Neurosarcoidosis - the Role of Magnetic Resonance Imaging in Diagnostics

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CASE REPORT

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ABSTRACT
Background: Sarcoidosis is a multisystem granulomatous disease of unknown etiology, characterized by presence of granulomas in affected tissues with variety in clinical presentations and presents a differential diagnostic and therapeutic dilemma. Clinical presentation of neurosarcoidosis is very variable. Diagnosis is based on clinical and radiological criteria and histological findings of disseminated non-necrotic granuloma followed by negative cultures for bacteria and fungi. MRI plays a key role in detection of lesions located in the brain parenchyma. Objective: The aim of this article was to present case of a 36-year-old male patient, who came to doctor with symptoms of fever, dry cough with whitish sputum, lymphadenopathy of neck region and neurological disturbances in form of headaches and vision problems. Case presentation: Patient underwent on pulmonary examination and results indicated presence of sarcoidosis. CT examination was performed (SIEMENS Somatom Definition AS, Erlangen, Germany), which confirmed presence of mediastinal and hilar lymphadenopathy. Ultrasound (US) examination of a neck region was also performed showed significantly enlarged and morphology altered lymph nodes. After biopsy of several neck lymph nodes, histopathological was proven diagnosis of sarcoidosis. Due to neurological disturbances in form of headaches and vision problems patient was examined by an ophthalmologist, neurologist and endocrinologist. Hormonal analysis showed an increase of prolactin and that raised suspicion for neurosarcoidosis. In further diagnostic evaluation it was indicated MRI examination of the brain with focus on sellar region. Conclusion: Contrast-enhanced MRI is the modality of choice for investigating suspected neurosarcoidosis. The versatility of MR recording and the amount of diagnostic informations obtained from MRI examination is huge. Comparison of MRI sequences obtained, facilitate interpretation of these findings. Obtained MRI information and available literature, correlating with other diagnostic modalities (ultrasound and CT) facilitate understanding of the specific pathology.

Keywords: Sarcoidosis, neurosarcoidosis, MRI.

1. BACKGROUND

Neurosarcoidosis is a rare form of sarcoidosis that affects the nervous system. Sarcoidosis is a multisystem granulomatous disease of unknown etiology, characterized by the presence of granulomas in affected tissues and which has variety in clinical presentations and presents a differential diagnostic and therapeutic dilemma (1). Granulomas may be present in all organs and tissues, including the nervous system. Symptoms are numerous, non-specific and they depend on which organ or organic system is affected by disease. The most common symptoms are fever, anorexia and weight loss, appearance of skin granulomas, cough and cardiac failure (1). Pulmonary form of sarcoidosis is presented in 95% of patients. Incidence of disease is about 10 patients per 100,000 inhabitants (2). Clinical presentation of neurosarcoidosis is very variable. Neurologic manifestations can occur in approximately 10% of all patients with sarcoidosis, and peripheral neuropathy is presented in 5-10% of patients (3-6). Clinical criteria for diagnosis of neurosarcoidosis are defined by interna-
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Sarcoidosis is a disease that is extremely difficult to diagnose. There is no specific test for sarcoidosis, but some studies indicate importance of elevated value of angiotensin converting enzyme (ACE), elevated value of calcium and biopsy (1, 8).

For diagnosis of this disease requires evidence of presence of systemic disease and exclusion of other neurological manifestations presented in other systemic diseases. Diagnosis is based on clinical and radiological criteria and histological findings of disseminated non-necrotic granuloma followed by negative cultures for bacteria and fungi.

MRI is a new non-invasive technique for displaying body structure without use of ionizing radiation. This is a highly sophisticated imaging diagnostic technique with a high diagnostic sensitivity and specificity for many pathological conditions.

Using Diffusion Weighted Imaging (DWI) brings a new possibility for extending capabilities for magnetic resonance imaging which can provide additional informations about changes visible on standard MRI sequences (9).

2. OBJECTIVE

The aim of this article was to present case of a 36-year-old male patient, who came to doctor with symptoms of fever, dry cough with whitish sputum, lymphadenopathy of neck region and neurological disturbances in form of headaches and vision problems.

3. CASE PRESENTATION

Patient underwent on pulmonary examination and results indicated presence of sarcoidosis. CT examination was performed (SIEMENS Somatom Definition AS, Erlangen, Germany), which confirmed presence of mediastinal and hilar lymphadenopathy. Ultrasound (US) examination was also performed (GE Voluson 730 ultrasound system). US examination of a neck region showed significantly enlarged and morphology altered lymph nodes. After biopsy of several neck lymph nodes, histopathological was proven diagnosis of sarcoidosis. Due to neurological disturbances in form of headaches and vision problems patient was examined by an ophthalmologist, neurologist and endocrinologist. Hormonal analysis showed an increase of prolactin and that raised suspicion for neurosarcoidosis.

In further diagnostic evaluation it was indicated MRI examination of the brain with focus on sellar region.

MRI examination was performed (SIEMENS Magnetom Essenza, 1.5 T, Erlangen, Germany) with MR protocol which included use of axial and sagittal plain T1W sequence, and T2W, FLAIR, HEMO axial sequences. Gadolinium contrast media was administrated in T1W sequence and we made dynamic analysis in all three plains. MR examination was supplemented with diffusion (DWI) sequences. On obtained MR images, in a supratentorial region, in visible area of the hypothalamus we noted T2W and FLAIR hyper intense lesion with approximative dimensions 14x11 mm (apx(ec) which showed intense post-contrast enhancement (Figure 1).

Both optical tracts had higher signal intensity on T2W and FLAIR sequences (Figure 2). In brain parenchyma, left and temporal, we spotted another (stipe like) area of increased T2W and FLAIR signal intensity with approximately 11x4 mm in diameter, which showed post-contrast enhancement (Figure 3). Described changes showed no signs of restriction of diffusion. In left temporoparietal area and closer to cortex on post-contrast sequences were visible few smaller hyper intense strip areas which did not have convincing correlates due to other sequences. Significant pathotmorphological changes in orbital region were not noticed. MR findings of pituitary region were not noticed.
gland, including infundibulum were normal. Optical chiasm had normal anatomical and morphological characteristics with higher signal intensity on T2W sequence.

4. DISCUSSION

Neurosarcoidosis can be asymptomatic and symptomatic, can be life-threatening, usually occurs in cases with systemic involvement, with about 5 to 10 percent of patients with sarcoidosis having neurological complications (10). The case demonstrates typical features of neurosarcoidosis, which often require good quality contrast images to identify. This patient had systemic sarcoidosis diagnosed on histology from a neck lymph nodes biopsy. Described changes in hypothalamus and in left temporal area correspond with infiltrative changes within neurosarcoidosis. In the patient with systemic sarcoidosis who develops new neurological symptoms and signs, the likelihood of any such neurological presentation being due to sarcoidosis is high with the proviso that individuals who have received immunosuppression as part of their previous sarcoid treatment may be at greater risk of CNS infections, which, therefore, must be rigorously excluded (11).

Lesions in neurosarcoidosis appear as:
- Parenchymal form—lesions DDx can remind as a MS (located in periventricular and subcortical white mass),
- Meningeal affection with a uniform dural thickening and post contrast signal enhancement,
- Focal infiltration of hypothalamus and/or pituitary stalk,
- Cranial neuritis (usually C II and C VII) (12).

Contrast-enhanced MRI is the modalit of choice for investigating suspected neurosarcoidosis (13).

Pachymeningeal disease often takes the form of pachymeningeal thickening with homogeneous postcontrast enhancement. Leptomeningeal involvement is characterized by prominent postcontrast changes may be inapparent on other sequences. Although pituitary and hypothalamic involvement are frequently seen as part of a more extensive leptomeningeal disease, it may also be encountered in isolation. Cranial nerves may be involved either as part of a more widespread leptomeningeal disease or in isolation. Parenchymal involvement is the most common finding and can be in many forms (14, 15).

Neurosarcoidosis has a wide spectrum of imaging features that mimic both benign and malignant conditions. Virtually any portion of the central nervous system and associated structures can be affected. MRI is highly sensitive for detecting neurosarcoidosis, but is not specific. Finally, the diagnosis is made by exclusion of other entities using a combination of imaging, diagnostic testing, clinical presentation, and sometimes tissue sampling (16).

Other differentials to consider for neurosarcoidosis include primary brain tumors, toxan ingestion, neuropathies secondary to monoclonal proliferation or paraproteinemic neuropathy, paraneoplastic neuropathy, vitamin deficiency, leptomeningeal carcinomatosis, central nervous system (CNS) lymphoma, or autoimmune conditions like systemic lupus erythematosus (10).

5. CONCLUSION

Contrast-enhanced MRI is the modality of choice for investigating suspected neurosarcoidosis. The versatility of MR recording and the amount of diagnostic informations obtained from MRI examination is huge. Comparison of MRI sequences obtained, facilitate interpretation of these findings. Obtained MRI information and available literature, correlating with other diagnostic modalities (ultrasound and CT) facilitate understanding of the specific pathology.

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