Autoimmune GFAP astrocytopathy diagnosed after long term diagnosis of CLIPPERS

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BACKGROUND

Cases of autoimmune glial fibrillary acidic protein (GFAP) astrocytopathy who were initially diagnosed with chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids(CLIPPERS) were rarely reported.

METHODS

Herein, we reported a case of 31-year-old woman. Her clinical history, symptoms, brain MRI enhancement features and response to treatment during each attack were reviewed.

RESULTS

A 31-year-old woman presented with 7 years of recurrent headache. Her initial brain MRI (figure 1a) demonstrated characteristic pepper-like enhancement of pontine and cerebellum and her symptoms resolved completely after taking high-dose of steroid. She was suspected with the diagnosis of CLIPPERS and experienced 5 relapses once the oral steroid was tapered below 20 mg per day(figure 2). During her last relapse, she experienced fever and psychosis, and GFAPα-antibodies were detected in her serum and cerebrospinal fluid by antigen-transfected HEK293 cell-based assay (figure 3). She obtained relief again after steroid therapy and her diagnosis converted to autoimmune GFAP astrocytopathy.

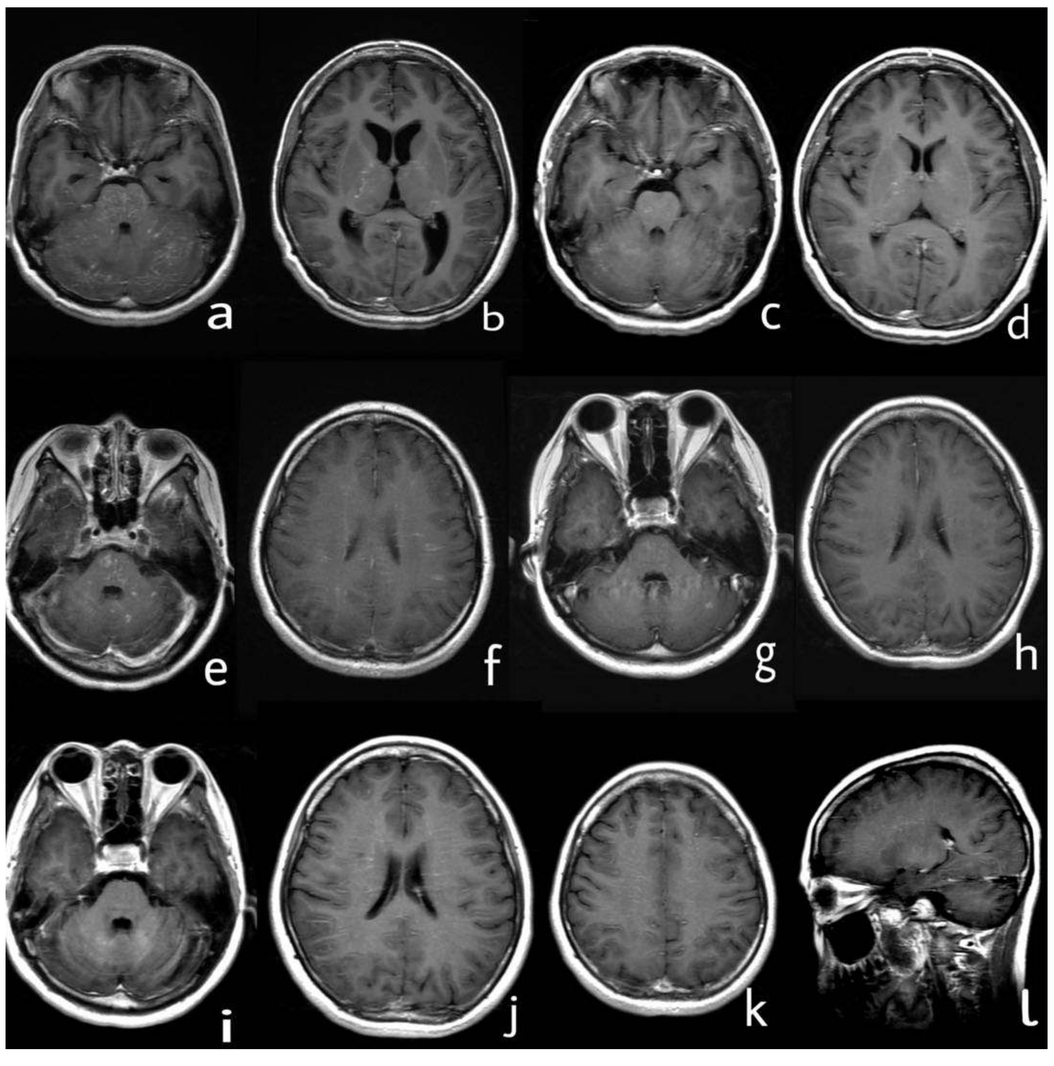

Figure 1. Series of brain MRI with contrast. (a-b) March 2012, curvilinear and punctate enhancement of pontine, cerebellum and basal ganglion was demonstrated. (c-d) June 2012, enhancement of pontine, cerebellum and basal ganglion improved after treatment with steroids. (e-f) June 2015, novel dot-like and liner enhancement in pontine, semiovale center and juxtacortical area was showed. (g-h) September 2015, enhancement in pontine, semiovale center and juxtacortical area improved again after treatment with steroids. (i-l) September 2019, novel dot-like and liner enhancement in both hemisphere of the cerebellum, semiovale center, basal ganglion, thalamus was demonstrated.

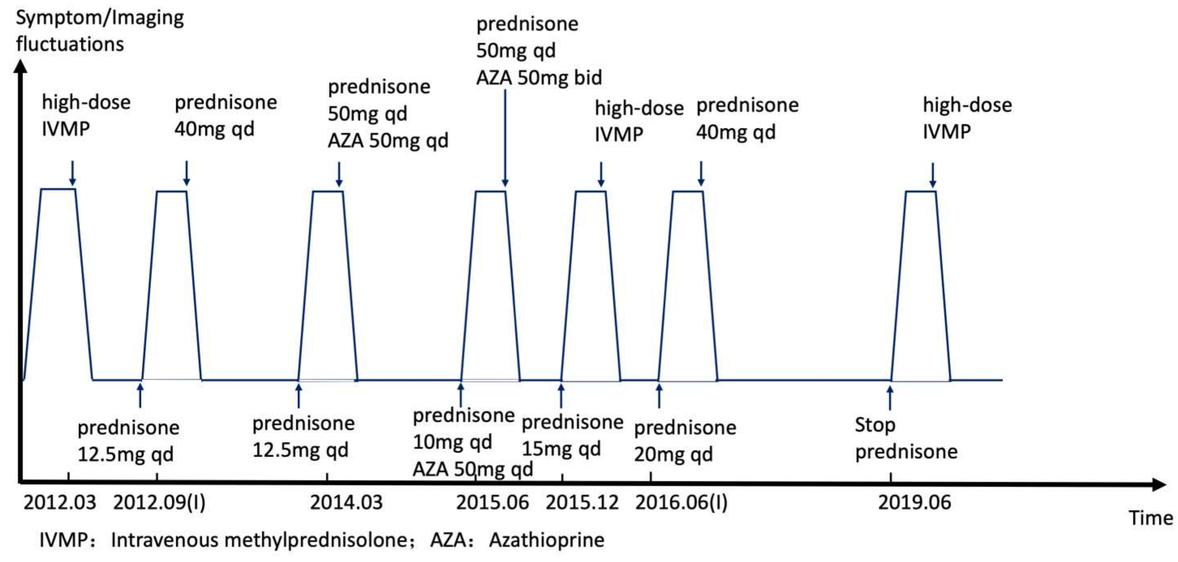

Figure 2. Clinical course of the case.

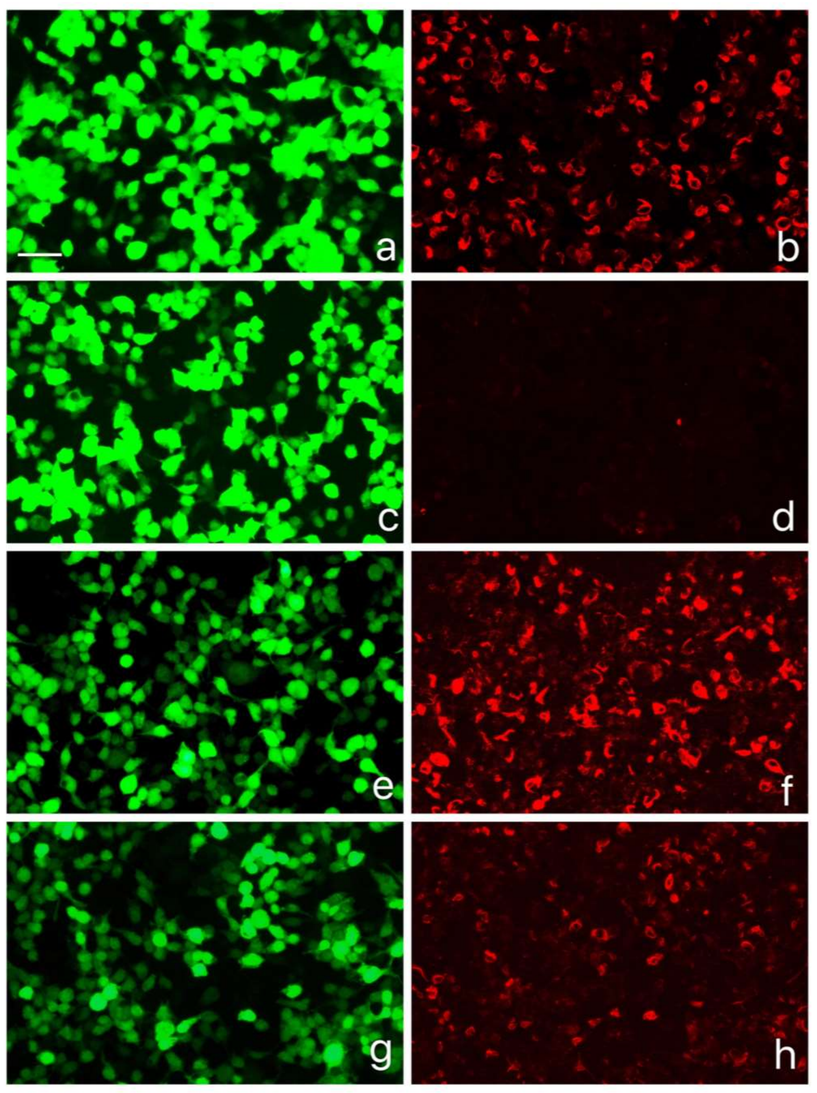

Figure 3. GFAPα-IgG test results by GFAP-transfected HEK293 cell-based immunofluorescence assay. (a, c, e, g) HEK293 cells expressing green fluorescent protein (GFP)-tagged GFAP (green) and (b, d, f, h) HEK293 cells immunostained with human IgG (red if positive). (b) positive control with human anti-GFAPα IgG, (d) negative with healthy control, (f) positive result of serum (titer at 1:320), (h) positive result of cerebrospinal fluid(CSF) (titer at 1:32). Scale bar=50um.

CONCLUSIONS

Autoimmune GFAP astrocytopathy may mimic CLIPPERS both clinically and radiologically. Long term follow-up is essential for necessary diagnosis revision at each new attack in patients with diagnosis of CLIPPERS.