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Progressive meningoencephalitis due to neurosarcoidosis

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Study concept and design: Adamec and Habek. Acquisition of data: Adamec, Ozretić, Zadro and Habek. Analysis and interpretation of data: Adamec, Ozretić, Zadro and Habek. Drafting of the manuscript: Adamec. Critical revision of the manuscript for important intellectual content: Adamec, Ozretić, Zadro and Habek. Administrative, technical, and material support: Adamec, Ozretić, Zadro and Habek.

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Introduction

Neurological manifestations of sarcoidosis can be the presenting feature of the disease and are reported to be present in up to 15% of patients with systemic sarcoidosis (1). The most common neurological manifestation is cranial nerve palsy with a special propensity for the facial nerve (1). Almost half of the patients have symptoms of parenchymal affection which is demonstrated as encephalopathy, mass effect or hypothalamic disorder (1).

We present a patient with progressive meningoencephalitis and diffuse brain MRI abnormalities due to neurosarcoidosis.

Case report

A 38-year-old woman was referred to our hospital because of a progressive neurological decline. Her first symptom was an unexplained fever that started four months prior. Pneumonia was suspected but the fever persisted despite introduction of two different antibiotics. There were no other general signs. A month later she was first seen by a neurologist because of a left peripheral facial palsy. During the following months she developed bilateral facial palsy, her hearing became impaired and walking was becoming increasingly strenuous. On admission, examination revealed spastic tetraparesis with more severe affection of the legs. Her speech was dysarthric. She wasn't able to protrude her tongue. Bilateral palatal muscle paresis was present and she had difficulty swallowing. Brain MRI showed diffuse T2 and FLAIR hyperintensity of the periventricular and deep white matter. Symmetrical hyperintensity in the deincephalic-mesencephalic boundary, tegmentum of the pons, middle cerebellar peduncle and hylus

of the nucleus dentatus was visible (Figure 1). The MRI of the cervical spinal cord was normal. MR spectroscopy showed increased Choline in the frontal regions and reduced NAA in the parietal region (Figure 2). MR angiography was normal. Sedimentation rate was elevated (26 mm/h), leukocytes, C reactive protein, liver enzymes and B12 level were within normal limits. Serum calcium levels were 2.06 mmol/l and calcium in the 24 h urine was 7.88 mmol/dU. Serum angiotensin-converting enzyme level was raised to 74 U/L. Cerebrospinal fluid analysis revealed 53 cells per cubic milliter, 99% of them being lymphocytes, a protein level of 1.13 g/l, CSF glucose of 2.2 mmol/l and oligoclonal IgG bands were negative. Extensive serological investigation (including serology for brucellosis, toxoplasmosis, borreliosis, EBV, CMV, HHV6, VZV, WNF, and JCV PCR), bacterial and tuberculosis cultures and Quantiferon test were negative. The results of the bone marrow aspiration were normal. Arylsulfatase A, very long chain fatty acid and GFAP genetic analysis were normal. Chest CT demonstrated multiple enlarged lymph nodes bilaterally in middle and lower mediastinum. Fiberoptic bronchoscopy was done with biopsy of the enlarged lymph nodes, which showed noncaseating epithelioid granulomas. Whole body ⁶⁷gallium scan showed bilateral hilar lymphadenopathy uptake with lacrimal and parotid gland uptake. The patient was diagnosed with neurosarcoidosis and she was treated with methylprednisolone and cyclophosphamide. She showed gradual improvement on follow-up six months later and significant resolution of diffuse MRI lesions.

Discussion

Our patient was diagnosed as “probable” neurosarcoidosis according to the accepted criteria (suggestive symptoms, positive findings on extra-CNS biopsy, laboratory results of CNS inflammation, exclusion of other diseases).

The differential diagnosis in presented patient is comprehensive. Tuberculosis was excluded with negative CSF cultures and Quantiferon test. Histiocytosis of Langerhans and non Langerhans type (Erdheim-Chester disease) were considered, however the absence of bone lesions and normal bone marrow argued against these diagnoses. Leukodystrophies or other toxic and metabolic causes were excluded with appropriate tests.

MRI can detect neurological lesions in 10% of patients with sarcoidosis and is the imaging modality of choice in the diagnosis and follow-up of nervous involvement of sarcoidosis.(1) The pathologic hallmark are leptomeningeal inflammation or parenchymal findings in a form of mass lesions or granulomas.(2) While the intraparenchymal T2 hyperintense lesions are the most common finding on brain MRI there are several characteristic MRI patterns that can help us in the diagnosis of neurosarcoidosis.(3) These include: 1) periventricular and white matter lesions on T2 sequences, mimicking multiple sclerosis, 2) single or multiple supratentorial and infratentorial brain lesions with various amounts of vasogenic edema, mimicking tumors and 3) leptomeningeal and ependymal enhancement usually in the basal, hypothalamic and periventricular regions. (4) Diffuse involvement of the brain parenchyma is rarely observed in neurosarcoidosis, and when present can pose a diagnostic difficulty. We performed MR spectroscopy of the diffuse

T2 hyperintensity in our patient on two locations but gained contrary results. NAA is a neuronal marker and decreases with any disease that adversely affects neuronal integrity, while Choline is a measure of increased cellular turnover and is elevated in tumors and inflammatory processes. Therefore these findings were of limited value in presented case and systematic studies on MR spectroscopy in neurosarcoidosis are lacking. Other imaging possibility is FDG-PET which was shown helpful in one case.(5)

Conclusion

In conclusion, although T2-hyperintense white matter lesions are not uncommon they are rarely diffuse and found bilaterally in brainstem as presented here. This report further broadens MRI spectrum of neurosarcoidosis.

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Figures

Figure 1.

Brain MRI, a,b,c) T2 sequences and d,e,f) FLAIR sequences showing diffuse hyperintensity of the periventricular and deep white matter. Symmetrical hyperintensity in the deincephalic-mesencephalic boundary, tegmentum of the pons, middle cerebellar peduncle and hylus of the nucleus dentatus is seen. There is no postcontrast enhancement.

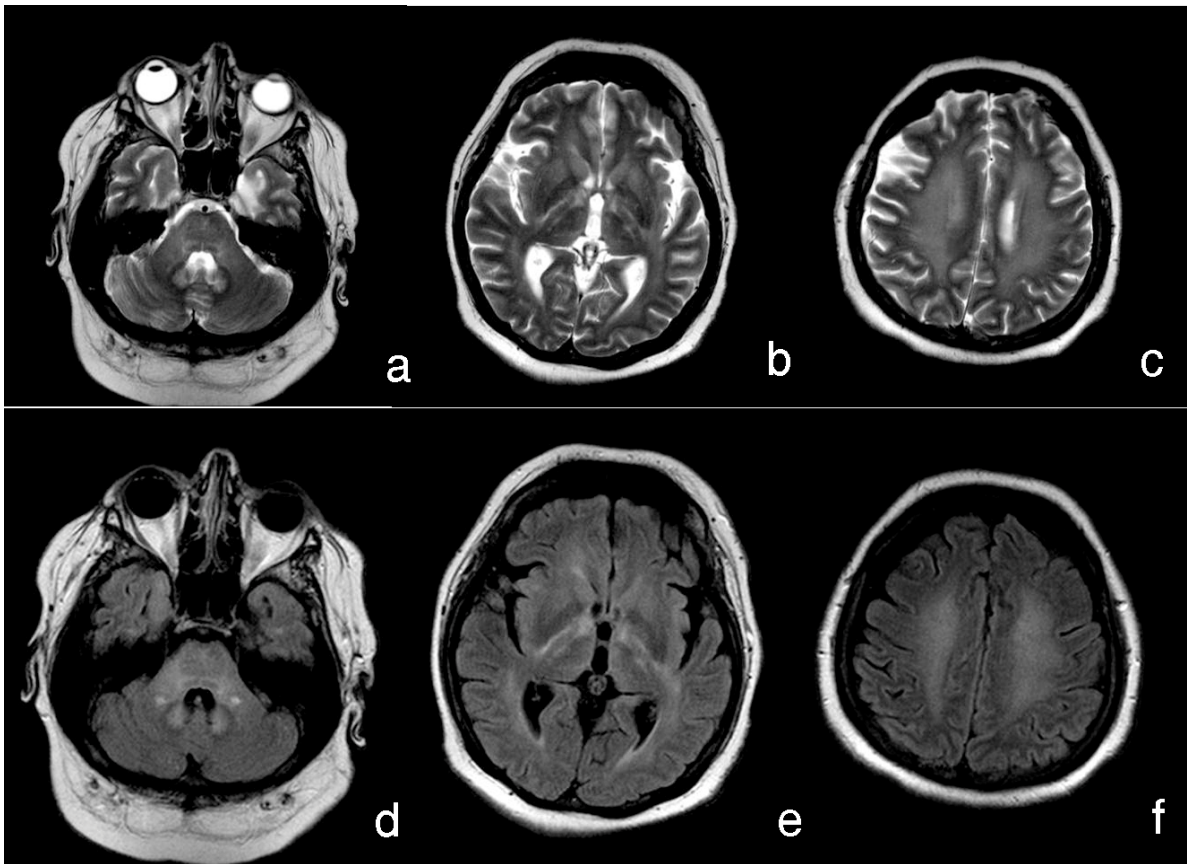


Figure 2.

MRI spectroscopy in the deep white matter showing a) increased Choline in the frontal regions and b) reduced NAA in the parietal region.

