

Case Report

Primary Central Nervous System Non Hodgkin's Lymphoma: A Case Report

Mohit Gupta^{1*}, Hansa Choudhary^{2**}, Sunita Kulhari^{3**}, Neelu Gupta^{4**}

¹Assistant Professor, ²3rd Year PG Student, ³Senior Demonstrator, ⁴Head of Department,
*Dept of Pathology, Shri Guru Ram Rai Institute of Medical & Health Sciences, Dehradun, Uttarakhand, India.

**Department of Pathology, Sardar Patel Medical College, Bikaner, Rajasthan, India.

Corresponding Author: Hansa Choudhary

Received: 10/12/2015

Revised: 23/12/2015

Accepted: 28/12/2015

ABSTRACT

Primary CNS Lymphoma is a rare extranodal non-Hodgkin's lymphoma. It is mainly located supratentorially with predilection for the cerebral hemisphere having poor prognosis. We reported extranodal non-Hodgkin's lymphoma in 55 year old female patient. The patient's physical examination demonstrated paralysis on right side of the body. Magnetic resonance imaging (MRI) study revealed a mass lesion measuring 69×62×48mm involving both frontal lobes. The patient underwent a left frontal craniotomy and tumor was removed. Histopathological examination revealed multifocal and diffuse solid sheets of small round cell having scanty basophilic cytoplasm, round to irregular nuclei having small nucleoli and occasional mitotic figures with little intervening stroma. The patient received chemotherapy with cyclophosphamide, adriamycin, vincristine, and prednisolone (CHOP) every 3 weeks for six cycles. After passing 6 months of chemotherapy there was no evidence of tumor recurrence. The treatment should be with multidisciplinary approach and regular follow-up are recommended for every patient.

Keywords: Primary, CNS, Extranodal, Non Hodgkin's Lymphoma, PCNSL.

INTRODUCTION

Primary CNS Lymphoma (PCNSL) is a rare form of extranodal non-Hodgkin's lymphoma that arises within and remains restricted to the craniospinal axis, without evidence of systemic disease.

^[1] It is an interesting B cell lymphoma as very few B lymphocytes, if any, are found in the CNS under normal circumstances. ^[2]

In immunocompetent patients its incidence is 0.4% of primary brain tumours, whereas in immunodeficient it constitutes 5% of the same. PCNSL, classically a disease of 6th and 7th decade (4th decade in immune-compromised) shows a male preponderance. Majority, about 60% of cases show a supratentorial location with

predilection for the cerebral hemisphere, whereas 10-15% is infratentorial. ^[1] Although the prognosis remains poor for the majority of patients, approximately 20-30% of cases can be cured. ^[3]

MATERIALS AND METHODS

A 55 year old female presented with complaints of headache, nausea, vomiting and seizures since one month. There were behavioral and emotional changes since 15 days and paralysis on right side of the body since 2 days. There was no lymphadenopathy and organomegaly.

MRI of brain showed large ill defined intra axial mass lesion seen in the

bilateral frontal lobe, measuring about 62x69x48mm in dimensions. This showed heterogeneous signal on all the pulse sequences, predominantly hyperintense on T2 and FLAIR (fluid attenuated inversion recovery) and hypointense on T1W sequence without diffusion restriction. There was moderate to large perilesional edema. There was compression and effacement of the adjacent sulci gyri and bilateral frontal horn of lateral ventricle, involving anterior corpus callosum, body, genu, and rostrum. Left frontal horn lateral ventricle choroid plexuses and bilateral anterior basal ganglia involving caudate. On administration of contrast the lesion showed intense heterogeneous enhancement with necrotic areas.

Hemogram: Peripheral smear examination- Erythrocyte microcytic hypochromic. leucocytes: Count within normal range. Polymorph- 67%, lymphocytes- 28%, monocytes -2%, eosinophil-3%. Platelets:- adequate in number.

Blood sugar (random):108mg/dl (ref. range 70-110mg/dl), Blood urea 24mg/dl (ref. range 15-45mg/dl), Serum creatinine 0.8mg/dl (ref. range 0.6-1.5mg/dl). HIV status was negative.

Left frontal craniotomy was done and intraoperative finding revealed that the mass was grayish white, soft, vascular and at places firm.

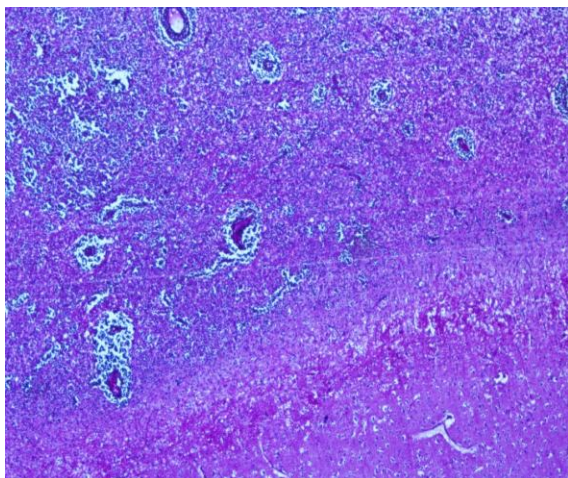


Figure 1: showing diffuse solid sheets of small round cell. Lower part showing brain tissue. (H&E Stain 4X)

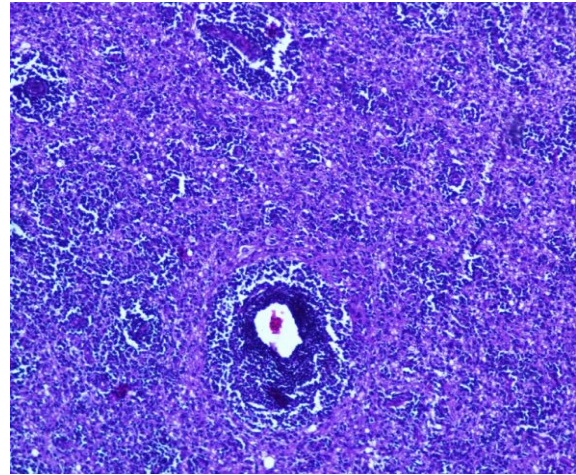


Figure 2: Showing diffuse solid sheets of small round cell little intervening stroma. These cells concentrated around blood vessels (perivascular cuffing) (H&E Stain 10X)

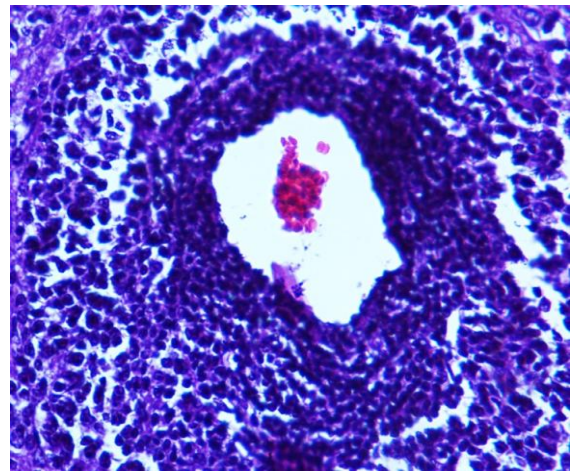


Figure 3: showing small round cell having scanty basophilic cytoplasm, round nuclei. These cells concentrated around blood vessels forming perivascular cuffing. (H&E Stain 40X)

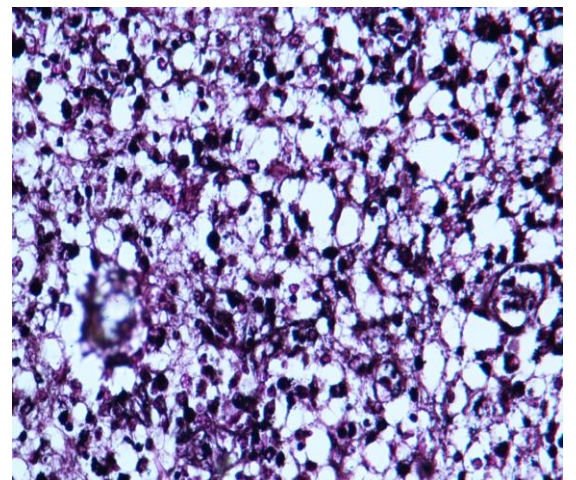


Figure 4: Showing concentric rings of reticulin fibres around the blood vessels with tumor cells entrapped within reticulin fibres. (H&E Stain 10X)

Pathological findings

Gross examination showed several greyish white and grey brown soft tissue

pieces of irregular size and shape collectively measuring 6×6 cm. then tissue was processed and stained with haematoxylin & eosin stain, Reticulin stain and Periodic acid Schiff (PAS) stain.

Microscopic examination showed multifocal and diffuse solid sheets of small round cell having scanty basophilic cytoplasm, round to irregular nuclei having small nucleoli and occasional mitotic figures with little intervening stroma (Figure 1 & 2). At many places these cells concentrated around blood vessels (perivascular cuffing) (Figure 3). These cells morphologically resemble lymphoid series cells. Reticulin staining showed concentric rings of reticulin fibres around the blood vessels with tumor cells entrapped within reticulin fibres (Figure 4). PAS staining showed negativity (Figure 5). Thus a diagnosis of primary CNS non Hodgkin's Lymphoma was made.

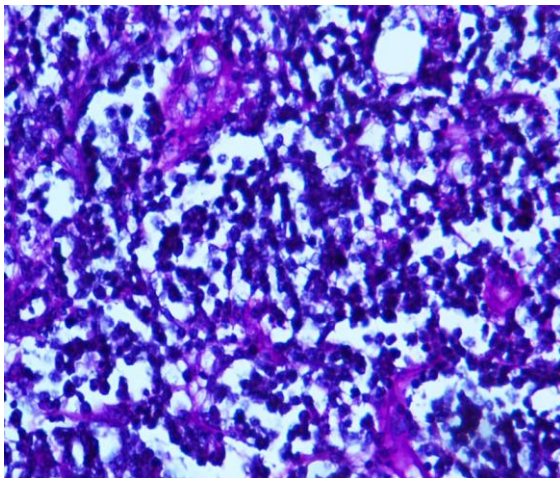


Figure 5: PAS staining showed negativity (H&E Stain 40X)

DISCUSSION AND RESULTS

This 55 years old female presented with symptoms of frontal mass lesion. There was no evidence for lymphoma elsewhere in the body. Hence it may be assumed to be a primary CNS lymphoma.

Although CNS lymphoma occurs more frequently in immunocompromised patients, incidence peaked in the mid-1990s and has now declined. This might be related to changes in HIV incidence and management. [3,4] In contrast, the incidence

remains high among older patients (>60 years) who are mostly immunocompetent. [5] Our patient was an immunocompetent patient presenting with frontal lesions.

PCNSL was first described by Bailey as 'Perithelial Sarcoma' in 1929. Subsequently it had varied historical pseudonyms like reticulum cell sarcoma (RCS), diffuse histiocytic lymphoma, RCS-microglioma and microglioma. [6] Indian scenario (study from northern India) revealed PCNSL cases constituted 1.2% of total intracranial tumors. [7] PCNSL, classically a disease of 6th and 7th decade (4th decade in immune-compromised) shows a male preponderance. Majority, about 60% of cases show a supratentorial location with predilection for the cerebral hemisphere. Whereas 10-15% are infratentorial. [1]

Our case showed a cerebral mass in bilateral frontal lobe location. In MRI it shows large ill defined intra axial mass lesion seen measuring about 62x69x48mm in dimensions. This showed heterogeneous signal on all the pulse sequences, predominantly hyperintense on T2 and FLAIR and hypointense on T1W sequence without diffusion restriction. There was moderate to large perilesional edema. There was compression and effacement of the adjacent sulci gyri and bilateral frontal horn of lateral ventricle, involving anterior corpus callosum, body, genu, and rostrum. Left frontal horn lateral ventricle choroid plexuses and bilateral anterior basal ganglia involving caudate. On administration of contrast the lesion showed intense heterogeneous enhancement with necrotic areas. In MRI, it was misdiagnosed as high grade glioma.

According to the National Cancer Institute of USA, non- Hodgkin's lymphoma is divided into three subtypes namely the small non-cleaved cell, lymphoblastic and large cell types. [8]

Histologically, In our case, multifocal and diffuse solid sheets of small round cell having scanty basophilic

cytoplasm, round to irregular nuclei having small nucleoli and occasional mitotic figures with little intervening stroma. At many places there was perivascular cuffing. Reticulin staining showed reticulin fibres around the blood vessels with tumor cells entrapped within reticulin fibres.

Although the prognosis remains poor for the majority of patients, approximately 20-30% of cases can be cured. [3] The best treatment strategy has yet to be defined. [9,10] However, biopsy followed by corticosteroids, high dose methotrexate (MTX) - based chemotherapy and/or radiotherapy are the current treatment options. PCNSL is a highly radiosensitive and chemosensitive tumor [3] which can result in a resolution of the tumors.

CONCLUSIONS

Primary CNS lymphoma is rare disease mainly located supratentorially. In MRI it may be misdiagnosed as glioma. Prognosis is generally poor for the majority of patients. The treatment should be with multidisciplinary approach and regular follow-up are recommended for every patient.

DECLARATIONS

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not Required

REFERENCES

1. Sarkar C, Deb P, Sharma MC. Primary central nervous system lymphoma: an update. Indian J Med Paediatr Oncol 2003; 24:17-24.
2. Anthony IC, Crawford DH, Bell JE. B lymphocytes in the normal brain:

- contrasts with HIV associated lymphoid infiltrates and lymphomas. Brain 2003; 126: 1058-1067
3. Sierra del Rio M, Rousseau A, Soussain C et al. Primary CNS lymphoma in immunocompetent patients. Oncologist. 2009 May; 14(5):526-539.
4. Hoffman S, Propp JM, McCarthy BJ. Temporal trends in incidence of primary brain tumors in the United States, 1985-1999. Neuro Oncol. 2006 Jan; 8(1):27-37.
5. Kadan-Lottick NS, Skluzacek MC, Gurney JG. Decreasing incidence rates of primary central nervous system lymphoma. Cancer. 2002 Jul 1; 95(1):193-202.
6. Schaumburg HH, Plank CR, Adams RD. The reticulum cell sarcomamicroglioma group of brain tumors. A consideration of their clinical features and therapy. Brain. 1972; 95: 199-212.
7. Powari M, Radotra B, Das A, Banerjee AK. A study of primary CNS lymphoma in Northern India. Surg Neurol 2002; 57: 113-116.
8. Babu Rafeeq, Narayanadas K, Aboobacker CM. Non Hodgkin's Lymphoma presenting as cardiac tumours. Indian Paediatrics.2001; 38:543-5.
9. O'Neill BP, O'Fallon JR, Earle JD et al. Primary central nervous system non-Hodgkin's lymphoma: survival advantages with combined initial therapy? Int J Radiat Oncol Biol Phys. 1995 Oct 15; 33(3):663-73.
10. Giustina A, Gola M, Doga M et al. Clinical review 136: Primary lymphoma of the pituitary: an emerging clinical entity. J Clin Endocrinol Metab. 2001 Oct; 86(10):4567-75.

How to cite this article: Gupta M, Choudhary H, Kulhari S et al. Primary central nervous system non Hodgkin's lymphoma: a case report. Int J Health Sci Res. 2016; 6(1):585-588.
