

Case Report

A Rare Presentation of Ovarian Lymphoma with Cerebral Leptomeningeal Metastasis: Clinical Insights and Treatment Outcomes

Nejmi Kiyamaz^{1*}, Nebi Yilmaz¹, Mustafa Kösem², Taner Yazici¹, Suleyman Alici³
and Abdulbaki Kozan¹

¹Department of Neurosurgery, University of Yuzuncu Yil, School of Medicine, Van, Turkey.

²Department of Pathology, University of Yuzuncu Yil, School of Medicine, Van, Turkey.

³Department of Medical Onkology, University of Yuzuncu Yil, School of Medicine, Van, Turkey.

Accepted 13 January, 2024

A 38 years old female patient is presented who applied to our neurosurgery clinic due to symptoms such as headache, ataxia, vertigo, vomiting and complaints related to a mass at the back side of her head. The examination of cranial magnetic resonance imaging (MRI) of the patient displayed a mass which obliterates the transverse sinus and extends from the scalp to the epidural space at a certain distance and to the occipital zone. The result of the biopsy report revealed an infiltration of malignant lymphoma. The patient received cyclophosphamide, adriablastine, vincristine and prednisolone for 1 - 5 days, at 21 days interval and as 6 cures. A complete response was obtained at the end of the 3rd cure. The patient was orientated for radiotherapy at the end of the sixth cure. This case is a rarely seen ovarian lymphoma and constitutes a sample for cerebral leptomeningeal metastasis which is a very rare condition.

Key words: Cerebral leptomeningeal metastasis, primary ovarian lymphoma.

INTRODUCTION

Nevertheless, in patients who are within the advanced stage of a lymphoma and leukemia the retention of organs of the female genital system such as the corpus and cervix of the uterus are frequently encountered; but these organs are rarely accepted as primary initiation sites. The primary ovarian location of a lymphoma is determined less than 1% (Chorton et al., 1974; Delgado et al., 1976; Monterroso et al., 1993) and the age range changes between 38 - 44 years. The majority of female genital system lymphomas are non - Hodgkin lymphomas according to their histological status. The metastatic diffusion of the systemic lymphoma into the cerebral parenchyma occurs generally during the late stage and the frequency is between 1 - 7%. Metastases commonly occur as leptomeningeal infiltration (Jellinger et al., 1976). The patient presented in this study is a rare case diagnosed with cerebral leptomeningeal metastasis due

to a primary ovary lymphoma.

CASE REPORT

A 38 years old female patient referred to our outpatient clinic because of symptoms including headache, ataxia, vertigo, vomiting and a complaint related to a mass at the back side of her head. The patient was operated six weeks ago because of an adnexial mass in the obstetric and gynecology clinic where she was referred due to abdominal pain and irregular menses that was continuing for a period of one year. During the operation trans - abdominal hysterectomy and bilateral salpingo- oophorectomy was carried out whereas the pathology report of the patient displayed a high-graded diffused large cell lymphoma (Figures 1 and 2). The neurological examination of the patient indicated Romberg (+) abnormal cerebellar tests including ataxia. Physical examination demonstrated a, soft mass with a diameter of 7 × 6 cm at the occipital mid-line. Contrast brain MRI and MR Angiography showed a mass was assessed at the

*Corresponding author. E-mail: nejmikiyamaz@yahoo.co.uk. Tel: 90.432.2164710/204190.532.4160980. Fax: 90.432.2167519.

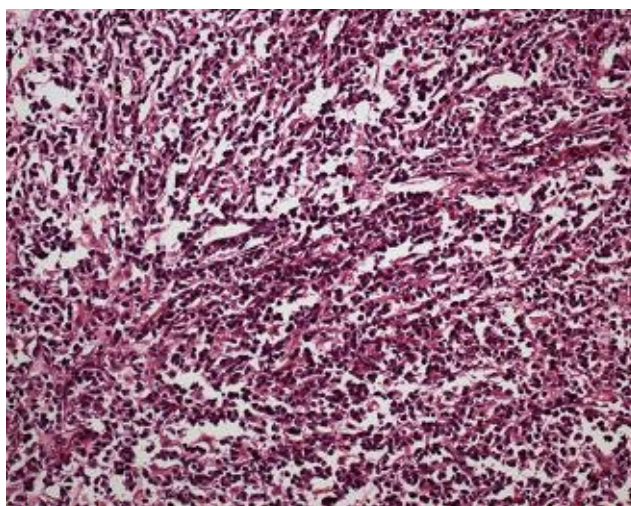


Figure 1. Diffuse staining for CD20 in tumor cells in ovarian tumor (Immunoperoxidase x200).

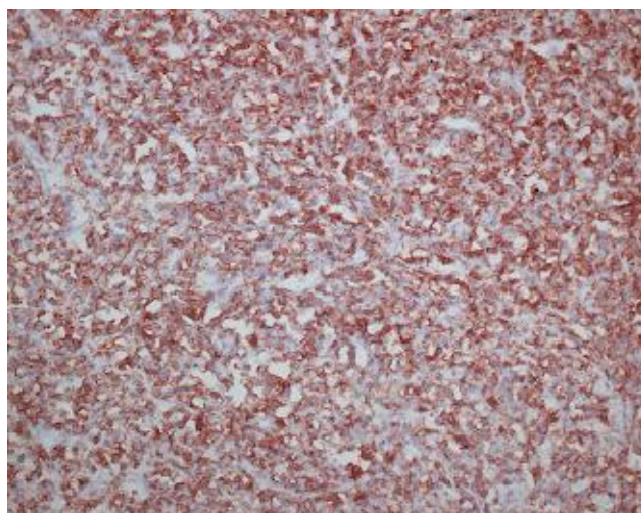


Figure 2. Malignant lymphoma of the ovary. Diffuse distributions of the tumor cells (H-E x200).

occipital mid-line (lymphoma?) that diffuses under the skin from the epidural distance after destructing the occipital bone that suppresses the transverse sinus (Figures 3 and 4). A biopsy was obtained from the occipital region. The report of the biopsy showed malign lymphoma infiltration (Figure 5). The thorax and abdominal computerized tomography of the patient displayed no abnormality related with bone marrow study. The patient was diagnosed with a diffused large cell extra-nodular non - Hodgkin's lymphoma after transferred to the Division of Internal Diseases, Department of Oncology and chemotherapy was planned. The patient received cyclophosphamide 750 mg/m^2 , adriablastine 50 mg/m^2 ,

vincristine 1.4 mg/m^2 and prednisolone 100 mg for 1 - 5 days, with 21 days interval and in 6 cures. A complete response was obtained at the end of the 3rd cure. At the end of the sixth cure the patient was referred for radiotherapy.

The patient was followed by control visits which were performed at every 3 months. However, the patient referred to our emergency clinic after 1 year due to instantaneous loss of consciousness. Neurological examination displayed a Glasgow Coma Scale at 3 and the contrasted brain tomography indicated a $5 \times 4 \text{ cm}$ mass at the left fronto - temporal region. The patient was hospitalized in our intensive care clinic and then she became

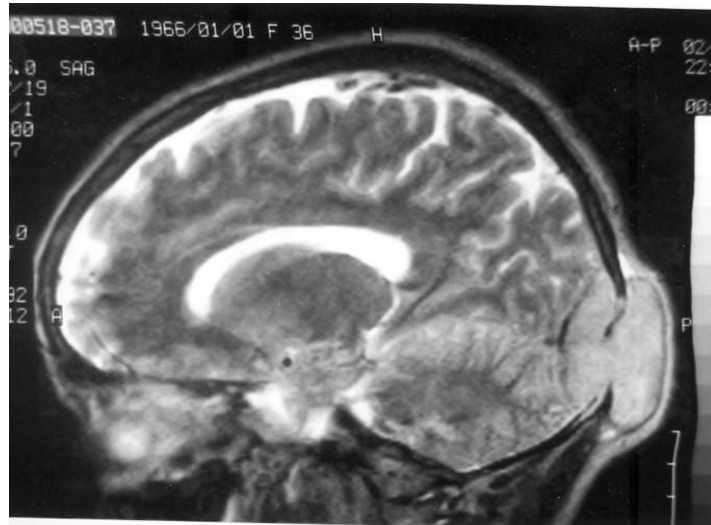


Figure 3. Sagittal T2-weighted MRI showing leptomeningeal methastasis of primary ovarian lymphoma located in the occipital area.

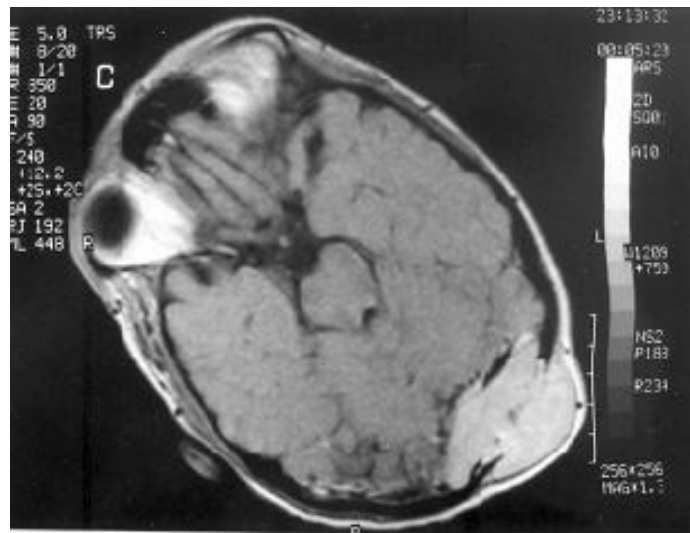


Figure 4. Contrast-enhanced axial T1-weighted MRI showing leptomeningeal methastasis of the primary ovarian lymphoma located in the occipital area.

exitus.

DISCUSSION

Systemic lymphoma is the 5th widespread cause of deaths due to cancer (Jellinger et al., 1976). Involvement of the central nerve system in lymphomas may be secondarily occurred as a systemic lymphoma or may be primarily occurred within the central nervous system.

Certain criteria's must be met in order to diagnose the patient with a primary lymphoma related to the genital system, both if the condition is a localized retention of an organ (such as ovarian lymphoma, cervix lymphoma) or if the condition is a diffused retention (such as corpus, cervix and vaginal retentions); 1) During diagnosis the lymphoma must be limited only within the mentioned region. A whole body scanning must be performed and no any other focuses must be determined; 2) No any atypical cell should be detected at the peripheral blood

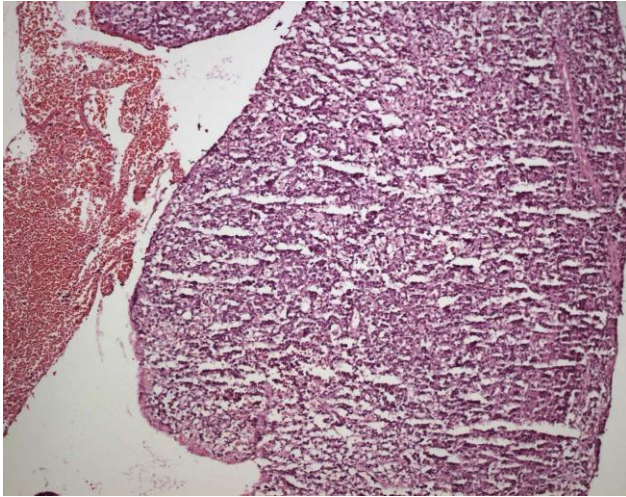


Figure 5. Malignant lymphoma metastasized to the leptomeningeal area (H-E x100).

system and bone marrow; 3) There must a minimum period of a few months between the occurrence of the lymphoma at the genital system and the secondary focuses (Fox et al., 1988; Maeda et al., 1988). No any other focuses were determined in the presented case based on physical examination and radiological imaging methods (MR and computed tomography) whereas no any atypical cells were seen in the bone marrow and; however, during the clinical follow-up of the patient, a cerebral mass was found by the means of physical examination and radiological imaging methods, nearly 1.5 months after an operation performed out due to an ovary tumor and the case was accepted as a cerebral metastasis sourcing from a primary ovarian lymphoma.

The metastatic diffusion of the cerebral lymphoma into the cerebral parenchyma commonly occurs at a late stage and shows a frequency ratio of 1 - 7% (Jellinger et al., 1976). Primary lymphomas may be deeply located multiple and bilateral tumors with a location site at the intra - parenchyma. In return, cerebral methastasis of lymphomas occurs as meningeal infiltration (Greenberg et al., 1995). The presented case is a patient with a lesion that demonstrates infiltration in the dura mater that causes bone destruction at the occipital region due to a cranial metastasis those sources from a primary ovarian tumor. Primarily the ratio of frequency of the lymphoma located at the ovary is less than 1% (Chorton et al.; Delgado et al., 1976; Monterroso et al., 1993). However,

cerebral metastasis of primary ovarian lymphomas is an extremely uncommon condition. In general, extra-nodular lymphomas have a better prognosis when compared to a nodular lymphoma at the same stage (Monterroso et al., 1993).

The five-year survival rate may differ according to the mentioned prognostic factors; however, variation can be 73% in patients with uterus located and 22% in patients with ovarian located lymphomas (Benjamin et al., 1995; Harris et al., 1984). The presented patient died 19 months after biopsy obtained after chemotherapy and radiotherapy were applied. The majority of female genital system lymphomas displays a histological structure of non-Hodgkin lymphomas (Monterroso et al., 1993).

The histological type in our case was a high - graded, diffused, large cell non - Hodgkin's lymphoma.

The presented case is an example of a cerebral leptomeningeal metastasis due to an uncommonly encountered primary ovarian lymphoma which was supported by physical examination, imaging and histopathological study and the site of location of the metastasis where symptoms are present pointing out the site of location as the ovaries.

REFERENCES

- Benjamin E, Isaacson PG (1995). Lymphoproliferative disease of the ovaries and female genital tract. In: Fox H (ed). Haines and Taylor's, New York, Churchill Livingstone: Obstet. Gynaecol. Pathol. 4(2): 1015-1041.
- Chorton I, Norris HJ, King FM (1974). Malignant reticuloendothelial disease involving the ovary as a primary manifestation. A series of 19 lymphomas and 1 granulocytic sarcoma. *Cancer* 34: 397-407.
- Delgado G, Smith JP, Declos G, Gallagher S (1976). Reticulum cell Sarcoma of the cervix. *Am. J. Obset. Gynecol.* 126: 691-694.
- Fox H, Langley FA, Govan ADT, Shirley HA, Bennet MH (1988). Malignant lymphoma presenting as an ovarian tumour a clinicopathological analysis of 34 cases. *Br. J. Obstet. Gynaecol.* 95:386-390.
- Greenberg JO (1995) Cerebral lymphoma. In: Adams RD. Neuroimaging: A Companion to A Victor's Principles of Neurology McGraw-Hill pp. 418-419.
- Harris NL, Scully RE (1984). Malignant lymphoma and granulocytic sarcoma of the uterus and vagina. A clinicopathologic analysis of 27 cases. *Cancer* 53: 2530-2545.
- Jellinger K, Radaszkiewicz T (1976). Involvement of the Central Nervous System in Malignant Lymphomas. *Virchows Arch (Pathol. Anat.)* 370: 345-62.
- Maeda T, Kamegai H, Mori H (1988). Malignant lymphoma presenting as initial symptom in the uterus. Case report. *Br. J. Ostet. Gynaecol.* 95: 1195-1197.
- Monterroso V, Jaffe ES, Merino MJ, Medeiros LJ (1993). Malignant lymphomas involving the ovary. A clinicopathologic analysis of 39 cases. *Am. J. Surg. Pathol.* 17: 154-170.