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Clinical Image

Chronic Lymphocytic Inflammation with Pontine Perivascular Enhancement Responsive to Steroids (CLIPPERS) Mimicking Posterior Circulation Infarction

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Abstract

CLIPPERS is a rare inflammatory condition of the central nervous system. It is pathologically characterised by white matter perivascular lymphohistiocytic infiltrate with or without parenchymal extension. A male patient in mid sixties presented with dysarthria and facial numbness underwent an MRI brain which supported a diagnosis of a subacute posterior circulation infarct. The patient re-presented with worsening of symptoms and a repeat MRI revealed changes atypical for ischaemia and consistent with CLIPPERS. Our case represents a common presentation of a very rare condition.

***Keywords:** CLIPPERS, Chronic Lymphocytic Inflammation; Pontine Perivascular Enhancement.*

Case Description

A male patient in his mid 60s presented with a two-week history of mild swallowing discomfort, a metallic taste in his mouth, right sided numbness of the lips and tongue and left face. He was dysarthric. At this stage there was no evidence of limb incoordination. The initial MRI (Figure 1) revealed signal abnormalities in the pons, left inferior cerebellar peduncle, and right anterior thalamus, suggesting subacute infarcts, particularly due to the timescale. An MR angiogram (Figure 2) showed an abnormal appearance of the left vertebral artery, supporting the diagnosis. The patient had a history of hypertension and was on amlodipine and lisinopril. Clopidogrel, simvastatin, and paracetamol were also added. Three months later there was left-sided weakness with numbness, worsening dysarthria and reduced sensation on the face. Examination revealed ataxia with bilateral gaze-evoked nystagmus, dysdiadochokinesis and intention tremor. Repeat MRI (Figure 3) showed worsening signal abnormalities in the cerebellum, midbrain, pons, and medulla. Curvilinear enhancement on post-contrast T1 suggested perivascular spaces, not typical for ischemic changes. Laboratory tests and lumbar puncture were normal. A diagnosis of Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS) was made based on the radiological findings. Steroids and immunosuppression in the form of cyclophosphamide for twelve months were initiated. At initial follow-up at two months, there was clinical and imaging improvement (Figure 4). After twelve months low dose prednisolone was continued and azathioprine was used for continued immunosuppression. The subsequent course was one of recurrent relapses followed by partial remission but with sustained disability. He continues to be severely ataxic and has residual dysarthria with nystagmus and impaired limb coordination.

The differential diagnosis for the radiological appearances includes neurosarcoidosis and lymphoma. The unique imaging features of this case were the diffusion abnormalities and the abnormal left vertebral artery which necessitated the exclusion of a vascular aetiology such as vasculitis.

CLIPPERS, an inflammatory CNS condition, is characterized pathologically by perivascular lymphohistiocytic infiltrate in white matter. The diagnosis involves clinical, radiological, and histopathological criteria and, responsiveness to glucocorticoid treatment. Due to limited evidence, there are no unified treatment strategies for CLIPPERS. Immunosuppressive therapies such as azathioprine, cyclophosphamide, mycophenolate mofetil, and rituximab can be considered based on available literature.[1, 2]

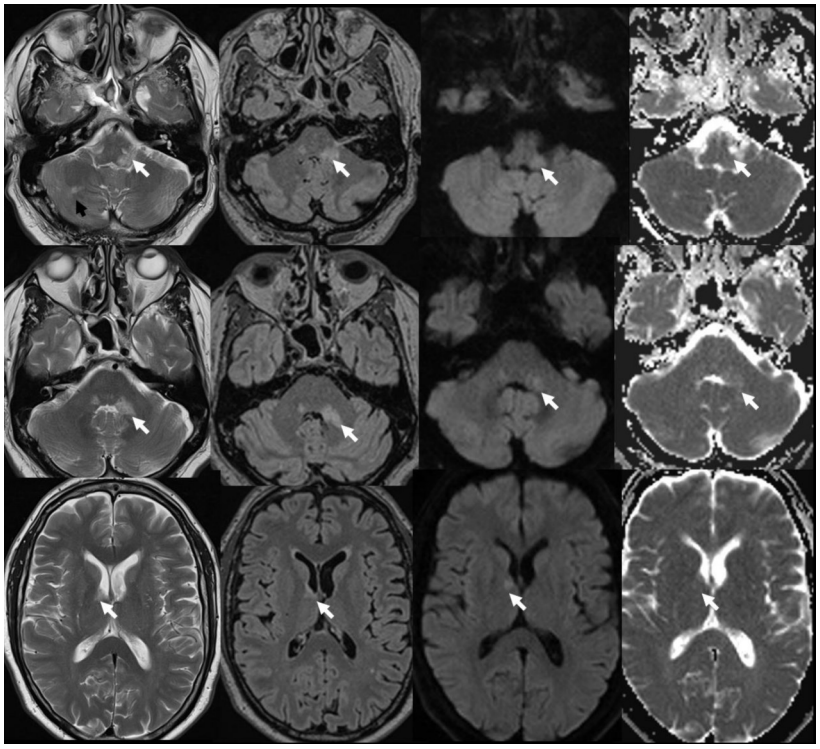

Figure 1: *Initial MRI. Signal abnormalities on T2 and FLAIR are noted in pons, inferior cerebellar peduncle on the left side and right anterior thalamus (white arrows) with associated subtle diffusion abnormalities.*

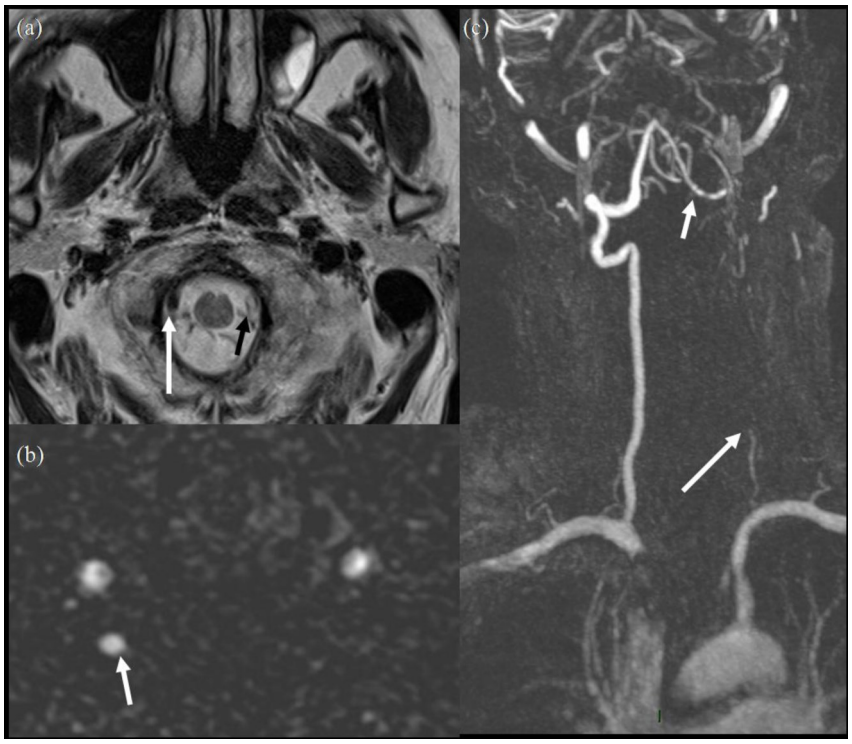

Figure 2: *Initial MRI. T2 axials (a) showing normal right vertebral artery (white arrow) and abnormal signal in the left vertebral artery (black arrow). Axial MRA reconstructions (b) show normal right vertebral artery (white arrow) while the left vertebral artery is missing. Maximum Intensity Projection (MIP) images of MRA (c) show a barely identified V1 segment of left vertebral artery (long white arrow) and an abnormal and irregular V4 segment of left vertebral artery (short white arrow).*

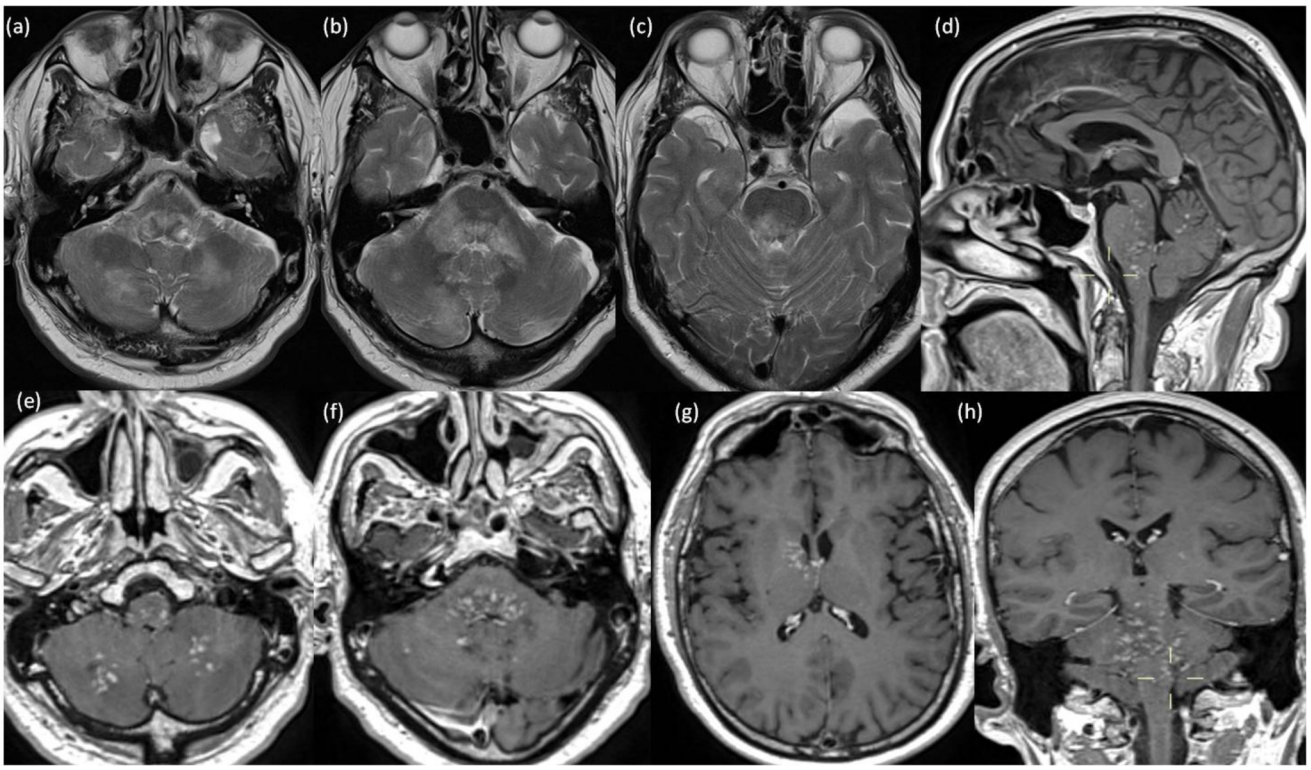

Figure 3: *Subsequent MRI at 3 months. T2 axial images through posterior fossa (a-c) and post contrast T1 images (d-h) showing linear and punctate areas of enhancement throughout brainstem, cerebellum and right basal ganglia with associated high T2 signal.*

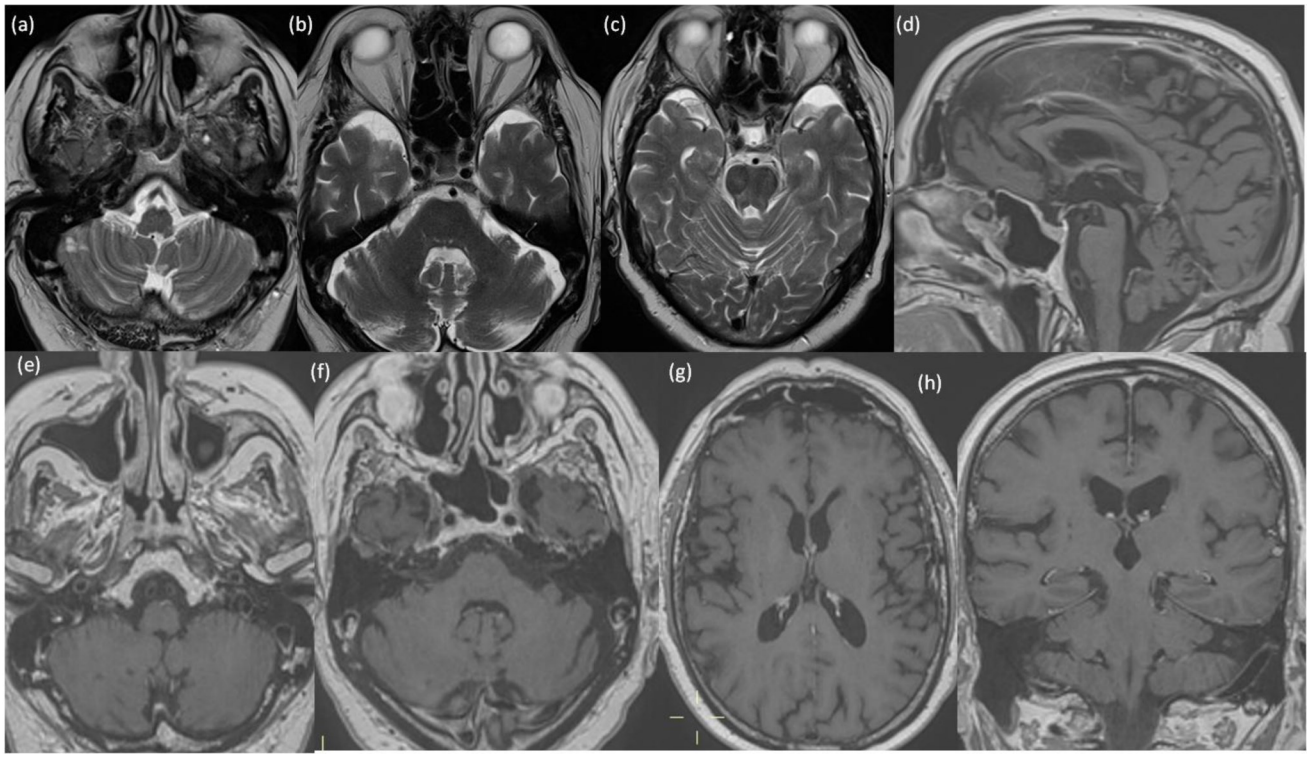

Figure 4: *Post treatment MRI after 2 months. T2 axial images through posterior fossa (a-c) and post contrast T1 images (d-h) showing resolution of the changes seen in Figure 3.*

Conclusion

CLIPPERS is a rare syndrome with striking radiological findings. While the initial presentation may mimic a cerebrovascular event magnetic resonance imaging shows characteristic findings. A differential diagnosis of lymphoma or other neuro inflammatory disorders such as neuro sarcoidosis or vasculitis needs to be excluded. It is an immune mediated disorder which is steroid responsive but almost invariably will require additional immunosuppression.

Conflict of Interest

None.

References

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