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Case Report

PRIMARY CENTRAL NERVOUS SYSTEM EFFUSION PLASMABLASTIC LYMPHOMA IN IMMUNOCOMPROMISED PATIENT: A RARE PHENOMENON

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ABSTRACT

Primary effusion lymphoma (PEL) is an aggressive neoplasm with a high rate of fatality. PEL cells are known to have morphological diversities, which range from immunoblastic or plasmablastic to anaplastic. Most of these cases are described in immunocompromised as well as immunocompetent patients. Plasmablastic lymphoma remains a diagnostic challenge, especially when encountered with the presentation as PEL. In spite of therapeutic advances, PEL remains an aggressive disease with a high rate of fatality. We describe one case of this extremely rare neoplasm in an immunocompromised patient presenting in the form of primary central nervous system effusion plasmablastic lymphoma. To the best of our knowledge, this is the first case ever been reported in the literature.

Keywords: Primary effusion lymphoma, Plasmablastic lymphoma, Papanicolaou smear, Flow cytometry.

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INTRODUCTION

Cancer is the most dreaded disease of the 21st century, and the frequency of its occurrence in adults is 2.7 times more than in children and lymphomas contribute to almost 18% [1]. Infection with human immunodeficiency virus (HIV) predisposes to the development of neoplasm including lymphoma [2,3]. Primary effusion lymphoma is one of the least common of the AIDS-related lymphomas, accounting for 1–4% of all cases [4,5]. Plasmablastic lymphoma (PBL) is a rare subtype of diffuse large B-cell lymphoma associated with HIV patients. Its presentation as primary central nervous system (CNS) effusion lymphomas is even rarer, in which there is no detectable tumor in the individual.

CASE REPORT

A 45-year immunocompromised male, presented to the neurosurgery department of Kasturba Hospital with complaint of headache, and altered sensorium for 15 days with no fever, neck rigidity, weight loss, organomegaly, or lymphadenopathy. Both plain computed tomography and magnetic resonance imaging of the brain yielded essentially normal findings. Papanicolaou-stained smears of cerebrospinal fluid (CSF) from the patients were examined. The smears showed increased cellularity with flourishing abnormal plasmacytoid cells (Fig. 1). Immunohistochemistry was attempted on cell block but was not processed due to scanty cell yield. Flow cytometry on fresh CSF sample was performed. The neoplastic cells showed plasmacytic differentiation with expression of bright CD38 and CD56 in approximately 67–70% of the total cells. These cells were also negative for CD79a and CD19. Interestingly, the neoplastic cells were not restricted for both kappa and lambda light chain, signifying non-secretory phenotype. Protein electrophoresis showed a normal pattern.

DISCUSSION

PBL is a rare phenotype presenting as primary CNS effusion lymphoma. To the best of our knowledge, till date, there is no reference in the literature of this type of presentation of PBL. PBL is known as one of the most aggressive forms of lymphoma and now has been accepted by the WHO as one of the subtypes of diffuse large B-cell lymphoma [6]. Since the first description of PBL, in 1997, by Delecluse *et al.* [7], the spectrum of PBL involvement of various organs has widened. Various sites from where it has been reported include skin [8], breast [9], gastrointestinal [10], central nervous system [11], male genitourinary system, and bone [12]. It is rarely found in children [13]. PBL is characterized by B-immunoblast-like cells with the expression of immunophenotype as explained in mature plasma cells [14]. Morphologically, PBL cells have two subtypes described in literature [15], first, which is most common is characterized by monomorphic neoplastic cells with minimal or no plasmacytic differentiation. The second type includes PBL with plasmacytic differentiation which shows tumor cells with differentiation to mature plasma cells, i.e., round-to-oval cells with an eccentric nucleus and some with prominent nucleolus. Sometimes, a very conspicuous perinuclear hof in the cytoplasm may be seen [16]. Epstein–Barr Virus, a human gamma herpesvirus, infects almost 90% of population worldwide and is associated with the development of B-cell lymphomas [17]. It is also found in most of the plasmablastic cases, whereas human herpesvirus 8 (HHV8) is absent [18]. We were unable to go further with immunology for Epstein-Barr virus or HHV8 as the patient was lost to follow-up.

CONCLUSION

Primary CNS effusion lymphoma of non-secretory plasmablastic type is the rarest phenomenon to be reported in the literature. When there is an established hematological or solid organ malignancy, the abnormal CSF cells can be easily subcategorized, but for a primary CNS effusion lymphoma, concurrent flow cytometry, and cytomorphological evaluation are the most appropriate to come to an acceptable conclusion.

AUTHOR'S CONTRIBUTION

Dr. Brij Mohan: Interpretation of cytomorphology and case report write up and has reviewed the article. Dr. Sushma: Interpretation of flow cytometry. Dr. Arijit: Extracting the case file with other case details. Dr. Tanvi: Write up and also reviewed the article. Dr. Pavithra: Reviewing literature for the occurrence of the case.

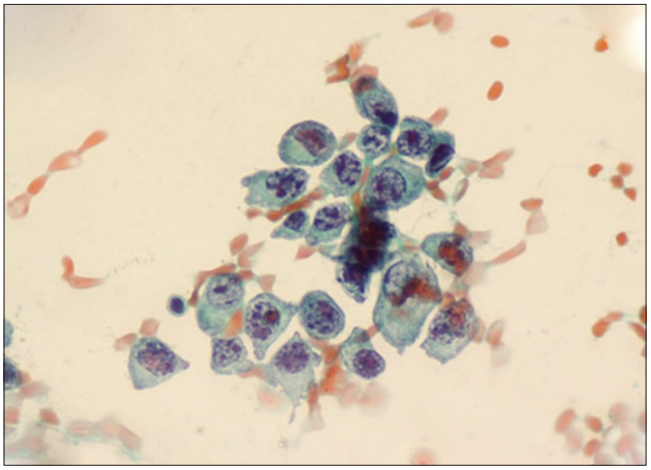

Fig. 1: Papanicolaou stain (×400) cerebrospinal fluid smear showing bizarre plasmacytoid cells

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

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