

A rare case of CLIPPERS presenting with mass effect and diffusion restriction on magnetic resonance imaging

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Abstract

Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS) is an inflammatory disorder of the CNS, most commonly involving brainstem and is characterized by punctate enhancing lesions on post contrast MRI. Alternative diagnosis such as Neurosarcoidosis, Sjogren's syndrome, neuro-Bechet's disease, vasculitis and lymphoma need to be excluded.

Keywords

CLIPPERS; pontine; inflammatory; contrast; punctate

Abbreviations

CNS: Central nervous system; MRI: Magnetic resonance imaging

Introduction

Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS) is a chronic inflammatory neurological syndrome which has gained increased recognition in recent times. It has a wide range of differentials, thus detailed investigation and workup is required to rule them out. The importance of this case is the atypical clinical & radiological presentation which made the diagnosis challenging. Atypical features for CLIPPERS in this case include mass effect, unilateral involvement, absence of spinal cord lesions, diffusion restriction, and larger size of T2 signal abnormalities compared to areas of contrast enhancement.

Case Report

A thirty-two- year old female presented with headache, vomiting and vertigo for five days. There was no history of loss of weight and appetite. On examination she was afebrile, and investigations like blood picture, platelet count and fundoscopy were normal. Neurological examination showed the patient to be

well oriented with normal pupillary reflexes, however there was gaze-evoked horizontal-torsional nystagmus. There was no neck rigidity or signs of peripheral nerve irritation. Cerebrospinal fluid analysis revealed normal cytology, protein and glucose with lymphocytic pleocytosis (lymphocyte 98 %). No malignant cells were reported. CSF culture was negative for tuberculosis and fungal infection.

On MRI, ill -defined T2 weighted and FLAIR hyperintensity with T1 hypo intensity was seen involving left side of dorsal pons, left middle cerebellar peduncle and the adjacent left superior cerebellar hemisphere. Surrounding edema & mass effect was seen causing partial effacement of fourth ventricle. There were no areas of hemorrhage. Mild patchy diffusion restriction was seen. Post contrast T1 weighted images show punctate, homogeneous foci of enhancement 'peppering' the brainstem, middle cerebellar peduncle and cerebellum. There was no abnormal leptomeningeal enhancement. Time of flight (TOF) angiography showed no abnormality. Screening of cervical cord was normal. There was no brain or spinal cord atrophy. Follow up MR imaging showed complete resolution of above findings and associated mass effect with no enhancing lesions on T1 weighted post contrast imaging performed after four months.

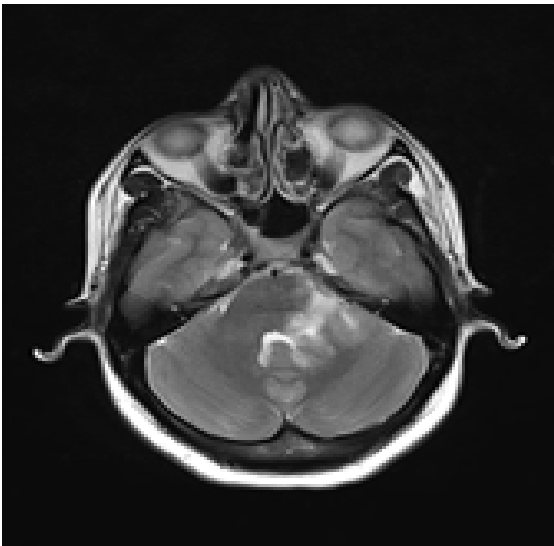


Figure 1: Axial T2 weighted MR image shows ill -defined hyperintensity involving left side of dorsal pons, left middle cerebellar peduncle and adjacent left superior cerebellar hemisphere. Surrounding mild mass effect is also seen.

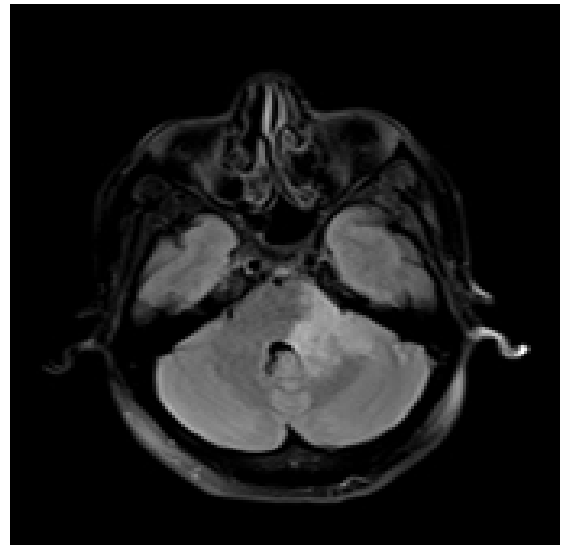


Figure 2: Axial FLAIR MR image shows ill -defined hyperintensity involving same areas of brainstem, middle cerebellar peduncle and adjacent cerebellum.

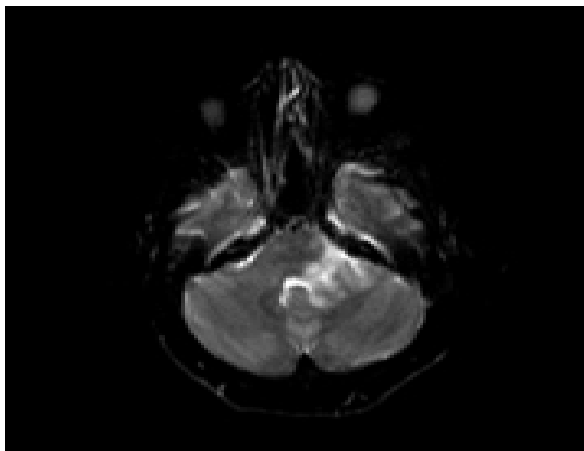


Figure 3: There is hyperintensity involving above areas on axial diffusion weighted MR image.

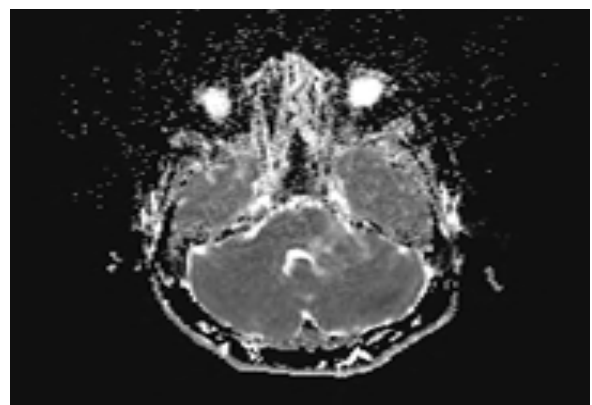


Figure 4: Hypo intensity in affected areas on corresponding Apparent diffusion coefficient (ADC) image, suggestive of mild patchy diffusion restriction was seen.

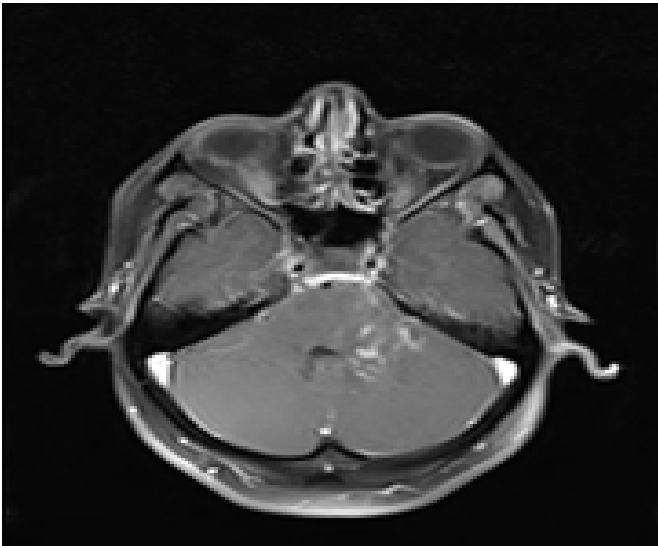


Figure 5: Post contrast T1 weighted image shows punctate, homogeneous foci of enhancement 'peppering' the brainstem, middle cerebellar peduncle and cerebellum. There was no pachymeningeal or leptomeningeal enhancement.



Figure 6: Follow up MR imaging showed complete resolution of enhancing lesions and mass effect.

Discussion

Diagnostic features of CLIPPERS are- (a) subacute progressive pontocerebellar dysfunction, (ii) bilateral punctate enhancing lesions on MRI brain, predominantly in the pons or cerebellum, (iii) absence of mass effect, (iv) reduction in number and size of enhancing lesions following corticosteroid treatment on follow up imaging and (v) exclusion of other differentials [1].

Clinical features include headache, vomiting, vertigo, dysarthria, diplopia and ataxia. It is mainly a diagnosis of exclusion. Differential diagnosis can include vasculitis, angiocentric lymphoma, neurosarcoidosis, neuro-Behcet's disease, neuromyelitis optica (NMO IgG was negative in this case), infective process (like tuberculosis), acute disseminated encephalomyelitis (ADEM), Posterior reversible encephalopathy syndrome (PRES), paraneoplastic syndromes and pontine glioma. It may be associated with other autoimmune diseases. [2] Lesions generally involve pons and middle cerebellar pedicles but can also involve cerebellar hemispheres, brain and spinal cord. Diffusion restriction is generally absent. Post contrast the lesions show homogeneous enhancement and measure less than 3mm in size. [3] IgM MOG antibodies have been linked with CLPPERS and lymphoma in literature. Cerebrospinal fluid (CSF) analysis typically shows lymphocytic pleocytosis with normal protein and glucose. Biopsy is not needed in typical cases and was not performed in this case because the patient improved clinically and radiologically after steroid treatment. [4] The initial treatment of choice is a relatively short course of high-dose intravenous methylprednisolone followed by oral glucocorticoids [5].

Final diagnosis: Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS)

Differential diagnosis: Angiocentric lymphoma, Pontine glioma

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