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A CASE OF PRIMARY CENTRAL NERVOUS SYSTEM ANGIOITIS

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INTRODUCTION

Primary angioitis of the central nervous system is a rare disease. It is a diagnostic and therapeutic challenge for clinicians because there are no specific findings. Its progression is usually acute or subacute. An aggressive workup is necessary in order to avoid poor prognosis.

CASE REPORT

A 62 years old man developed left facial and neck numbness, dysphonia, diplopia and ataxia with gait imbalance. He underwent a brain MRI which showed a T2/Flair hyperintense and T1 hypointense lesion in the left side of bulbar, pontine an upper cervical region (C1-C2), with enhancement, bulbar haemorrhagic hypointense spot in T2*, without restriction in DWI (Fig. 1). The other region of the spinal cord were normal. He was submitted to extensive radiological and laboratory investigations which excluded haematological or solid neoplasm and infectious diseases. Isoelectrofocusing demonstrated oligoclonal bands, cytology and flow cytometry were normal.

A diagnosis of CLIPPERS (chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids) was supposed and the patient was treated with high doses of steroids followed by oral prednisone 1 mg/kg with improvement of symptoms and neuroradiological findings.

About three weeks after discharge the patient acutely developed aphasia and right hemiparesis. A Ct scan showed a left fronto-parietal acute vascular lesion (fig 2). Brain MRI showed in the frontal left region a large area of hypointensity in T1 and hyperintensity in T2, with enhancement and slight restriction in DWI, oedema, mass effect and haemorrhagic spots inside. Another similar lesion was detected in the right cerebellar hemisphere (fig. 3). A diagnosis of cerebral vasculitis was supposed and the patient was treated with metilprednisolone 1 gr iv, then with ciclophosphamyde 1500 mg iv.

The development was complicated by a Sepsis by Lysteria monocitogenes, the neurological picture worsened rapidly and the patient died after two months from the onset of symptoms. Autopsy confirmed the diagnosis of PNCSA (lymphocytic variant).

Fig. 1

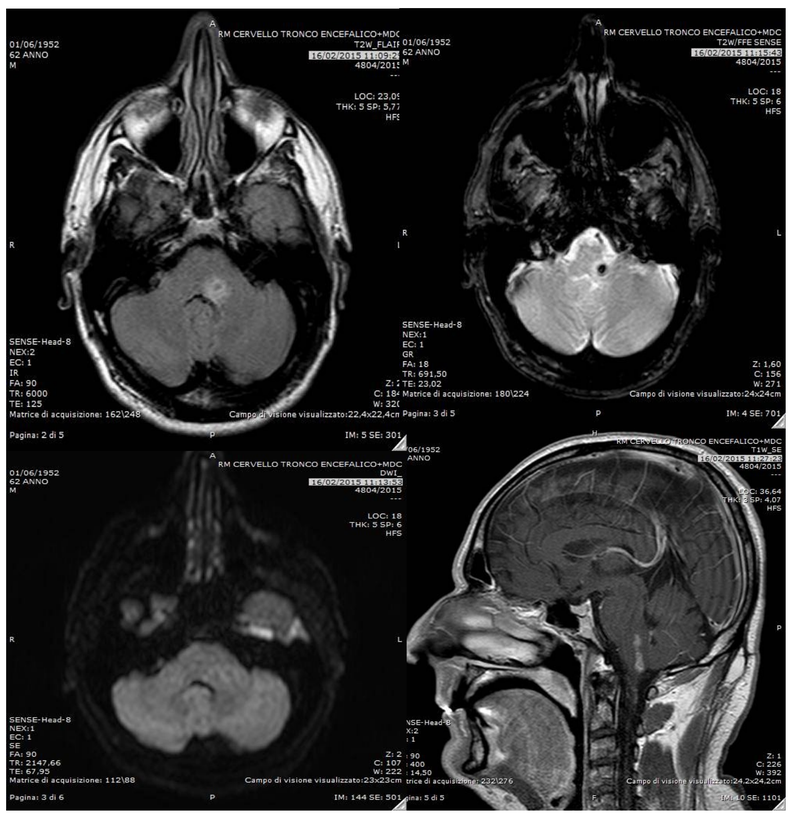


Fig.2:

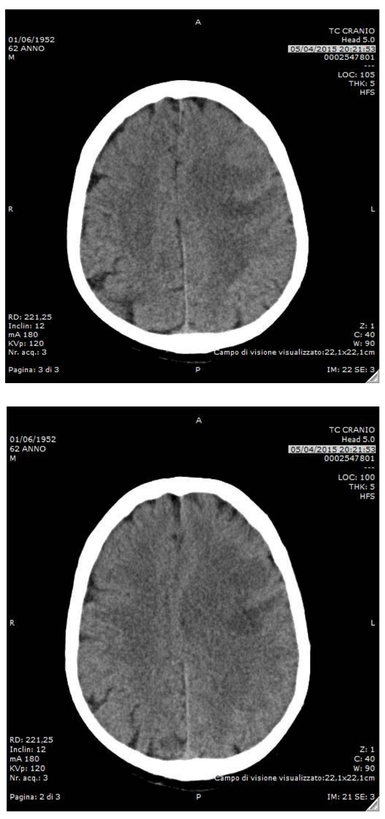
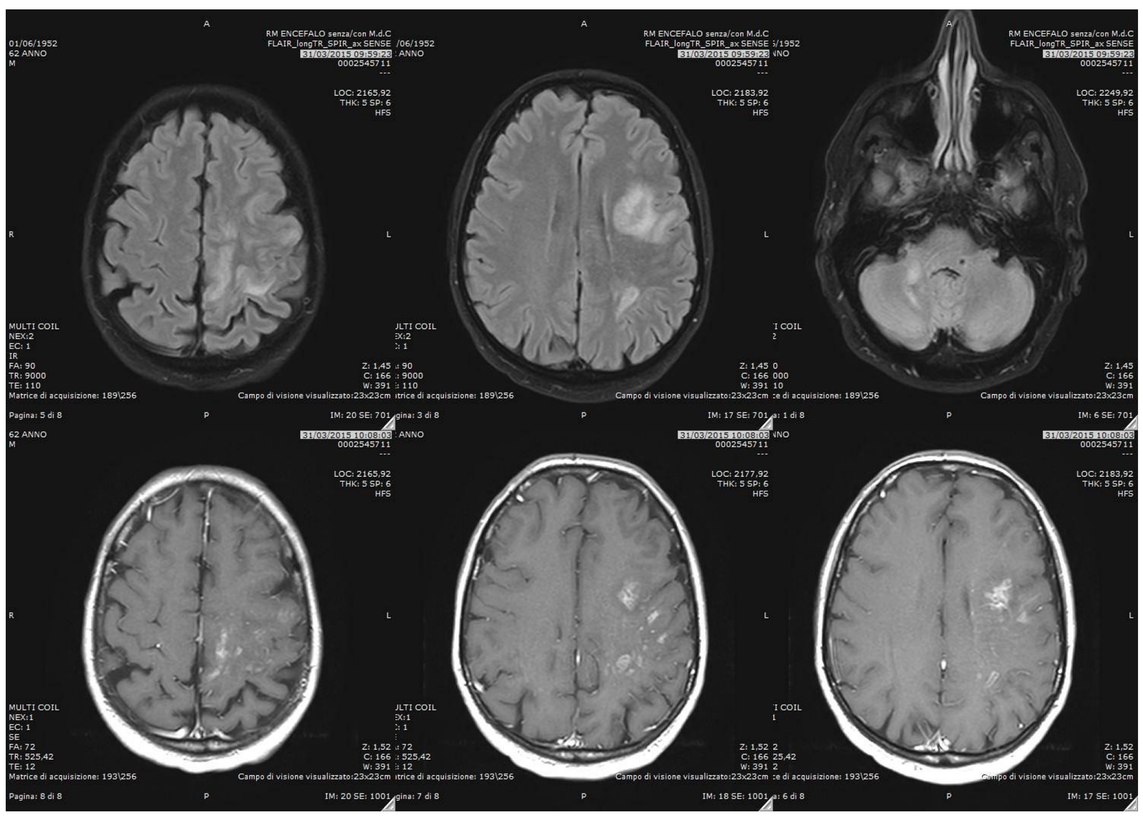


Fig.3:



DISCUSSION AND CONCLUSION

Clinical manifestations of PACNSA include non specific symptoms of variable severity and progression. Also diagnostic tests are not specific. Additionally PACNSA may mimic different disorders such as infections, malignancies, inflammatory diseases, non-inflammatory vasculopathies, demyelinating diseases. In the case reported, considering clinical presentation and diagnostic tests we supposed a diagnosis of inflammatory disease of CNS and the improvement with steroid treatment supported this hypothesis. Only the following clinical evolution oriented to a more aggressive disease and despite the immunosoppressive therapy the course was rapidly fatal. We think that in similar cases, when laboratory and neuroradiological findings are not diagnostic, a cerebral biopsy should be taken in account very quickly, in order to start a specific therapy as soon as possible and in order to prevent poor prognosis.

BIBLIOGRAPHY

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