案例報告: 神經貝賽特氏病症—影像及病理發現

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Diffuse MRI: Apparent Diffusion Coefficient and Pathologic Findings in a Case of Neuro-Behçet's Disease

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Background :

Behçet's disease (BD) is known with inflammatory perivasculitis which may involve multiple organs, and rarely invade central nervous system. Here, we reported a case with neuro-Beçhet's disease with acute onset of right cerebellar symptoms.

Case Report:

A 45-year-old man with a history of Behçet's disease (BD) came to our emergency department because of deteriorated ataxia for one week. BD was diagnosed one year ago with the initial presentation of repeated oral, genital ulcers and skin rashes. Uvietis with left eye total blindness were noted half a year ago. He regularly took immune modulation medication (cyclosporine 200mg, cyclophosphomide 50mg, levamisole 50mg, prednisolone 7.5mg, colchicine 1mg per day) for disease control.

One week prior to admission, he suffered from ataxia with right side deviation. Initially there was no focal weakness, but progressive right side clumsiness was noted within 2-3 days. On neurologic examination, direct fundoscopy showed only mild pale on optic disc of left eye, and dysmetria on right extremities. On the magnetic resonance imaging (MRI) study, T2-weighted images and fluid attenuated inversion recovery (FLAIR) images showed multiple hyperintensitivity ovoid spots in bilateral periventriuclar white matter of cerebrum and cerebellum without obvious enhancement, and apparent diffusion coefficient (ADC) showed round lesions with increased intensity on right cerebellum.

Brain biopsy from the lesion on left frontal lobe was performed and the histologic examination found mild gliosis with abundant foamy histiocytes and no accumulation of inflammatory cells within vessels on H-E stain, favored demyelinating diseases. Pulse therapy with methylprednisolone 1000mg QD for 5 days was given and immune modulation therapy with Cyclosporin 100mg 1# BID; Levamisole 50mg 1#QD; Prednisolone 5mg 8# QD were maintained. The neurological deficits got gradually improved. He followed up at our OPD for two months, without further deterioration of neurologic deficits. Then, he got lost of follow up.

Conclusion:

We reported a case of neuro-BD with initial manifestation as acute onset of right cerebellar symptoms. The diagnosis of BD is based on clinical manifestation. In our case, brain MRI detected multiple hyperintensitivity ovoid lesions on bilateral cerebrum and cerebellum on FLAIR and T2-weighted image, whereas, only one lesion on right cerebellum showed increased intensity on ADC. This lesion detected on ADC was compatible with clinical manifestations. Increased intensity in ADC suggested vasogenic edema in this brain lesion. Vasogenic edema in this brain lesion implied acute change of permeability of involved vessels which may be secondary to perivasculitis, an important pathologic marker of BD. In summary, our case showed clinical manifestation of BD with brain involvement confirmed by clinical symptoms and brain image findings, so neuro-BD was impressed.

According to the McDonald Criteria 2010, at least one episode of attack and multiple cerebral and cerebellar ovoid lesions which disseminated in at least two different times in this case suggested probable MS. However, the location of biopsy was not active lesion in this case, and the differential diagnosis of neuro-BD from MS in this case still needed more clinical data including longer follow-up period.

We suggested ADC sequence may contribute to detection of acute brain lesion with vasogenic edema in neuro-BD. And there still have diagnostic dilemma in differential diagnosis of neuro-BD from MS by McDonald Criteria.

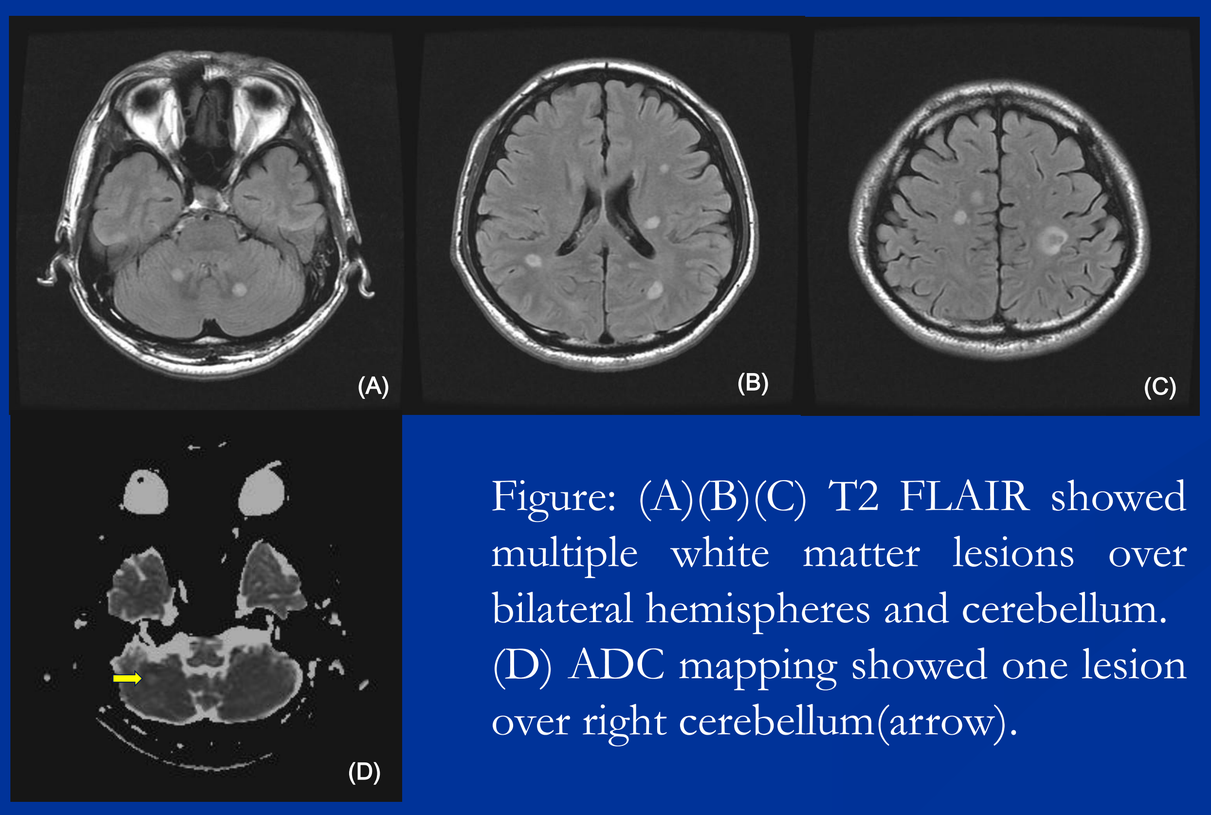

Figure: (A)(B)(C) T2 FLAIR showed multiple white matter lesions over bilateral hemispheres and cerebellum. (D) ADC mapping showed one lesion over right cerebellum(arrow).

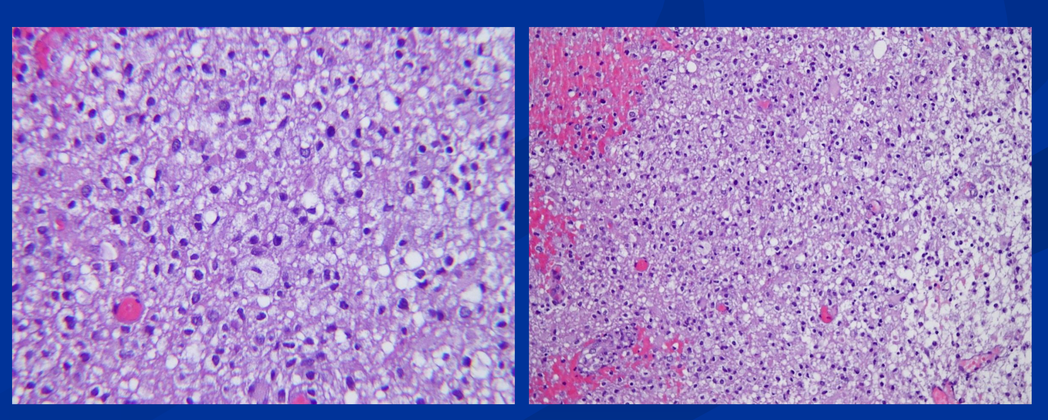

Figure: Brain biopsy, left frontal area. Mild gliosis with abundant foamy histiocytes, in favor of demyelination.