ISOLATED NEUROLOGICAL WHIPPLE'S DISEASE PRESENTING WITH RHOMBENCEPHALITS AND CERVICAL MYELITIS.

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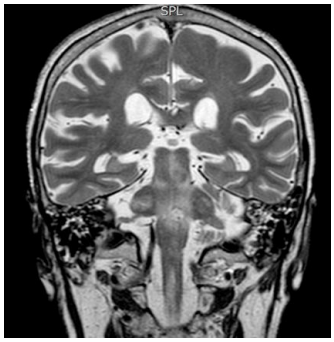
INTRODUCTION

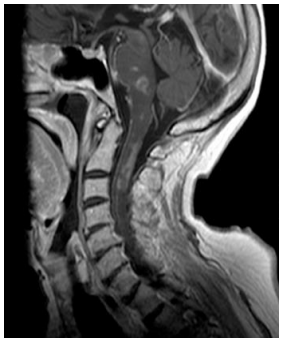
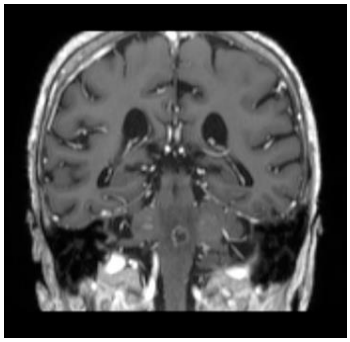
Whipple's disease is a very rare infection and chronic immunosuppressive treatment influences the course of disease. We report a case of a immunosuppressed patient with isolated neurological involvement.

CASE REPORT

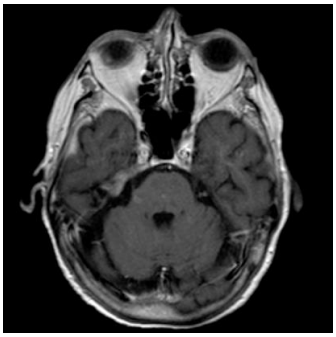
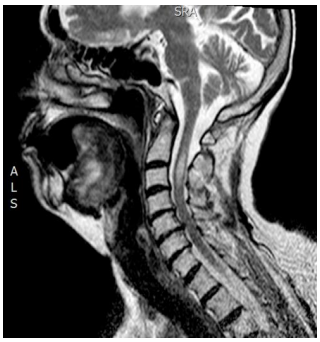
A 76-years-old woman was admitted to our Neurology Department for subacute onset of diplopia, vertigo, postural instability and headache associated to a febrile episode. She had 9-years history of rheumatoid arthritis in therapy with Methotrexate and oral steroids. She reported to be allergic to penicillin.
On admission neurological examination revealed ptosis, diplopia, right dysmetria without cognitive or strength or sensitive impairments. Patient's clinical conditions rapidly worsened in three days with mild consciousness deterioration, multiple cranial nerve deficits and right ataxic paresis.
Brain CT scan was negative. Lumbar puncture showed clear fluid with moderate pleiocytosis, elevated proteins and normal glucose levels. In suspect of viral encephalitis acyclovir was introduced and immunosuppressive was ceased. Three days later results of viral PCR performed on CSF were negative so acyclovir was stopped. Culture and microscopic examinations on CSF excluded common infectious.

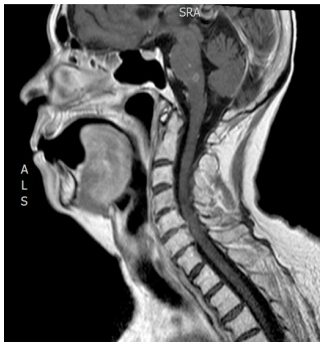
Although negative exams, blood tests showed increasing infectious signs and her neurological conditions progressively worsened with severe consciousness deterioration and signs of brainstem involvement: considering patient's allergy antibiotic treatment with Meropenem was started.



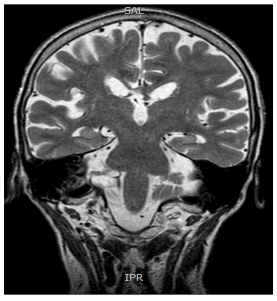
On 7th day after admission
MRI showed multiple T2 and FLAIR iperintense contrast-enhancing lesions involving pons, medulla, cerebellum and proximal tract of the cervical spinal cord. Antibiotic spectrum was widened introducing Linezolid and Cotrimoxazole. Further examinations excluded the most common infectious (listeria, toxoplasmosis, cryptococcus, brucella, borrelia, TBC), autoimmune and neoplastic disorders.

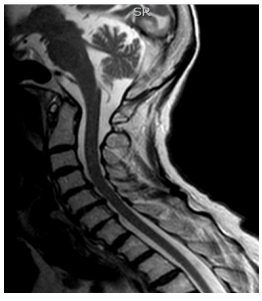





Two weeks later patient experienced gradual and progressive clinical and radiological improvement. CSF analysis confirmed general improvement showing lower cellularity and proteins concentration.

CSF was sent for further molecular analysis and PCR for Tropheryma Whipplei was found to be positive. Antibiotic treatment was de-escalated continuing cotrimoxazole only.





Follow up 6 months later showed gradually clinical and instrumental improvement. In particular she is able to walk with single support and brain MRI showed complete resolution. She complains arthalgia as a resumption of rheumatic disease for which she has resumed low dose of oral steroids.

Although she had not gastrointestinal signs and symptoms. She performed multiple duodenal biopsy, that are now ongoing.

A folllow up at 1 years will be included MRI and CSF controls in order to make a treatment decision.

DISCUSSION

Rhomboencephalitis have several causes, that included infections, autoimmune and paraneoplastic disorders. Tropheryma whipple is a rare cause and isolated CNS involvement is very difficult to diagnose due to its different presentations and the lack of sensitive diagnostic tests. Early diagnosis is very important as the disease is rapidly progressive and fatal if left untreated.

References
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