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Review Article

Epstein Barr Virus and its Causality in Hodgkin Lymphoma

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Abstract

Hodgkin lymphoma (HL) is a malignant disease that usually presents in young adults, with an indolent and variable course. Its presentation usually occurs with adenopathy and B symptoms; several risk factors have been pointed out as important precursors of HL. Regarding Epstein Barr virus (EBV), literature points to its presence in 20-50% of classical HL, mostly the scleronodular type.

With this case, we show causality between EBV infection and Hodgkin lymphoma development, something rare but well documented in the literature. By showing a typical presentation of a hematologic malignant process, with adenopathies and B symptoms that are very unspecific. With all the exams that were performed, we diagnosed a classical HL, and within the prolonged symptoms, we can assume that the EBV subacute infection was the most likely predisposal factor to this malignant process. The final diagnosis was made promptly, and early referral permitted the best outcome possible, with sustained remission.

Keywords: epstein barr virus; hodgkin lymphoma; adenopathy; immune system; b symptoms

Learning points:

1. Instructive errors in the rare presentation of a common condition, given the fact the viral etiology of Hodgkin lymphoma is usually forgotten.
2. Failure to detect serious illness because of clinical judgment and or experience, given the fact it is a young and healthy patient with no risk factors.

Introduction:

Hodgkin lymphoma (HL) is a malignant disease that usually presents in young adults, with an indolent and variable course [1]. Its presentation usually occurs with adenopathy (over 60%) mainly within the cervical area, and B symptoms (~40%) are more common with advanced stages [1-2]. Several risk factors have been pointed out as important precursors of HL, the most common Epstein-Barr virus (EBV), obesity, tabagism, and immunosuppression [3-4]. Regarding EBV literature points to its presence in 20-50% of classical HL (mostly the scleronodular type), and even more common if immunosuppressive conditions are present (HIV) [4]. The treatment of HL is under large investigation, and today the cure is achieved in approximately 75% of patients [5].

With this case, we pretend to bring more knowledge about this virus and to show a case where the causality of EBV and HL is suggestive. And also want to show that these cases' outcomes depend mostly on prompt diagnosis and treatment.

Casedescription:

A 27-year-old woman, with no relevant previous clinical history, was admitted to the emergency department with inguinal and cervical adenopathies, anorexia, asthenia, and night fever (maximum 38,1ºC), being the first symptoms reported within the previous 4 months.

The analytic study showed normocytic and normochromic anemia (hemoglobin 10.8g/dL), a thrombocytosis (410000/uL), and an augmented sedimentation velocity (57mm). She performed a total body computed tomography that described hepatosplenomegaly (spleen with 15cm) and unspecific supra and infra diaphragmatic

adenopathies (cervical, mediastinal, periaortic, and inguinal), as shown in Figure 1.

To complement the study a positron emission tomography was made, showing an abnormal glycolytic hypermetabolism in several supra and infra diaphragmatic adenopathies, suggesting a malignant lymphoproliferative process in course. A narrower analytic study was performed showing positivity to IgM and IgG of EBV on the serologies, all the other study was negative.

Given the EBV subacute infection, it was made a course of antiviral therapy with acyclovir.

The microscopic analysis of one cervical ganglia excised showed the presence of a classical HL, with a scleronodular subtype, the presence of EBV on the ganglia wasn't detected.

The referral of the patient to our reference haemato-oncologic center was immediately made and she started quimiotherapic therapy. The complete remission was achieved after the 6th cycle, and she maintains surveillance until the present time, after one and a half years.

Discussion:

This case shows a typical presentation of a hematologic malignant process, with adenopathies and B symptoms being the most noted signs, that go according to what is referred to as the most common features at presentation [1-2]. However, those are very unspecific, and with the suspicion of malignant neoplasm, the final diagnosis must be made promptly, to start treatment and achieve a cure in approximately 75% of patients [5].

With all the exams that were performed, we diagnosed a classical HL, and within the prolonged symptoms, we can assume the causality with EBV subacute infection, being the most likely predisposal factor to this malignant process. The causality between EBV and HL is well described in the literature, pointing to about 20-50% of the cases, and the scleronodular type is the most commonly associated [4], as we show in our case.

Besides predisposing factor, EBV has also been studied for its prognostic significance in HL survival [6-7]. When is present usually determines poor overall survival and disease- specific survival, therefore is being appointed as a valuable prognostic marker [7]. Regarding our case, other factors may be involved in the good outcome, respecting the young age and fast diagnosis of it.

Conclusion:

EBV has been extensively studied and is associated with several types of lymphomas, including HL. The relationship between both is complex and not fully understood. However, it is known that EBV plays a role in the development of a specific subtype of HL, triggering an immune response and leading to the characteristic presence of inflammatory cells in the affected lymph nodes.

With this case, we showed an example where this causality may play a big role in HL development, but the exact causal relationship between the virus and the development of the disease is still being investigated. Further research is needed to fully understand the mechanisms by which EBV contributes to developing HL and identify potential therapeutic targets.

But with a fast diagnosis and treatment, the cure is possible, as we have shown in our patients with sustained remission after one and a half years.

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