Neurosarcoidosis as an atypical Onset of Systemic Disease

Anetta Lasek-Bal1, Mike Smertka2, Małgorzata Piwkowska3, Małgorzata Cisowska-Babraj1 and Michał Holecki2*

1Department of Neurology, Medical University of Silesia Hospital, Poland
2Department of Internal Medicine and Metabolic Diseases, Medical University of Silesia Hospital, Poland
3Department of Radiology, Medical University of Silesia Hospital, Poland

CLINICAL IMAGE

We present a 28 year old male patient with a primary neurosarcoïdosis. His initial symptoms found on physical exam include acute onset of sensorimotor aphasia on the right side (central nerve paresis) of the face, as well as the right upper extremity, with pain localized in the occipital region. Computer Tomography (CT) of the head revealed a 20 cm tumor in the left fronto-parietal region with extensive edema, prompting a potential diagnosis of neoplastic disease. Further diagnostics with magnetic resonance imaging was suggestive of meningioma with malignant transformation (Figure 1a,1b). Steroid therapy was initiated resulting in rapid clinical improvement and the tumor removed by craniotomy. Subsequent histopathology surprisingly revealed an inflammatory tumor containing tuberculoid granulomas surrounded by fibrous tissue bands. Subsequent evaluation excluded tuberculosis and syphilis from the differential diagnosis. CT of the chest (Figure 2), along with bronchofibroscopy, and histopathology established the diagnosis of sarcoïdosis.

DISCUSSION

Sarcoidosis is a granulomatous disorder of unknown etiology affecting multiple organs, most commonly the lungs, but can also involve the nervous system (in approximately 5% of cases) [1]. Isolated neurological disease accounts for approximately 1% of individuals with sarcoïdosis [2]. Neurological symptoms are rare in sarcoïdosis; however the post mortem examinations

reveal nervous system involvement in 27% of all cases [3].
Sarcoidosis can affect any location in the central nervous
system and manifests by a broad spectrum of symptoms and
radiological findings. The most common neurologic symptoms
are associated with cranial neuropathy, especially the facial
nerve, optic nerve, trigeminal nerve, or a combination thereof.
Additionally neurosarcoidosis may present in the form of aseptic
meningitis, hydrocephalus, seizure, neuropsychiatric syndromes,
and neuroendocrine dysfunction [4]. The common radiological
findings include infiltration of the dural matter, subarachnoid
space, hypothalamus, pituitary, and chiasma of the optic nerves
area with gadolinium-enhancement [3].

There is great difficulty in diagnosis sarcoidosis until the
symptoms of systemic disease presents and it is very rare that
a biopsy of neural tissue is available for diagnosis. For example,
in the ACCESS study, biopsies were performed from tissues
originating in the lung, lymph nodes, hiliius, bronchi, and trachea.
In the events of extra pulmonary origin, biopsy tissue was often
taken from skin, peripheral lymph nodes, and the liver [5].

Due to the broad clinical spectrum of sarcoidosis, including
central neural origin, sarcoidosis should be considered in the
differential diagnosis of atypical neurological presentations to
facilitate rapid diagnosis and initiation of appropriate therapy.

REFERENCES
1. Segal BM. Neurosarcoidosis: diagnostic approaches and therapeutic
Semin Respir Crit Care Med. 2010; 31: 419-427.
4. Tabuchi S, Uno T. Hydrocephalus with panventricular enlargement as
the primary manifestation of neurosarcoidosis: a case report. J Med
5. Teirstein AS, Judson MA, Baughman RP, Rossman MD, Yeager H Jr,
Moller DR. Case Control Etiologic Study of Sarcoidosis (ACCESS)
Writing Group. The spectrum of biopsy sites for the diagnosis of