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Central and Peripheral Nervous System Vasculitis as Devastating Manifestation of Cryoglobulinemia

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Introduction

Several neurological disorders have been described in chronic HCV infection, as cerebrovascular acute events and mono- or polyneuropathies, consequence of small and/or medium-sized vessels vasculitis. It remains unclear whether deposition of cryoglobulins plays a direct pathogenic role or whether it simply represents an epiphenomenon of the immune response.

Case Report

A previously healthy 59-year-old Caucasian woman presented to the emergency department in March 2015, with a 4-mo history of malaise, arthralgia migrants, and last week fatigue, muscle weakness, low-grade fever, finger cyanosis (**fig. 6**). Flaccid quadriparesis and symmetrical distal sensory loss were noted.

Results

An electroneuromyographic study revealed sensorimotor multineuropathy **(Fig. 1).** She performed routine blood tests, tumor markers, immunological screening, lumbar puncture, brain MRI sequences with Angio, echocardiography, whole-body CT and specialist consult. The rheumatologic panel was negative. Further testing showed negative serology for hepatitis B virus, HIV, syphilis, cytomegalovirus, and human T-lymphotropic virus 1/2. HCV antibodies were positive, as well as serum HCVRNA. Serum cryoglobulins were positive with a criocrito of 8%. We then considered peripheral multineuropathy related to HCV infection and the patient was treated with intravenous methyl-prednisolone. After two weeks, she became increasingly disoriented. Brain MRI showed high-signal lesion in the left thalamus and small foci of increased signal intensity at the semioval center and subcortical white matter on T2 and FLAIR sequences **(Fig. 2 e 3)**. After 15 days she showed drowsiness, mental confusion and motor focal seizures. Thus it was initiated therapy with cyclophosphamide at dosage of 150 mg and apheretic sessions. The control brain MRI highlights the appareance of areas of alterated signal of the white matter subcortical, predominantely in the occipital and parietal lobes and less marked in the right frontal lobe, with signal confluent patches referred to interstizial edema **(Fig. 4)**. The patient get worse towards coma with absence of response to painful stimuli and worsening of peripheral involvement. Nevertheless she continued apheretic sessions, but a new brain MRI showed a severe deterioration with alterated signal areas affecting both cerebral hemisferes and vertebrobasilar district, leveled furrows in the occipitoparietal regions bilaterally to be referred to ischemic lesion in the acute phase. Angio MRI showed diffuse luminal alteration of cerebral vessel of Willis circle in the carotid and vertebrobasilar district with marked reduction of caliber of the anterior and middle cerebral arteries more evident on the right posterior cerebral artery (**Fig 5**). After some days she dead.

Conclusions

The description of this case was suggested by

a) **severity and rapidity of evolution** besides the singular contemporary association of the damage both peripheral and central
b) an **unusual severe progressive multineuropathy**
c) concerning the **central injury**, the **large caliber vessels involvement**

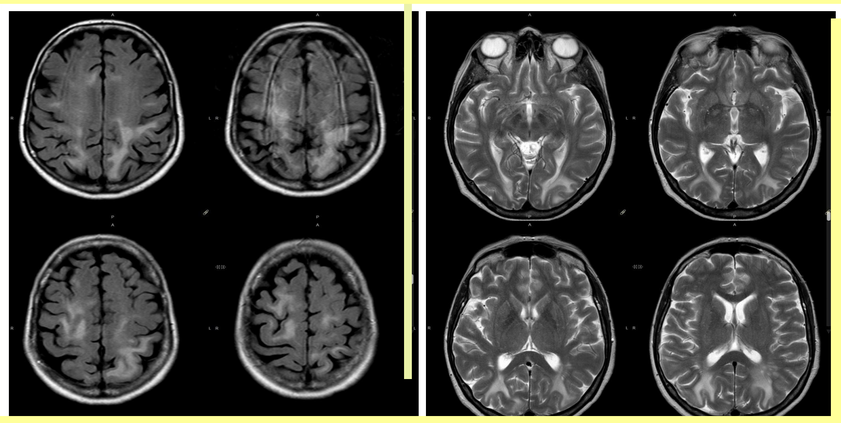

Fig. 2

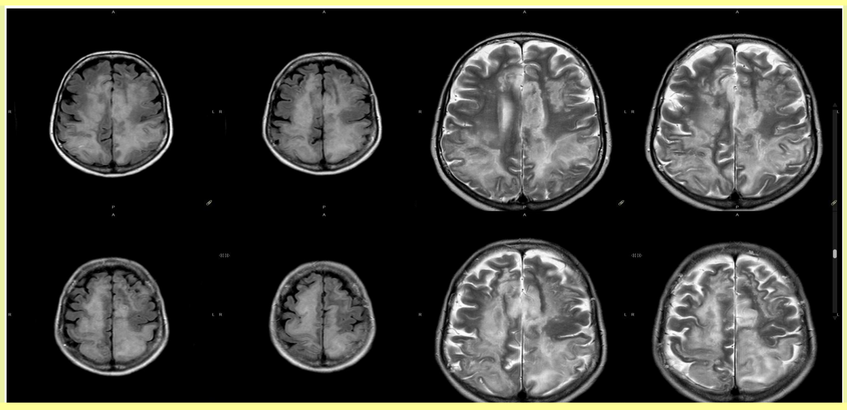

Fig. 3

Fig 2 e 3. High-signal lesions in the left thalamus and small foci of increased signal ntensity at the semioval centers and subcortical white matter on FLAIR and T2 sequences

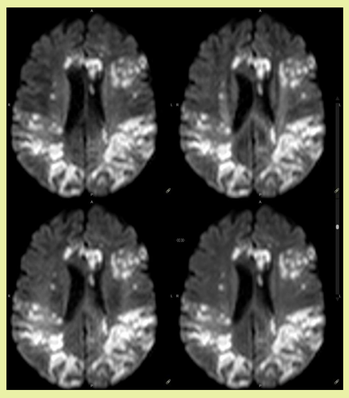

Fig. 4

Fig. 4. Areas of alterated signal as diffusion restriction in DWI sequences of the white matter subcortical, predominantely in the occipital and parietal lobes and less marked in the right frontal lobe, with signal confluent patches

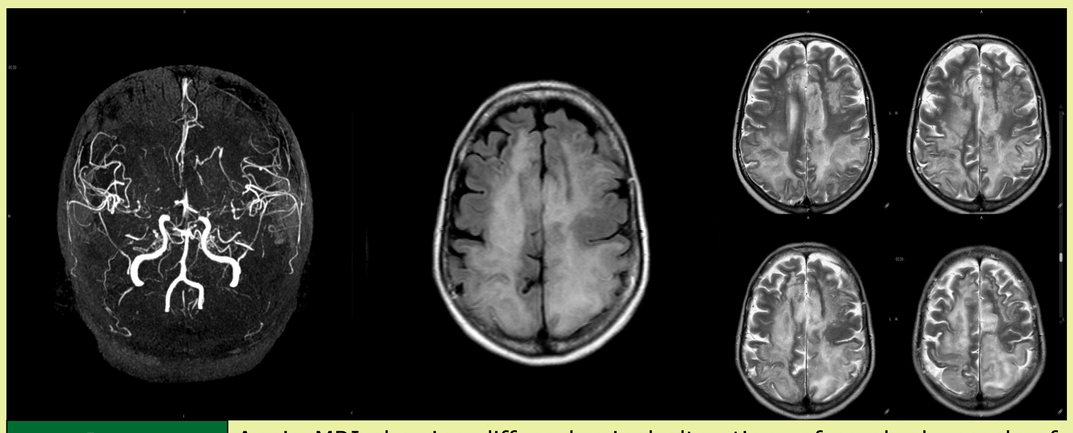

Fig. 5

Angio MRI showing diffuse luminal alteration of cerebral vessels of Willis circle in the carotid and vertebrobasilar district with marked reduction of caliber of the anterior and middle cerebral arteries. Alterated signal areas affecting cerebral emisferes and vertebrobasilar district in FLAIR and T2-sequences

Fig. 6

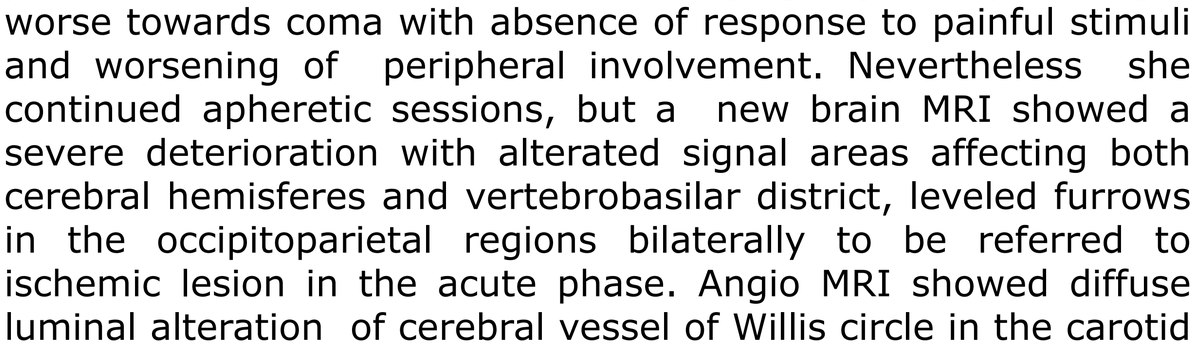

Fig. 1

References

1) Origgi L et al. Central nervous system involvement in patients with HCV-related cryoglobulinemia. Am J Med Sci. 1998.
2) Cacoub P et al. [Central nervous system involvement in hepatitis C virus infection] Gastroenterol Clin Biol. 1998.
3) Monaco S et al. HCV-related nervous system disorders. Clin Dev Immunol. 2012.
4) Serena M et al. Peripheral and central nervous system involvement in essential mixed cryoglobulinemia: a case report. Clin Neuropathol. 1991
5) Bonetti B et al. Hepatitis C virus infection of peripheral nerves in type II cryoglobulinaemia. Virchows Arch. 1999.
6) Vital C et al. Combined nerve and muscle biopsy in the diagnosis of vasculitic neuropathy. A 16-year retrospective study of 202 cases. J Peripher Nerv Syst. 2006.
7) Gemignani F et al. Cryoglobulinemia is a frequent cause of peripheral neuropathy in undiagnosed referral patients. J Peripher Nerv Syst. 2002.
8) Gemignani F et al. Clinical spectrum of cryoglobulinaemic neuropathy. J Neurol Neurosurg Psychiatry. 2005.
9) Bonetti B et al. T-cell-mediated epineurial vasculitis and humoral-mediated microangiopathy in cryoglobulinemic neuropathy. J Neuroimmunol. 1997.

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