease in the size of the molecular layer, proliferation of Bergmann's glia and loss of granular layer neurons.

The hypothesis that PCAs may be of autoimmune origin was first proposed in 1961 by Russel [6], who suggested that an antigen-antibody reaction could explain the presence of the inflammatory perivascular infiltrates observed in the brains of patients. . This hypothesis has led several groups to systematically search for the presence of anti-nervous system antibodies in the serum and CSF of patients with paraneoplastic neurological syndrome. In 1976, Trotter et al. [7] first identified an anti-Purkinje cell antibody in the serum of a patient with cerebellar ataxia and Hodgkin's disease. Following this first observation, several different autoantibodies have been reported in association with paraneoplastic neurological syndromes [2]. Depending on the clinical and immunological criteria, different subtypes of paraneoplastic cerebellar ataxias can be defined. In some types, cerebellar ataxia is isolated, while in others it is only part of a larger nervous system involvement.

Although there are many arguments in favor of autoimmune disease, only occasional observations have reported benefit from immunosuppressive therapy in paraneoplastic neurological syndromes [1].

Several observations of paraneoplastic cerebellar ataxias have been reported in association with other autoantibodies, the presence of which is of uncertain significance, but it indicates the existence of an unusual activation of the immune system which could play a role in the occurrence of the cerebellar ataxia. It is nevertheless interesting to note that in an extremely large percentage (probably more than 50%), of patients with confirmed paraneoplastic cerebellar ataxia no circulating anti-neuronal antibody is identified [4] and this is the case of our observation. The etiology of cerebellar ataxias in these patients without autoantibodies remains speculative. It is nevertheless very probably of autoimmune origin since most of these patients have inflammation of the cerebrospinal fluid but not in our patient and the pathological lesions are not different from those observed in patients who have antibodies. Surprisingly, the clinical course of patients without antibodies appears to be slightly different from patients with antibodies [4]. Indeed, patients with PCA with anti-Hu antibodies generally die due to the rapid spread of neurological disorders and not to tumor development, while the reverse is observed for patients without antibodies. These results suggest that the immune system may be involved in the occurrence of cerebellar ataxia in different ways in patients without and with antibodies.

CONCLUSION

Although the pathological role in cerebellar ataxia of anti-cerebellar antibodies has not been able to be formally demonstrated, apart from anti-GluR1 antibodies, it appears extremely important to seek them systematically because they allow an accurate etiological diagnosis. In addition, depending on the type of antibody, it is possible to predict the clinical course.

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