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 craniocerebral 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 outline [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 reticulin 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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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 revealed PCNSL cases constituted 1.2% of total intracranial tumors and microglioma[3,4]. Indian scenario (study from northern India)


