ANAPLASTIC ASTROCYTOMA MIMICKING NEUROMYELITIS OPTICA: A CASE REPORT

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BACKGROUND

The intramedullary anaplastic astrocytoma is a rare tumour representing only 0.5-1% of the spinal cord tumours, which affects almost exclusively children and adolescents and it is characterized by a rapid progression. Only few case-reports are described in literature with an intracranial dissemination. We report a case with evidence of spinal cord and brain lesions suspected at first as an inflammatory origin and later diagnosed as anaplastic astrocytoma.

CASE REPORT

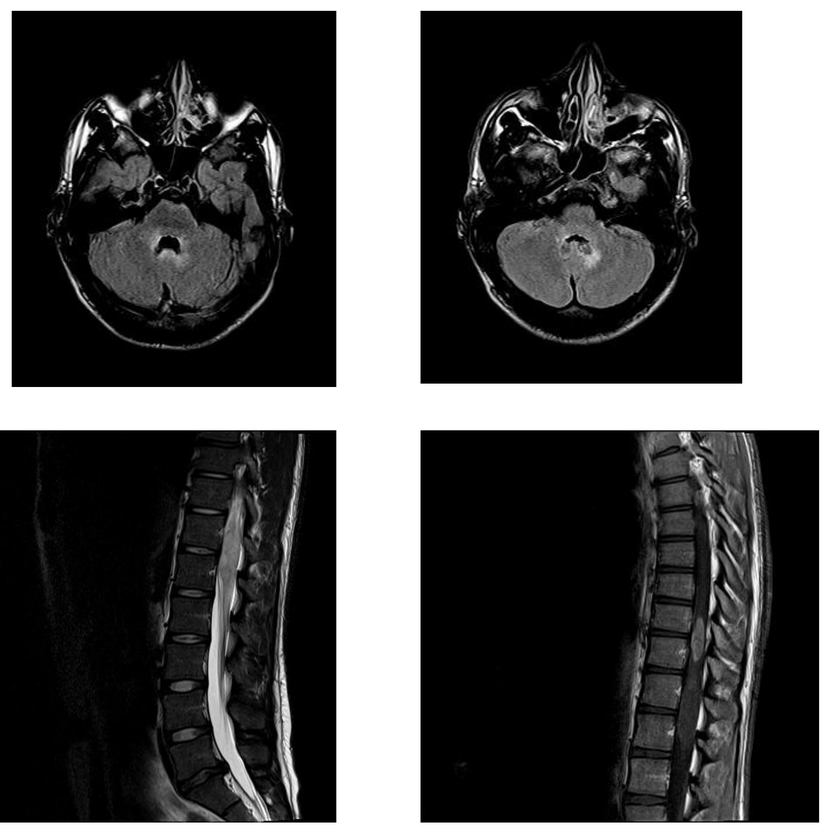
ANAMNESIS

A 18 years old man, without previous remarkable medical history, developed an acute weakness in the distal part of the right leg. At the neurological examination, an impairment in the dorsiflexion of the right foot was noted. The neurophysiological study showed findings consistent with the damage root.

During the following months he suffered from a progressive worsening of the weakness extending to the knee and accompanied by an extension of the tingling and an altered feeling of the floor.

After two months the patient developed acute urinary retention and he was admitted to our neurological department, where he presented with a sensory level from the 11th thoracic dermatome and a complete foot drop to the right limb.

MRI FINDINGS



SEROLOGIC AND CSF FINDINGS

CSF analysis: cytochemical (proteins 1.24 g/l, G.B. 1/mm3), immunochemical (no intrathecal Ig synthesis), immunoblot (NO oligoclonal bands on CSF and serum).

Flow cytometry immunophenotyping of peripheral blood: normal

Considering the characteristics of the spinal cord lesion (centromedullary, swollen and involving more than 3 metamers) and the localization of the brain abnormalities, a diagnosis of Neuromyelitis optica was supposed.

THERAPIES

- intravenous steroid therapy (1g/day for 5 days)
- intravenous steroid therapy tapering (0,5 g/day for 3 days + 0,25 g/day for 3 days, +0,125 g/day for 3 days)

! Sudden worsening characterized by the involvement of the left limb and a new episode of urinary retention !

- Rituximab 1g/m^2
- IvIg 0,4 g/kg/day for 5 days

! Patient's clinical conditions worsened with paraplegia and total anaesthesia and hypotrophy in both lower limbs. !

Anti-MOG and anti-Aqp4 Ab: negative

Considering the clinical and radiological lack of response to the therapies, the progression of symptoms and the anti-Aqp4 and anti-MOG antibodies negative values, we decided to reconsider the diagnosis.

CSF analysis: cytochemical (proteins 2.75 g/l, G.B. 2/mm3), immunochemical (no intrathecal Ig synthesis), immunoblot (one oligoclonal band on CSF and serum).

CSF cytomorphological analysis: presence of isolated atypical cellular elements characterized by enlarged nuclei, sometimes with irregular profile.

BIOPSY

Microscopic Description: Biopsy fragments of neoplasia characterized by high cellularity and cellular atypia. Absence of mitosis and necrosis in the material under examination.

Immunohistochemistry: GFAP: 5; SYNAPTOPHYSIN: 1; CK AE1-AE3 +: 0; EMA : 0

Diagnosys: IDH1 wild type anaplastic astrocytoma (WHO grade III)

CONCLUSIONS

In this case the diagnosis of neuromyelitis optica has been formulated mainly on the characteristics of the MRI images.

Two issues led us to reformulate the diagnosis: the progressive chronic course of the disease and the fact that, as reported in literature, intracranial dissemination of anaplastic astrocytoma is rare but possible through the spinal subarachnoid space and ventricles.

In conclusion, as reported here, the differential diagnosis of a longitudinally extensive medullary lesion is a challenge; the correlation of clinical, bioumoral and radiological data and the evaluation of the response to the therapy approached are necessary to make the right diagnosis.

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