Journal of Cytology & Histology
ISSN: 2157-7099

Bhuyan et al., J Cytol Histol 2011, 2:6
DOI: 10.4172/2157-7099.1000127

Case Report Open Access

# Primary CNS Lymphoma- a Squash Diagnosis

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## Abstract

Primary CNS Lymphoma (PCNSL) is a rare form of extranodal non-Hodgkin's lymphoma. It has a varied presentation and cytomorphology. We diagnosed a case of PCNSL in a 52-year old male in intra-operative squash cytosmear differentiating from other lesions. In cytosmear even gradient of cellular density, monomorphic tumour cells with lack of cellular cohesion on a background showing good no. of tingible body macrophages and lymphoglandular bodies were the characteristic features. Histomorphology showed the features of diffuse large cell lymphoma and was immunohistochemically confirmed as B-cell type (DLBCL).

**Keywords**: Primary lymphoma; CNS

## 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]. In immunocompetent patients its incidence is 0.4% of primary brain tumours, whereas in immunodeficient it constitutes 5% of the same [1].

Morphologically, primary CNS lymphomas resemble systemic lymphomas and are generally large B cell type or immunoblastic type [2]. The cytosmear shows a diffuse monomorphic population of lymphoid cells with high N:C ratio, prominent nucleoli and often irregularities in nuclear membrane out line [2]. The lack of cellular cohesion or cytoplasmic processes are useful to distinguish the tumor from metastatic carcinoma and glioma [2].

We present one such case diagnosed in intraoperative squash smear and a brief discussion on differentiating PCNSL from other entities in cytology.

## Case Report

A 52-year-old male presented with complaints of headache, vomiting, unstable gait falling to one side for a duration of 6 months. CT scan of brain revealed a single, deep seated hyperdense, intraparenchymal lesion in the left cerebellar hemisphere. The mass was of size (3x2) cm with well defined border showing homogeneously contrast enhancement and exhibiting peripheral edema along with features of obstructive hydrocephalus (Figure 1).

Other investigations revealed no abnormality in hemogram and peripheral smear examination. HIV status was negative. The patient had no palpable enlarged peripheral lymph node. No deep seated lymph node was detected even after thorough searching by Ultrasonography & CT Scan. Preoperative finding revealed that the mass was unencapsulated, grayish, soft and was richly vascular.

Squash cytosmears showed easy spreading of tissue. On lower magnification smears were cellular showing even gradient of cellular density with clustering at places. Intermediate magnification revealed monomorphic population of cells with lack of cellular cohesion admixed with good number of tingible body macrophages (Figure 2). Background showed large number of degenerated nuclei, apoptotic bodies along with numerous lymphoglandular bodies. Higher magnification revealed large round cells with thin rim of basophilic cytoplasm, coarse granular chromatin, nuclear convolution and prominent nucleoli in many cells (Figure 2). Few mature looking lymphocytes were also noted. With this cytomorphology a diagnosis of large cell lymphoma was given.

In tissue section solid sheets of large round cells with little intervening stroma clustering around the blood vessels was the picture (Figure 3). Higher magnification revealed nuclear membrane convolution, prominent nucleoli and mitotic figures along with angioinvasion by the tumor cells. Reticulin staining showed concentric rings of retirulin fibres around the blood vessels with tumor cells locked within reticulin fibres (Figure 3). Thus a diagnosis of primary CNS lymphoma diffuse large cell type was made. It was later confirmed by immunohistochemical markers CD20 and MUM1 positivity.

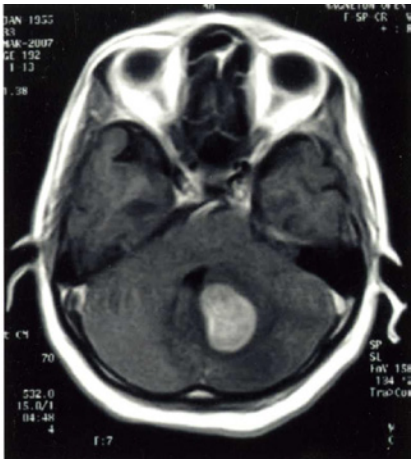

**Figure 1:** CT Scan of brain showing Lt. Cerebellar space occupying lesion.

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**Received** August 20, 2011; **Accepted** September 15, 2011; **Published** September 20, 2011

**Citation:** Bhuyan P, Mohanty P, Pattnaik K, Mishra A, Biswal P, et al. (2011) Primary CNS Lymphoma- a Squash Diagnosis. J Cytol Histol 2:127. doi:10.4172/2157-7099.1000127

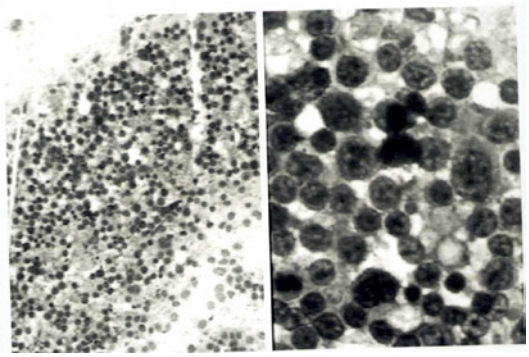

**Figure 2:** Cystosmears showing (a) monomorphic, discohesive cells with tingible body macrophages (H & E stain, 100x) (b) large round cells with prominent nucleoli, thin rim of cytoplasm and lymphoglandular bodies (H & E stain, 400x).

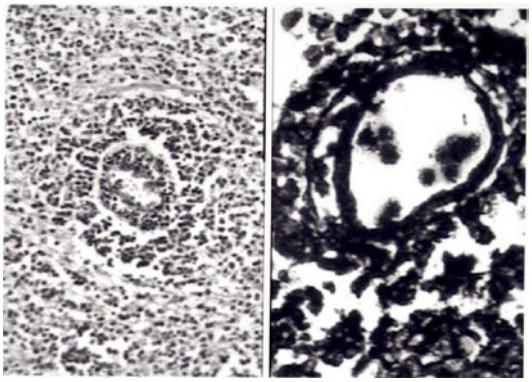

**Figure 3:** Tissue sections showing (a) Angiocentric distribution of Lymphoma cells (H & E stain 100x), (b) Angionvasive lymphoma cells, locked within concentric rings of reticulin fiberes (Reticulin stain, 400x).

## Discussion

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[3,4]. Indian scenario (study from northern India) revealed PCNSL cases constituted 1.2% of total intracranial tumors [5]. A similar study undertaken by All India Institute of Medical Science (AIIMS) and National Institute of Mental Health and Science (NIMHANS) found PCNSL cases to be constituting 0.95% and 0.92% of the total intracranial tumors at the two institutions respectively [6,7].

PCNSL, classically a disease of 6$^{th}$ and 7$^{th}$ decade (4$^{th}$ 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 an intracerebellar mass periventricular in location – a site for primary brain lymphoma. Secondly lymphomas are extrapial, limited to the brain exterior [8]. In CT scan it shows a hyperdense mass with homogeneously contrast enhancement. Multifocal lesions, often centrally necrotic appearing as ring enhancing lesion resembling glioblastoma, are characteristic features in immunocompromised patients. Surprisingly corticosteroid therapy leads to complete disappearance of the tumor in CT scan (ghost tumor) as well as in cytosmears / biopsy [8].

In squash cytosmears the characteristic features are easy spreading (liquid tumor), even gradient of cellular density, monomorphic population of discohesive cells, thin rim of basophilic cytoplasm, coarse granular chromatin, prominent nucleoli along with good number of lymphoglandular bodies and tingible body macrophages in the background [8,9]. All these features were present in this case. Further more, it was differentiated from metastatic small cell carcinoma and small cell glioblastoma. Small cell carcinoma classically presents as small or medium sized cells with little or no cytoplasm dispersed in clusters with salt and pepper chromatin and nuclear moulding. Single file (piles or pennies) or money rolls (onion skinning) arrangement, tear-drop cells, engulfment of apoptotic bodies along with numerous mitotic figures are additional findings [5,7]. At times metastatic melanoma also mimics lymphomas. However, eccentrically placed large nucleus, prominent macro nucleolus and absence of lymphoid globules are the differentiating features [9]. Small cell glioblastoma was ruled out by absence of gliofibrillary background, microvascular proliferation, greater degree of nuclear pleomorphism and necrosis [8]. Besides this though affinity for the blood vessels is a feature of both glioblastoma and lymphoma, cells in glioblastoma don't permeate the vascular wall.

At times lymphoma is misdiagnosed as glioma when biopsy is taken from the periphery or infiltrating margin of the tumor where marked gliosis and lymphocytic infiltration is a prominent picture. In this situation lymphoma cells hiding on a glial meshwork and entangled around the blood vessels gives a clue to the diagnosis of lymphoma [8].

Histologically diffuse architecture, large lymphoid cells with nuclear membrane convolution, prominent nucleoli and angiocentric distribution were the identifying features. Most characterstic features were angioinvasion by lymphoma cells and concentric rings of reticulin fibres within and around the blood vessels locking the lymphoma cells within it simulating the annual growth rings of a tree [10].

The affinity of tumor cells for blood vessels is the most distinctive architectural features of PCNSL especially DLBCL type i.e. composed of immunoblasts or centroblasts [4,10]. The large majority (98%) of PCNSL are diffuse large B-cell lymphomas (DLBCL) showing positive reaction to CD 20 and are of high grade type. The rest are poorly characterized low grade lymphomas, Burkitt lymphomas and T-Cell lymphomas. Our case was confirmed as DLBCL by CD20 and MUM1 in tissue section.

This case is reported here because of its rare incidence and varied cytomorphology in squash cytosmears mimicking other entities.

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