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Plasmablastic Lymphoma: Retrospective Study of Cases from The Hematology Sector of The Santa Casa of São Paulo

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Introduction

Plasmablastic Lymphoma was first described in 1997 by Delecluse and collaborators. In this description there were 16 patients, of whom 15 were HIV positive. The most common presentation was extranodal and in the oral cavity [1,11]. Even today it is extremely related to HIV infection, with this virus being present in 72.2% of Plasmablastic Lymphoma cases [6], and this lymphoma accounting for about 2% of HIV-related lymphomas [10]. However, nowadays, there are numerous cases in HIV-negative patients, especially when there is some degree of immunosuppression (28.1%), such as malignancy or prior transplant [2].

Plasmablastic Lymphoma affects more men, around 75% [10]. The average age changes according to HIV serology. In seropositive patients the average age at diagnosis is around 38 years [1] and in those with HIV negative around 58 years [2].

In addition to the close relationship with HIV infection, Plasmablastic Lymphoma also exhibits a high association with EBV infection (about 60% of cases) [1,7,10]. It is believed that infection by such a viral agent, which presents antiapoptotic properties [10] associated with the presence of Myc gene rearrangements, which occur in about 40-50% of cases [11], are essential participants for the occurrence of Plasmablastic Lymphoma [10].

The most common clinical presentation remains extranodal, maintaining a predilection for the oral cavity [5]. Other frequent involvements are the gastrointestinal tract and skin [10]. Bone Marrow involvement occurs in about 30-40% of cases [10,11].

Pathologically it has monomorphic proliferation of round or oval cells with abundant cytoplasm, eccentric nucleus and evident nucleolus with a background of lymphocytes, cells apoptotic, mitotic and macrophages [11].

The immunophenotyping is similar to that of plasma

cells: positive for CD79a, MUM1, CD38, CD138 and BLIMP1 and negative for PAX5, CD19, CD20 and CD45. Ki67 is usually close to 100% [10]. About 39% of patients may express CD30 [11], a fact that has been taken into account in more recent studies to evaluate the efficacy of using Brentuximab for treatment of Plasmablastic Lymphoma [10].

Due to its rarity and the peculiar characteristics that merge Multiple Myeloma and Lymphoma, the diagnosis of Plasmablastic Lymphoma is usually difficult [3]. Important differential diagnoses must be ruled out such as:
Poorly differentiated carcinoma, Multiple Plasmablastic Myeloma or Anaplastic, Primary Effusion Lymphoma, ALK-positive Anaplastic Lymphoma, Plasmablastic Microlymphoma/ Castleman Disease [1,10].

The treatment of Plasmablastic Lymphoma, both for HIV positive and for HIV negative remains a challenge, especially due to the scarcity of cases and the negativity of CD20. CHOP has shown to be ineffective and there is no standard treatment [7]. The NCCN recommends the use of more aggressive chemotherapies such as EPOCH, CODOX-M-VAC, HYPERCVAD [8]. Case reports Havw shown that Bortezomib in combination with EPOCH, both in HIV positive and negative, can generate longer remissions (up to 24 months) [7].

The results for cases of relapsed and refractory Plasmablastic Lymphoma are disastrous. In HIV positive, relapse can generate an overall survival of 3.5 months [11]. Studies show that in cases of refractory lymphomas, 60 % will progress before undergoing auto-HSCT [11]. In this scenario, auto-HSCT can be viable and have benefit. The suggestion is that high-risk patients (IPI> 2, HIV negative, presence of MYC rearrangement, TP53 deletion and any response inferior to CR with chemotherapy) should undergo auto-HSCT in first remission. Refractory patients should undergo transplantation in second remission [11].

The data regarding survival are divergent, but in general show an average of 8 months of overall survival [10]. There is also divergence regarding factors that can alter these results. The use of ART seems to improve disease-free survival, but the reasons for this remain uncertain, it is believed to be related to an increase in CD4 [10]. The IPI seems to relate well to the risk of these patients, especially according to LDH level, disease stage, age and performance status [10]. Both in HIV positive and negative, EBV negativity is related to worse outcomes [1,2].

This study seeks to perform a review of the characteristics of presentation and treatment of Plasmablastic Lymphoma cases from the Hematology and Hemotherapy service of Santa Casa de São Paulo

Objectives

To evaluate clinical, epidemiological, histological and therapeutic response characteristics of patients diagnosed with Plasmablastic Lymphoma at the Hematology and Hemotherapy service of Santa Casa de São Paulo.

Methods

Study Design

Retrospective Longitudinal Study

Study Population

Patients diagnosed with Plasmablastic Lymphoma at Santa Casa de São Paulo

Sample

07 cases diagnosed with Plasmablastic Lymphoma at Santa Casa de São Paulo

Inclusion Criteria

All cases with biopsy and immunohistochemistry compatible with Plasmablastic Lymphoma

Exclusion Criteria

Cases with inconclusive biopsy, with medical records unavailable.

Materials

Retrieval of patient medical records under analysis.

Ethical Issues

The researchers involved commit to maintaining confidentiality regarding the data obtained. The project will be submitted to the Ethics Committee in Research.

Project Budget

This project does not have sponsorship funds, with expenses being the responsibility of the researchers. An estimated cost of around R$ 100.00 for stationery and printing materials.

Schedule

Year	**2017**												**2018**		
Months	J	F	M	A	M	J	J	A	S	O	N	D	J	F	M
Bibliographic research	x	x	x	x											
Data					x	x	x	x							
Database construction and Statistical analysis							x	x	x						
Discussion of data										x	x				
Writing the scientific article												x	x		
Presentation														x	

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