eISSN 2255-0569

CASE REPORT

Not everything is what it seems

No todo es lo que parece

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Received: 15 - VI - 2022
Accepted: 26 - VI - 2022

doi: 10.3306/AJHS.2022.37.04.174

Abstract

We report a case of a 36-year-old man, with previous medical history of marihuana consumption, high blood pressure and asthma. He came to the emergency room with a 4-month history of altered behaviour and a fall from his own height. After conducting complementary tests, diagnoses were acute renal failure secondary to rhabdomyolysis and brain space occupying lesions (SOLs) under study. Initial suspicion of brain metastasis radically changed with the results of brain magnetic resonance imaging and thoraco-abdomino-pelvic computed tomography. This lead us to focusing on inflammatory-granulomatous aetiology, with our first option being neurosarcoidosis, later confirmed by the histological results of an adenopathic biopsy performed by fiberoptic bronchoscopy.

Resumen

Se presenta el caso de un hombre de 36 años, con antecedentes de consumo de marihuana, hipertensión arterial y asma. Acudió a urgencias con una historia de 4 meses de alteración del comportamiento y una caída desde su propia altura. Tras la realización de pruebas complementarias, los diagnósticos fueron de insuficiencia renal aguda secundaria a rabdomiólisis y de lesiones cerebrales ocupantes de espacio (SOL) en estudio. La sospecha inicial de metástasis cerebral cambió radicalmente con los resultados de la resonancia magnética cerebral y la tomografía computarizada toraco-abdomino-pélvica. Esto nos llevó a centrarnos en la etiología inflamatoria-granulomatosa, siendo nuestra primera opción la neurosarcoidosis, posteriormente confirmada por los resultados histológicos de una biopsia adenopática realizada por broncoscopia de fibra óptica.

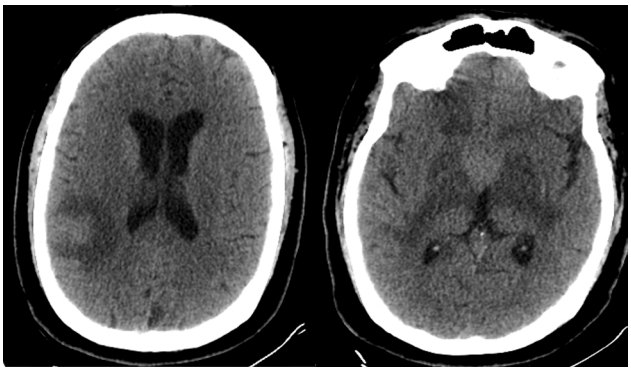
Case description

This is a 36-year-old man, a marijuana smoker (between 5-6 cigarettes a day), with a medical history of arterial hypertension treated with enalapril (unknown blood pressure controls) and bronchial asthma treated with salbutamol on demand.

The patient is brought to the emergency room by the emergency services after being unable to get up after a fall from his own height while at his home. During anamnesis, the patient and his relatives explained a 4 month history of blurry vision, behavioral alteration, difficulty walking and a 30 kg weight gain. He had been evaluated 72 hours previously by a psychiatrist in the emergency department, ruling out an eating disorder and recommending follow-up by Primary Care.

Upon initial assessment at the emergency department, the patient presented with a fever of 37,8ºC, with no clear signs of infection in the physical examination, displaying only spatial-temporal disorientation, verbhorrea, a cushingoid phenotype (full moon face, abdominal striate and buffalo hump), increased base of support and functional impotence in both shoulders due to pain. Blood tests showed acute renal failure with creatinine of 3.54 mg / dl in the context of rhabdomyolysis (creatine kinase of 3879 U / l). A brain computed tomography (CT) without contrast was requested, highlighting two intracranial lesions on the hypothalamic and right parietal level (**Figure 1**).

Figure 1: Simple head CT without contrast. In the image on the left, we can see a slightly hyperdense image of approximately 20x17mm at the right parietal level, isodendal with the brain parenchyma, with perilesional hypodensity suggestive of edema. On the right, a slightly hyperdense image of approximately 23x20x22mm is observed located at the hypothalamic level and extending towards the third ventricle, with perilesional hypodensity that extends to both internal capsules, suggestive of vasogenic edema.



With these results, the decision was made to admit the patient in the Internal Medicine department with initial diagnoses of acute renal failure secondary to rhabdomyolysis and brain lesions to be studied in a patient with a cushingoid phenotype.

Our main suspicion when faced with two brain lesions was a metastatic neoplastic aetiology, for which tumor markers, urinary cortisol, a full body CT with contrast and brain magnetic resonance imaging (MRI) were requested to better characterize the lesions. The first results showed negative tumor markers, normal urine cortisol and a CT scan that reported 5 pulmonary micro-nodules, inframillimetric mediastinal and retroperitoneal lymphadenopathies, without other notable alterations. Before obtaining the brain MRI results and due to the now low suspicion of a tumor, it was decided to expand the study at hormonal, infectious and autoimmune levels. It was decided to request quantiferon, serologies for HIV, toxoplasma, Treponema pallidum, HCV and HBV, which were negative. Likewise, an autoimmunity screen was requested (ANA, rheumatoid factor, C3, C4, CH 50, ECA and ANCA) which were also negative. Regarding the hormonal study, it showed hypogonadotropic hypogonadism and a prolactin of 92 ng / ml without macroprolactinemia. It was decided to extend the study of haematological neoplasia with a serum proteinogram, LDH and beta2-microglobulin, which were normal.

Finally, the brain MRI reported a main lesion in the hypothalamic region that cranially compressed the 3rd ventricle and fornices (**Figure 2** and **3**), infiltrated the bilateral retrochiasmatic optic pathway, sparing the hypothalamus. The imaging of the second lesion visualized on brain CT, was discussed with radiology, informing us that it was a small lesion with significant linear leptomeningeal hyper-enhancement at the parietal and temporal level.

Figure 2: T1 axial brain MRI. The left image shows a lesion in the bottom of the sulcus with vasogenic edema. In the image on the right, a lesion occupying the hypothalamic region can be seen which cranially compresses the 3rd ventricle and fornices.

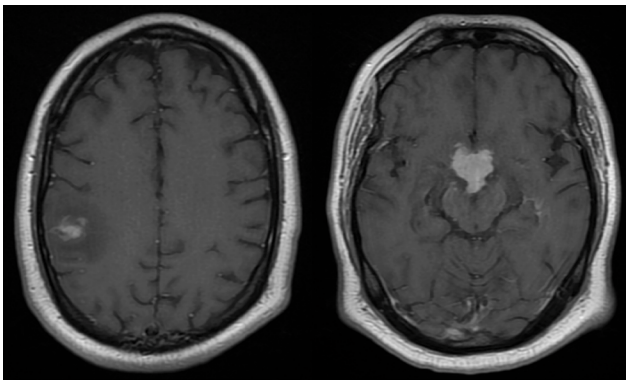
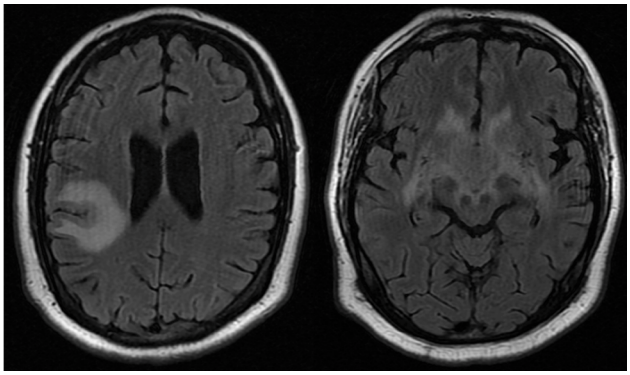


Figure 3: FLAIR T2 brain MRI. The image on the left shows thick linear leptomeningeal enhancement (we did not see a lesion at the bottom of the sulcus in this image). As for the image on the right, we appreciate a lesion in the hypothalamic area that compresses the 3rd ventricle and fornices cranially with significant edema.



Before requesting further diagnostic tests, the patient presented with an acute hypernatremia of 161 mEq/l, compatible with diabetes insipidus of central origin, showing good response to treatment with desmopressin.

Discussion

The importance of this clinical case lays not only in the correct differential diagnosis of brain SOLs, but also on the correct interpretation of the imaging tests requested. Initially, due to the performance of a brain CT without contrast in the context of acute renal failure, the diagnostic orientation was a possible neoplasm with paraneoplastic Cushing's syndrome and brain metastases. The results of the thoraco-abdomino-pelvic CT and brain MRI changed our view of the case completely.

A new differential diagnosis began, focusing on an inflammatory-granulomatous aetiology, with the most likely being neurosarcoidosis, Langerhans cell histiocytosis, and less likely granulomatosis with polyangiitis. Due to location, we also took into account primary neoplasia such as pituicytoma and suprasellar germinoma, with primary brain lymphoma being less likely.

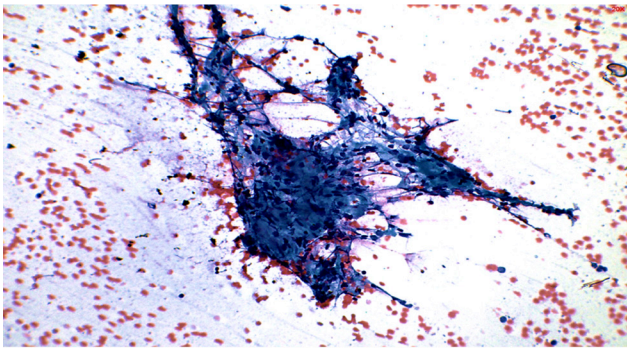
Our main suspicion focused on sarcoidosis with extra-pulmonary involvement, with between 5-10% presenting brain involvements. On the one hand, cerebral location and infiltration of the optic pathway is typical in neurosarcoidosis. This became our main diagnostic option due to its cerebral location and infiltration of the optic pathway. Likewise, due to hypothalamic inflammation, neuroendocrine alterations such as diabetes insipidus, appetite alteration and hypogonadotropic hypogonadism are also typical, as in the case of our patient. It is important to note that meningeal involvement can lead to confusion regarding infectious meningitis. Functional impotence in both shoulders also stood out upon physical examination, and is consistent with proximal myopathy that usually appears in cases of sarcoidosis.

Regarding the second option, Langerhans cell histiocytosis presents brain involvement in 6% of patients. In our clinical case, brain involvement aside, the typical picture of cystic pulmonary lesions did not match. In addition, the patient did not present osteolytic lesions (cranial being the most typical) or skin lesions such as eczematous rash or oral involvement.

Final clinical judgment

Pulmonary involvement and mediastinal lymphadenopathies visualized in the computed tomography lead us to request a fiberoptic bronchoscopy to perform a bronchoalveolar lavage and biopsy of the mediastinal inframillimetric adenopathy. While the CD4 / CD8 ratio of 1.60 with 10% of the lymphoid population obtained did not meet the criteria for sarcoidosis, pathological analysis non-caseating granulomas (**Figure 4**). Taking into account these histopathological results, the imaging tests performed and the patient's clinical history, the diagnosis of sarcoidosis with brain involvement was confirmed, beginning treatment with bolus of methylprednisolone 125mg every 24h for 3 days and subsequently rituximab 1g and mycophenolate mofetil 500mg every 12h. When the patient attended a check-up one month later, he presented a decrease in the size of the brain lesion and resolution of the myopathy.

Figure 4: Non-necrotizing granulomatous lymphadenitis. Aggregates of epithelioid-like histiocytes accompanied by lymphocytes and isolated plasma cells are observed in a hematic background. (20x Papanicolaou stain).



Conflict of interest

Authors do not have any conflict of interest to declare.

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