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Case report

Isolated angiitis of the central nervous system: A case presented with atypical psychiatric symptoms

Cicek Hocaoglu[a,*], Meliha Tan[b]

[a]Department of Psychiatry, Karadeniz Technical University Medical School, Trabzon, Turkey
[b]Department of Neurology, Karadeniz Technical University Medical School, Trabzon, Turkey

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Abstract

Isolated angiitis of the central nervous system (IACNS) is a rare form of angiitis limited to the central nervous system. The clinical finding of the combined series revealed that headache was the most common symptom within a combination of focal and diffuse neurological deficits. The case, a 28-year-old man, is presented; the clinical presentation and diagnostic difficulties are discussed. The patient's symptoms began with an obvious atypical depression. In spite of an antidepressive treatment, his symptoms continued to worsen with personality, mood changes and euphoria added to the clinical picture. Meanwhile after several transient ischemic attacks, 6 months later, he was admitted with neurological symptoms including headache, diplopia, and cerebellar ataxia. The radiological investigation was mimicked by primary brain lymphoma. The brain biopsy excluding of lymphoma revealed parenchymal hemorrhage with nonspecific degenerative changes. In systemic investigation, no underlying cause for vasculitis could be found. Neurological but not psychological deficits and radiological lesions of the patient improved with steroid therapy. Since we could not find features of systemic vasculitis, the patient's lesions responded to corticosteroid treatment and neuropathological investigation revealed no lymphoma, We concluded that the most probable diagnosis would be IACNS.

Keywords: Atypical psychiatric symptoms; Diagnosis; Isolated angiitis of the central nervous system; Vasculitis

1. Introduction

Isolated angiitis of the central nervous system (IACNS) is an inflammatory vascular disease of unknown origin, restricted to the CNS which responds to immunosuppressive treatments well (Moore and Richardson, 1998; Fieschi et al., 1998;Younger, 2004). Historically, the disease was called granulomatous angiitis on the basis of granulomas present on postmortem examination. But in antemortem pathologic investigations granulommas are not always found. Also, in vasculitis which develops secondarily related to some viral and neoplastic conditions intracranial granulommas can be seen. Therefore 'isolated angiitis of the CNS' is the suggested term (Moore and Richardson, 1998). The symptoms of IACNS are typically persistent headaches, encephalopathy and multifocal signs. Strokes may be prominent in some patients. In the absence of pre-existing disease, behavioural or psychiatric features are recognised increasingly. For ruling out systemic vasculitis, wide range of clinical and laboratory investigations must be made. Definite diagnosis is made by brain biopsy including cortical and leptomenengial vessels (Wiebers et al., 1997; Greenan et al., 1992, Hashimoto et al., 2002).

We present a 28-year-old man with atypical psychiatric symptoms, transient ischemic attacks and multiple strokes. In the light of current literature, we discussed the clinical and laboratory findings.

Abbreviations: ANCA, antineutrophil cytoplasm antibodies; CSF, cerebrospinal fluid; CT, computerized tomography; DSM-IV, Diagnostic and Statistical Manual of Mental Disorders, fourth edition; EEG, electroensalography; EKG, electrocardiography; IACNS, isolated angiitis of the central nervous system; MRI, magnetic resonance imaging; MS, multiple sclerosis; tbc, tuberculosis.

* Corresponding author. Karadeniz Teknik Universitesi Tip Fakultesi Psikiyatri AD, 61080 Trabzon, Turkey. Tel.: +90 462 3775393.
E-mail addresses: cicekh@meds.ktu.edu.tr (C. Hocaoglu), chocaoglu@veezy.com (M. Tan).

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2. Case report

A 28-year-old man, a university student, was admitted to our psychiatry policlinic with symptoms of oversleeping, not enjoying life, boredom, finding life meaningless, not attending school since 1998. According to the Diagnostic and Statistical Manual of Mental Disorders, fourth edition (DSM-IV) (American Psychiatric Association, 1994), the diagnosis was major depression and treatment begun with alprazolam 0.5 mg/day and sertralin 50 mg/day. Within 30 days, he had two transient attacks with diplopia and severe headaches, especially in the frontal region.

He was hospitalized because of sudden diplopia, nausea, and vomiting. He had restricted abduction and diplopia in his right eye. He was afebrile, there was no meningism, and general examination was normal. No fever, chills, or other clinical evidence of infection was observed. Routine laboratory studies, EEG (electroensalography), EKG (electrocardiography), hematologic investigation and sedimentation rate were normal. A cranial computerized tomographic (CT) scan, performed without and with the intravenous injection of contrast material, was negative. One day, his diplopia and neurological signs had disappeared. He was discharged from the hospital with his antidepressive treatment. In spite of the treatment, his symptoms continued, being relatively worse with personality, mood changes, euphoria and increasing agitation added to the clinical picture. He made obscene jokes to ladies and family members. Thioridazin 200 mg/day was administered for these psychotic symptoms by a psychiatrist. Six months later, he was admitted to a neurology policlinic because of suddenly developed right oculomotor nerve palsy, left hemiparesis, dysarthria and right cerebellar ataxia. An MRI (magnetic resonance imaging) examination of the brain showed a 3×4×3 cm diameter hypo-intensity on T1-weighted images and hyper-intensity on T2-weighted images in the right thalamus and basal ganglia. After the administration of gadolinium, there was no enhancement. The patient was sent to another hospital for brain biopsy, because of a probable primary brain lymphoma. Biopsy revealed intra-parenchymal hemorrhage and excluded lymphoma. However, no vessels could be observed in small tissue samples. In the hospital, the patient was given steroid treatment, after which the palsy of the third cranial nerve completely and the cerebellar signs as well as the left hemiparesis were partially improved. In May 1999, he was re-examined by one of us. After that, we performed all of the clinical and laboratory investigations such as lupus anticoagulant, antiphospholipid antibodies, antineutrophil cytoplasm antibodies (ANCA), autoantibody screen, antinuclear factor, hemoglobin S and C, homocystine, carotid and vertebrobasilar ultrasound, coagulation screen, EEG, ECG syphilis, HIV and hepatitis, to examine the possible causes of the systemic vasculitis and the other causes of stroke in young adults. All of these investigations were found to be within the normal ranges.

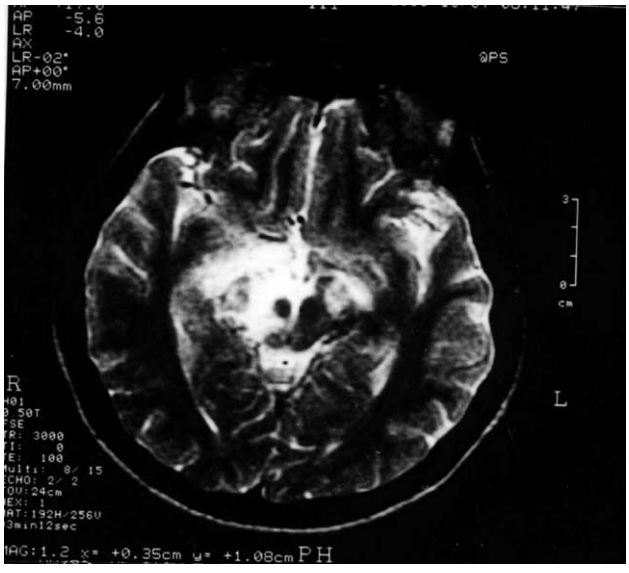

Fig. 1. In the second attack of the patient's illness, foci of increased signal intensity on the T2-weighted images.

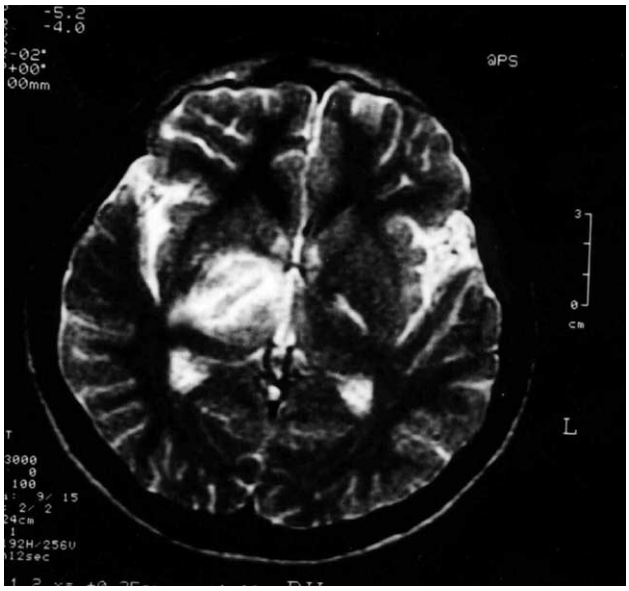

Fig. 2. In the second attack of the patient's illness, foci of increased signal intensity on the T2-weighted images.

After 5 months following this illness, the neurological findings became worse. An MRI examination of the brain (Figs. 1 and 2) revealed foci of increased signal intensity on T2-weighted images in the right midbrain and a major portion of the pons, extending from temporal lobe to posterior limb of internal capsule and within the posterior limb of the left capsula interna. The coronal T1-weighted images showed a hypo-intense area with small foci of hyper-intensity within the right temporal lobe extending to thalamus (Fig. 3).

The patient was admitted a second time to the hospital where the biopsy was previously made. He was given steroid again, after which he improved considerably. A