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Olgu Sunumu

Primary Central Nervous System Lymphoma (PCNSL) Presenting as Behçet's Disease: A Case Report

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Primary central nervous system lymphoma (PCNSL) may be hardly distinguishable both clinically and radiologically from some white matter diseases, such as Behçet's syndrome, multiple sclerosis, demyelinating or inflammatory disorders. Misdiagnosis of these cases leads to delay the accurate treatment of PCNSL that is one of the most malignant tumors. This report describes a rarely detected case of PCNSL mimicking a neuro-Behçet disease in young patient. The patient's MRI and clinic onset as same as his familial history (his father was diagnosed as Behçet disease, depression and died at an early age of 51) lead to misdiagnose PCNLS as Behçet disease. No improvement was noticed after his prescribed medical treatment. This led his doctors to decide performing new MRI. Repeated MRIs showed progression of the initial lesions. The neurosurgical team decided to perform biopsy under stereotactic guidance. Histopathological examination demonstrated that the lesion was diffuse large B-cell lymphoma. Herein, the neurosurgical team points out the importance of the stereotactic biopsy to distinguish PCNSL from other white matter diseases as well as to avoid the complications that may result from open craniotomy surgery in such deep located lesions.

Keywords: Large B-cell lymphoma, Behçet disease, stereotactic biopsy

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Radyolojik Olarak Behçet Hastalığı Düşünülen Beyin Lenfoma: Olgu Sunumu

Primer santral sinir sistemi lenfomalarının (PCNSL), klinik ve radyolojik olarak multiple skleroz, demyelinizan hastalıklar, inflamatuar hastalıklar ve Behçet hastalığı gibi ak madde hastalıklarından ayırt edilmesi zor olabilir. Bu olgularda yanlış teşhis koymasıyla PCNSL gibi malign tümörlerin doğru tedavi yaklaşımından uzak kılmaktadır. Bu makale, ender olarak saptanıp Behçet hastalığı olduğu düşünülen PCNSL'li genç hasta sunmaktadır. Hastanın MRG'si ve soygeçmişi (babsında Behçet hastalığı ve depresyon tanısı konulup erken yaşta ölmüştü (51) doğru tanısı geciktirmiştir. Hastanın doktorları tarafından yeni MRg'si çekilmiş, Behçet hastalığın tedavisine yanıt vermemiş ve nöroşirürji hekimleri tarafından hastadan biyopsi alınmasına karar verilmiştir. Stereotaktik biyopsi alınarak histopatolojik testlerinde diffüz B-hücreli lenfoma tanısı konulmuştur. Bu makale amacı, derin yerleşimli lezyonlarda biyopsi alınmasının önemi vurgulayarak açık kraniyotomi cerrahi görülebilecek komplikasyonlarında uzak tutulmasına yardımcı olabilmektedir.

Anahtar kelimeler: B hücreli primer lenfoma, Behçet hastalığı, stereotaksik biyopsi

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INTRODUCTION

PCNSL may be indistinguishable clinically and radiologically from some white matter diseases, including Behçet's disease (BD), multiple sclerosis, demyelinating or inflammatory disorders [1,2]. Establishment of diagnosis in such cases is difficult. To determine the final diagnosis, neurosurgeons often use biopsy procedures rather than less invasive means of diagnosis such as LP or vitrectomy [7]. Unfortunately, delayed diagnosis and treatment of PCNSL can negatively affect the diagnosis.

Neuro-Behçet disease is one of the most aggressive diseases that mimick a PCNSL. Behçet syndrome (BS) is a multisystemic, chronic, relapsing, vascular inflammatory disease of unknown etiology [11]. In this syndrome there is involvement of CNS in about 5% of the cases [10]. This report describes a rare case of PCNSL mimicking a neuro-Behçet disease.

CASE REPORT

A 26- year-old right-handed man had been referred to emergency department with right hemiparesis and acute loss of the sensation of the right side, after tooth extraction before three weeks ago. Neurological examination showed motor deficit in the right side which was evaluated as grade 3/5 weakness in the upper and grade 4/5 weakness in the lower limb, hypoesthesic right upper facial spasm, diplopia associated with bilateral restriction of upward gaze, right central facial palsy, and gait impairment.

Radiological investigation revealed multiple hyperintense lesions on T1, T2, and Flair weighted images in both of the left and right thalamus, as well as mildly hyperintense lesions in all of the corona radiata, centrum semiovale, frontoparietal lobe, periventricular and subcortical white matter (Figure 1). The radiologists was suspected the presence of neuro-Behçet disease or malign tumor.

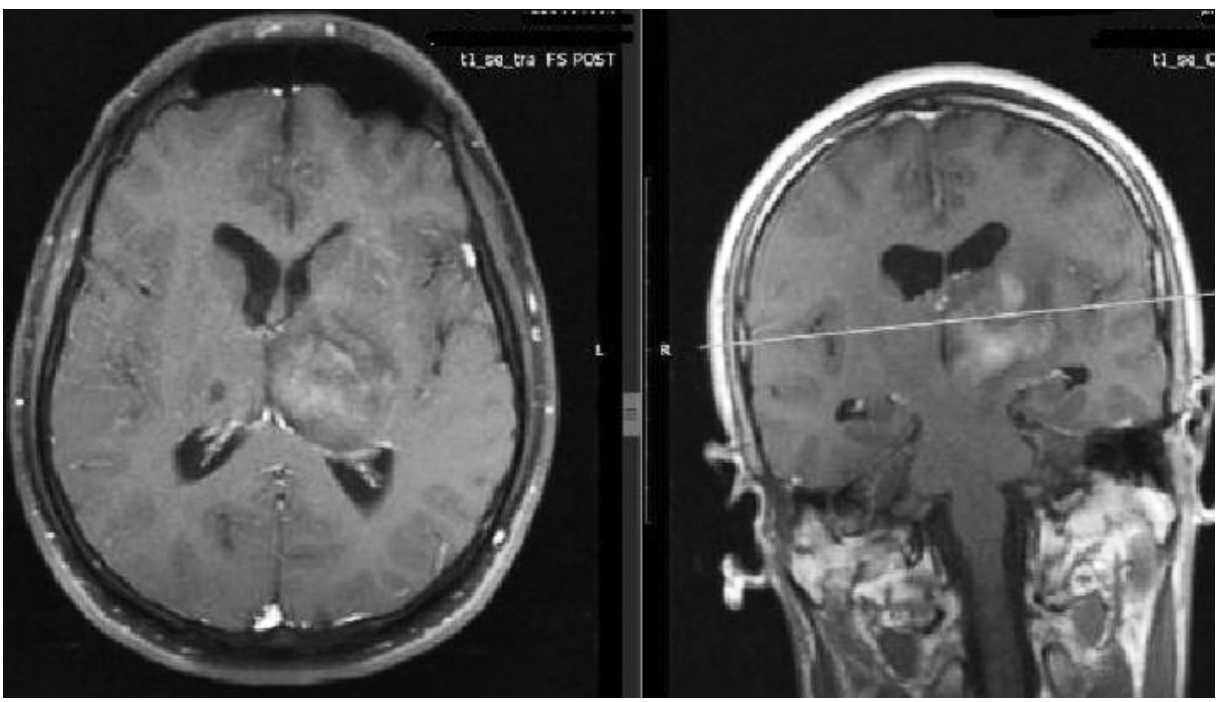

Figure 1. Contrast-enhancing MRI; T1- weighted images showed mild contrast enhanced multiple hyperintense lesions in both of the left and right thalamus.

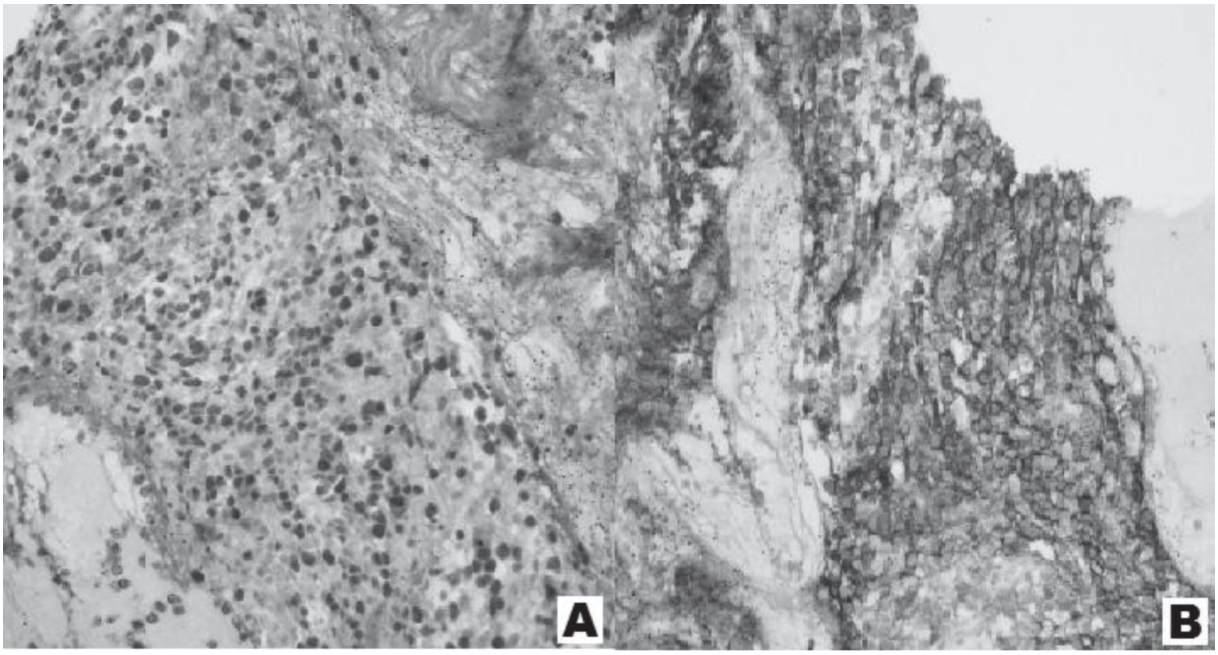

Figure 2. Histopathological examination shows; hypercellular nodules showing a MIB labeling (Ki-67) index of 70-80%, all of CD3, CD2, CD5, CD7, CD34, and CD20 were positive. [A]: CD20 (+) >80%; [B]: pax-5 (+); that is consistent with diffuse large B-cell lymphoma.

His father died at an early age of 51 after diagnosed as BD when he was on medication for major depression. His past medical history was unremarkable. His laboratory test results (white blood cell count: 4900/mm^3; C-reactive protein (CRP): 5.71 mg/dL, and erythrocyte sedimentation rate: 6 mm/h, 15 mm/2 hours) did not indicate any evidence of increased inflammatory reaction. Serologic tests for syphilis, antinuclear antibody, anti-DNA antibody, and Brucella agglutination test (Rose Bengal) were negative. The immune and complement systems were normal. The angiotensin-converting enzyme (ACE) level was within normal limits. CSF showed a mild pleocytosis of 33.6 mm^3 (all mononuclear cells) and normal levels of protein, sugar, chlorine, and myelin basic protein, and oligoclonal bands. Neither atypical cells nor bacteria were found cytologically in the CSF. But, human leukocyte antigen-B51 (HLA-B51) was positive.

After evaluation all these, the patient had been diagnosed as BD. Therefore, he received steroid therapy (high doses of dexamethasone) for 15 days, but any clinical response could not be detected. So his attendant physicians decided to perform new MRI and Spectroscopy MRI. Repeated MRI showed progression of the initial lesions. Spectroscopy MRI showed decreased N-acetylaspartate, increased choline levels , and minimally increased inositol and glutamine-glutamax complexes. Whereas, lactate peak was seen, which firstly suggested the presence of glioma. Lymphoma and vasculitis as neuro-Behçet disease and a demyelinating disease were included in the differential diagnosis. The neurosurgical team decided to perform biopsy using stereotactic guide. Because of the deep localization of lesions - the biggest one being in the left thalamus -and to avoid major complication (such as seizure), the patient had undergone stereotactic biopsy under general anesthesia.

Histopathological examination demonstrated hypercellular nodules showing a MIB labeling (Ki-67) index of 70-80%, CD3, CD2, CD5, CD7, CD34, and CD20 positivities. CD20 was >80% which was consistent with pax-5 (Figure 2). The

patient was referred to oncology department. On follow-up after 12 months the patient was presented to emergency room with headache, and deterioration in his consciousness. It was learnt that patient did not receive radiotherapy for socioeconomic reasons. The patient died three months after he received radiotherapy.

DISCUSSION

PCNSLs account for 1 to 2% of all non-Hodgkin lymphomas and for 2 to 7% of all primary CNS tumors [4]. These tumors can involve the brain, spinal cord, meninges, or orbit. The incidence of PCNSL has increased threefold since the 1970s [5], and most of the cases consist of non-Hodgkin B-cell lymphomas [4]. In contrast, Hodgkin disease rarely involves the CNS [3].

Diagnostic criteria require recurrent oral aphthae plus any two of the following: genital ulcers or scars; uveitis or retinal vasculitis; skin lesions such as folliculitis; acneiform lesions, or erythema nodosa; and hyperactivity of skin to nonspecific physical insult such as pinprick (skin pathergy test) [8]. Other organ systems also may be involved including the gastrointestinal tract, blood vessels (mainly the venous side), and lungs [10]. This coupled with a similar variation in HLA B51 (human leukocyte antigen), which has been reported to be strongly associated with the disease in highly endemic areas [9,11,13].

Tension-type headache, depression and neurological complications of BD treatments are among the indirect neuropsychiatric consequences of BD [8,11]. Turkey and the Far East are considered as endemic BS regions [13]. Neurological complications in BD occur more commonly in males who have more severe course [11,13]. Among cases with neurologic involvement, the male to female ratio is nearly 4:1 [6].

In cranial MRI, lesions of BD are generally located within the brainstem, occasionally with extension to the diencephalon, or within the periventricular and subcortical white matter. The pattern of parenchymal lesions is suggestive of small-vessel vasculitis [9]. With the exception of the rarely occurring lesions only affecting periventricular and subcortical white matter, cranial MRIs in CNS BS are rarely confused with multiple sclerosis [11]. Our patient was a young male, the onset of disease, MRI findings, familial history of his father, and HLA B51 (+) resulted in misdiagnosis of the patient as BD.

Even with treatment, the prognosis of PCNSL is still poor [12]. After only whole-brain radiation therapy, the 2-year- overall survival rate is 28% [12]. Non-AIDS-related PCNSL frequently presents with multiple enhancing lesions in the periventricular white matter and with signs and symptoms that are similar to many white matter diseases. In such diseases, determination of the final diagnosis is difficult. Delayed diagnosis and treatment of PCNSL can negatively affect the prognosis. For socioeconomic reasons, our patient did not receive radiotherapy. Although the survival time for patients with a PCNSL who do not undergo treatment is approximately 3 months after diagnosis [12]. Our patient survived for 12 months till his family brought him to oncology department. Unfortunately, he died 3 months after he started to receive radiotherapy.

CONCLUSION

Although all of laboratory data, clinical features and radiological investigations were leading to the diagnosis of BD, the true diagnosis was made by biopsy. To avoid major or new complications stereotactic biopsy is preferred.

Competing Interest

The authors declare that they have no competing interests. All authors certify that they have NO

affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

Patient Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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