A CASE OF BICKERSTAFF BRAINSTEM ENCEPHALITIS WITH ISOLATED SERUM ANTI-GM2 ANTIBODIES

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INTRODUCTION

Bickerstaff brainstem encephalitis (BBE) is a rare, severe, and rapidly progressive inflammatory process, most commonly linked to anti-GQ1b antibodies. Clinical manifestations typically include ataxia, altered consciousness, and ophthalmoplegia that usually recover within a 12-week period[1]. Our aim is to report a case of BBE associated with anti-GM2 antibodies to analyze the clinical feature of this rarer form.

MATERIALS AND METHODS

A 58-year-old male presented with sudden-onset dysarthria. Multimodal CT imaging ruled out cerebrovascular etiology. During hospitalization, the patient developed progressive dysphagia, right-sided limb dysmetria, gait ataxia, diplopia, divergent strabismus, bilateral ptosis, and abolition of pupillary light reflex with fixed mydriasis. Progressive drowsiness necessitated transfer to the intensive care unit. Thereafter he developed tetraparesis and then required tracheostomy and gastrostomy to support living functions.

RESULTS

Serial CSF analyses showed increased proteins and leucocytes while excluded any infectious etiology. Serological testing showed positivity for anti-GM2 ganglioside antibodies. Brain MRI revealed a symmetrical mesencephalic hyperintensity on T2-weighted images with vasogenic edema extended into the pontine tegmentum and both superior cerebellar peduncles. It displayed intense, homogeneous and bilateral contrast enhancement. Intravenous methylprednisolone was administered which led to regained consciousness and loss of contrast enhancement on the MRI. Differential diagnosis with lymphoma was considered but since the location of the lesion excluded a biopsy, we decided to insist on the immunotherapy and so administer intravenous immunoglobulins and then Rituximab; ketogenic diet was also associated. The patient showed a progressive but very slow improvement: after 6 months from onset, he is still in rehabilitation, needing bilateral support for ambulation.

DISCUSSION

This case highlights the peculiar course of BBE associated with anti-GM2 antibodies. They have been previously associated with dysimmune peripheral neuropathies where they correlate with a more severe course[2] and our case suggests that this may be valid for BBE as well.

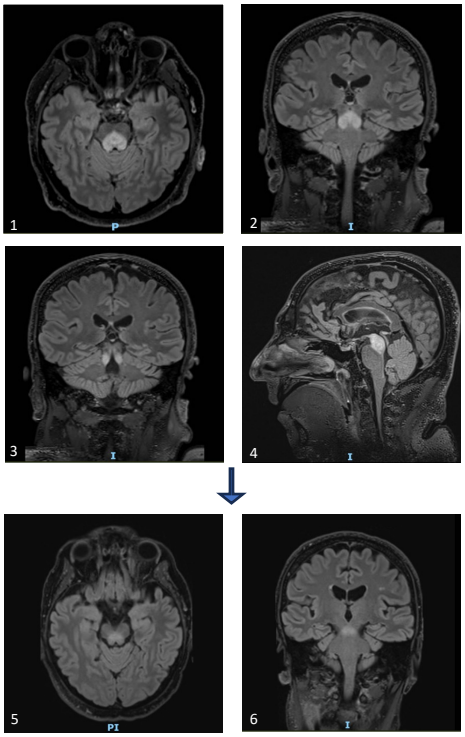

Fig.1,2,3,4: MRI symmetrical mesencephalic T2 hyperintensity
Fig.5,6: MRI improvement after 3 months

CONCLUSION

Diagnosing BBE is often challenging and serological and CSF analyses and neuroimaging are essential to establish the underlying etiology, which is fundamental for initiating targeted treatment. Nonetheless, scarce response to treatment like our case could be deceiving and wrongfully lead clinicians to abandon the autoimmune hypothesis for a neoplastic one. This case however shows that refractory course is not exclusive for tumors but could be referred to more aggressive antibodies such as anti-GM2.

Bibliography

1)Graus F, Titulaer MJ, Balu R, Benseler S, Bien CG, Cellucci T, Cortese I, Dale RC, Gelfand JM, Geschwind M, Glaser CA, Honnorat J, Höftberger R, Iizuka T, Irani SR, Lancaster E, Leypoldt F, Prüss H, Rae-Grant A, Reindl M, Rosenfeld MR, Rostásy K, Saiz A, Venkatesan A, Vincent A, Wandinger KP, Waters P, Dalmau J. A clinical approach to diagnosis of autoimmune encephalitis. Lancet Neurol. 2016 Apr;15(4):391-404. doi: 10.1016/S1474-4422(15)00401-9. Epub 2016 Feb 20. PMID: 26906964; PMCID: PMC5066574.

2) Kim JK, Kim YH, Yoon BA, Cho JY, Oh SY, Shin HY, Kim JS, Park KH, Kim SY, Suh BC, Seok HY, Yoo JH, Bae JS. Clinical Heterogeneity of Anti-GM2-Ganglioside-Antibody Syndrome. J Clin Neurol. 2018 Jul;14(3):401-406. doi: 10.3988/jcn.2018.14.3.401. PMID: 29971981; PMCID: PMC6031993.