B Cell Immunoblastic Lymphoma of Central Nervous System: Case Report

Primer B Hücreli Santral Sinir Sistemi Lenfoması: Olgu Sunumu

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Abstract: Primary central nervous system lymphoma (PCNSL) is defined as the lymphoma effecting only spinal axis and cranium without systemic symptoms. PCNSL is very rare. Hystologic type of lymphoma that arising in the CNS is most commonly B cell lymphoma. In our study we reported a 47 years old female patient with B cell non-hodgkin's lymphoma of CNS with no evidence of lymphoma outside the CNS. There was no evidence of systemic acquired immundeficiency syndrome.

Key Words : Primary, central nervous system, B cell non-hodgkin lymphoma.

Özet: Primer santral sinir sistemi lenfoması (PSSSL), sistemik belirti olmaksızın sadece spinal bölge, intrakranial alanda yerleşen ve sınırlanan lenfoma olarak tanımlanır. PSSSL, oldukça nadirdir. Patolojik olarak B hücreli tipi santral sinir siteminde gözlenir. Çalışmamızda histopatolojik ve immünohistokimyasal olarak B hücreli diffüz non-hodgkin lenfoma tanısı alan, santral sinir sistemi yerleşimli, başka sistemik belirti ve bulguları olmayan, immün sistem bozukluğu bulunmayan 47 yaşında bayan olgumuzu sunduk.

Anahtar Kelimeler: Primer, Santral Sinir Sistemi, B hücreli lenfoma

INTRODUCTION

Primary central nervous system non-hodgkin's lymphoma (PCNSL) is defined as the lymphoma located to the cranial-spinal axis without systemic disease (5,6). Although PCNSL was previously a rare tumor several studies have reported an increase in the incidence over the past 10 to 15 years (8,10,15). The peak incidence is in the fourth through sixth decades of life, with a mean age of about 55 years.There is also an early peak incidence in the first decade (6,16). The median age at diagnosis of PCNSL is 31 years for patients with AIDS.

PCNSL may involve numerous regions, including the brain parenchyma, the meninges, the intradural spinal cord and the vitreous and the retina of the eye. PCNSL can occur in all areas of the brain. The cerebral hemispheres are the most

commonly involving sites, especially in the periventricular area (1,2,13,16). Although PCNSL is frequently seen as a solitary lesion in patients with AIDS. It is seen as multiple lesions in other groups with a percentage of 25% (6,9). It is usually originated from B-cells (2,3,4,6,7,8,10,14,15,16). Clinically PCNSL may present with a wide variety of signs and symptoms because of variations in tumor involvement of the brain. Some of the symptoms are because of elevated intracranial pressure (2,6,8).

CASE

A 47 year-old woman was admitted with a 15-day history of progressive bilateral weakness dominantly on the left side of her body and right side of her face.

Neurological examination showed central facial palsy on the right side, other cranial nerves were normal. Muscle strength was 2/5 on the left and 3/5 on the right side. Deep tendon reflexes were considered to be normal. There were no other neuropathological findings. Physical examination showed no pathological finding as well. CT revealed two focal mass lesions, with a size of 3x2 cm and in deep right parietal region 1x1 cm. in size (Figure-1). Homogenius enhancement was detected following IV contrast injection in the lesions and there was a shift about 1 cm.across the midline. MRI showed the same masses with mild surrounding eudema in T2-weighted images (Figure-2).

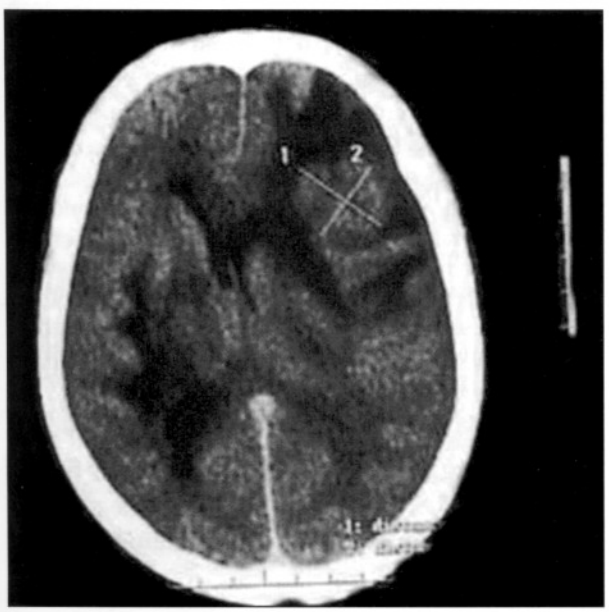

Figure -1 : Preoperative CT scan of the patient.

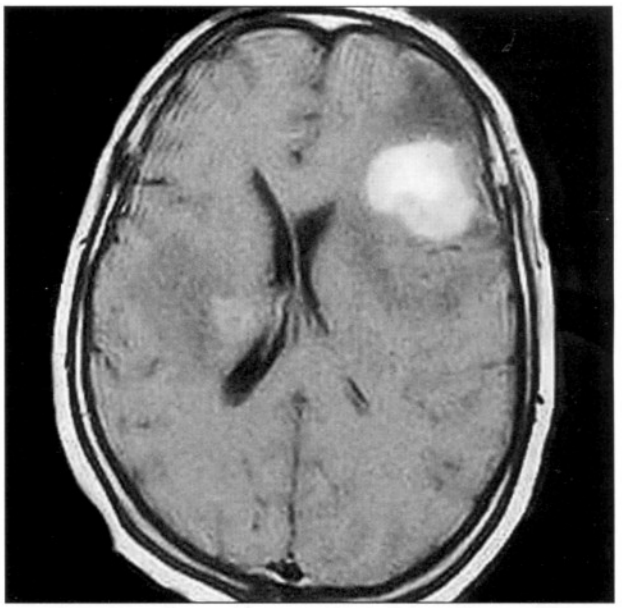

Figure - 2: Preoperative MR imaging of the patient.

The patient underwent left frontal craniotomy and subtotal removal of the tumor was performed. We did not perform any operation for deep right parietal mass.

Post-operatively slight improvement in muscle weakness as 3-4/5 on the left side, 4-5/5 on the right side were detected. Other neurological findings were the same.

Histopathological examination revealed that the tissue was composed of large lymphoid malignant cells infiltrating cerebral parenchyma diffusly and the cells had global or elyptoid shape with medium size cytoplasm. Some of the cells were in the form of dichrotic cells. Most of them had vesicular nuclei and some of them had visible nucleoli (Figure-3). It was seen that the tumor cells were stained with CD20 strongly and in diffusly (Figure-4). The patient was considered as B-cell diffuse non-Hodgkin primary central nervous system lymphoma and referred to hematology department for further treatment.

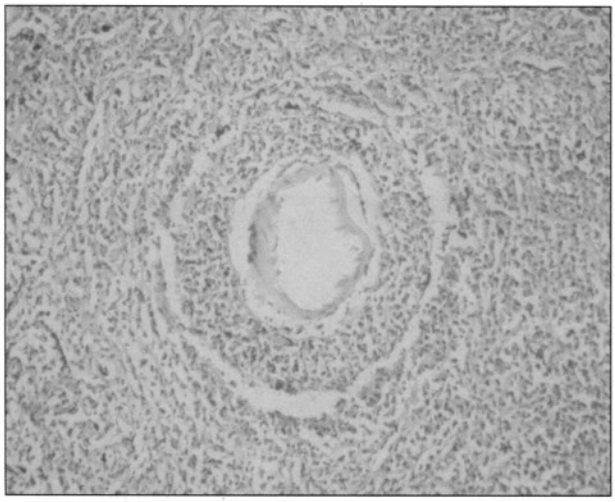

Figure – 3 Atypic lymphocyts in brain parecyhma (HE, x100)

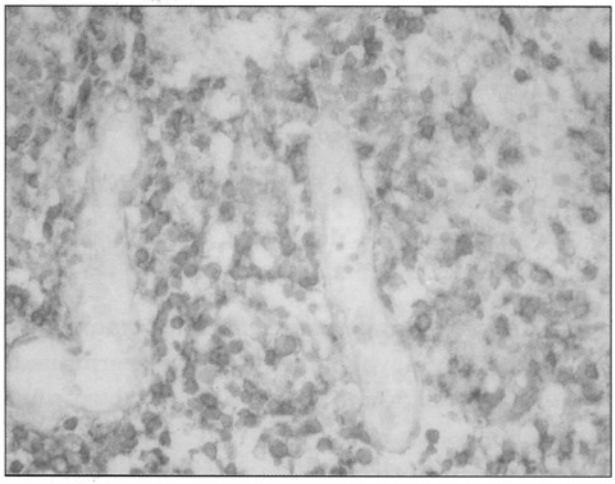

Figure – 4 : CD immunohistochemical stain for CD20 in tumor cells (x200)

DISCUSSION

Primary central nervous system lymphoma (PCNSL) is defined as the lymphoma effecting only spinal axis and cranium without systemic symptoms (5,6).

PCNSL is a rare condition. It consists less than 5% of all intcranial tumors and 1-2 % of all lymphomas (2,6). Several studies have reported an increase incidence because of AIDS in last 10-15 years (8,10,15). There is a correlation with Epstein-Barr Virus (3,7,10,11). It has been also reported that there is an increase at incidence in patients with organ-transplantation because of immune suppresion. Although PCNSL is usually seen in patients with congenital or acquired immunodeficiency, it can also be seen in patients with adequate immunity and with no risk factors (3,6). In our case we could not find any congenital or acquired immunodeficiency or systemic risk factors after hematological and serological evaluation.

The mean age for PCNSL is 55 year. The incidence is higher in the sixth decade and in the first decade respectively (6,16). Our case was 47 years old.

Variable clinical symptoms are seen according to the involved site of central nervous system. The symptoms are usually the result of the mass effect of the tumor or intracranial pressure (ICP) increase. Clinical presentation may occur in less than three months. Regarding focal neurological symptoms and / or mental deteroriation, epileptical seizures or headache, vomitting-nausea related to ICP increase (2,6,8). In our case, presentation of the clinical symptoms occurred in a even shorter period of time which was 15 days and the dominant symptoms and findings bilateraly were hemiparesis and central facial paralysis.

PCNSL can be located in spinal intradural space, cerebral parenchyma, meninges and even in retina and vitreus. Although PCNSL may be seen in all cerebral regions, it is frequently located in periventricular region of cerebral hemispheres (1,2,16). Brainstem involvement was also reported (13). In our case the lesions were located in frontal and deep parietal regions.

Although PCNSL is frequently seen as a solitary lesion in patient with AIDS, it is seen as multiple lesions in other groups with a percentage of 25% as far as the central nervous system is concerned (6,9).

Although PCNSL is usually originates from B cells (2,3,4,6,7,8,10,14,15,16). There are some other reports that pointed T cell as the origin of PCNSL (9).

In regard to therapeutical measures; surgery,

radiotherapy and chemotherapy can be used. Absolute curative therapy has not yet been found (1,2,6,8,12,15). Postoperative survey is 12-16 month and 5 year survey is as low as %5 (1,5,6).

CONCLUSION

Although PCNSL is rare entity found especially in patients with AIDS, Epstein Barr Virus infection,organ transplantation; it can also be seen in patients with no systemic disease and with adequate immune system such as our patient. Surgery for lesions with suitable localization and/or radiotherapy and chemotherapy are therapeutical choices. However, survival is not so long despite of all these treatment modalities.

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