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Case Report

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Rare Evolution of an Epstein-Barr-virus-Positive Atypical B-Lymphoproliferative disorder Into Classic Hodgkin Lymphoma

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Abstract

A 78 years-old woman exhibited a group of axillary lymph nodes, the largest reaching 2 cm in diameter. The histopathology was consistent, a priori with advanced-age related, Epstein-Barr virus-positive, atypical B-cell lymphoproliferative disorder. A revision with additional immunostaining seems to favor a progression into classic Hodgkin lymphoma. A proposed conversion may account for such clinical features as the deterioration in the patient's condition. It might indicate, moreover, the mitigation for an early misdiagnosis.

Keywords: Advanced-Age-Related Ebv-Positive B-Cell Lymphoproliferative Disorder, Atypical Hyperplasia, Revision, Evolution, Classic Hodgkin Lymphoma

Introduction

Classic Hodgkin lymphomas (cHL) in the Western World affects mainly young adults. In flagrant contrast, elderlies are the most frequently spared by the malignancy [1,2]. The nodular sclerosis (NScHL) type of cHL is the most frequent, occurring between 15-35 of age, while older people represent a minority thereof. It is of note that the Hodgkin-Reed-Sternberg (HRS) tumor cells in NScHL are infrequently infected by the Epstein-Barr virus (EBV), as documented by *in situ* hybridization, and compared with other subtypes [3].

The report of an older female patient, presenting with axillary lymphadenopathy, but without symptoms at first, is exhibited hereby to display a possible diagnosis. A rare incidence, that of cHL of the elders, is noted for mimicking a reactive condition, that is extranodal in 70% of the cases, and which affects the skin, tonsil, lungs, and stomach. The reactive entity is composed of large, atypical B-RS-like cells in an inflammatory background with frequent necrosis. The large, atypical cells express CD20+, CD30+, LPM1+, EBER+, CD15- and CD3- [4]. This condition, designated age-related EBV-associated B-cell lymphoproliferative disorder, is non-immune-deficiency-related, but, since it occurs well beyond 50 years of age, it might be linked to immune senescence [5]. The age ranges most often from 70 to 79 years. The female-to-male ratio is 3.3:1.4 [1].

Case Description

A 78-old-woman, member of a kibbutz, exhibited an axillary lymphadenopathy, but no symptoms. The morphologic picture raised at first, the suspicion of angioimmunoblastic T-cell lymphoma. This diagnosis was not retained, however, in the absence of pan-T-cell restriction, and in the presence of a negative TCR-γ. Follicle-formation and a lack of mantle zones, together with intrafollicular evidence of HRS-like cells, positive for EBER expression, underlined by the patient's age, and evoked by our consultant, as a probable advanced-age related Epstein-Barr virus (EBV)-associated atypical B-LPD [1].

The histopathology was revised at this stage. Additional immunostainings were performed notably releasing CD30 ++; CD15 + and MUM1++. Together with the revision, they pointed out at a probable progression to cHL [3].

While the proposed condition of advanced-age related, EBV-positive, atypical B-cell LPD, may persist as such, it might also transform rarely into a lymphoma. An attempt at classifying this patient's malady as a lymphoma was pursued [1].

Several features of the present biopsy led us favor an unusual conversion of the suspected condition, into a rare cHL transformation, so far described in two cases only [1].

Discussion

Lymph nodes enlargement with no associated symptoms, is a priori in favor of a benign illness. Large B cells, most of which are positive for EBER, distributed in a partially preserved lymph node, caught our attention. When underlined by the older patient's age it might raise the possible diagnosis of advanced age of related

EBV-associated atypical B-LPDV, as mentioned previously [1]. A majority of the infrequent aEBVLPD reports originate from the Far East, but a significant description of aEBVLPD cases, highlighted the distribution of the disease entity in Western countries [6].

Although, aEBVLPD was favored at first, a diagnosis of cHL in the elderly was finally established, substantiated by the details presented in Table 1.

Despite its rarity in the elderly, a diagnosis of cHL has been both suggested and confirmed by repeated immunohistochemistry. This patient would represent a third case of the transformation of an EBV-positive atypical B-LPD to cHL of the elderly.

Conclusion

Classic Hodgkin lymphoma occurs rarely in the elderly. Wherever this diagnosis is evoked, the 5th WHO Classification of Hematolymphoid Tumours (2022) [7], highlights a benign condition that should be chosen instead: the advanced-age related, EBV-positive, atypical B-cell LPD [7].

Nevertheless, one should recall, that this condition is prone, though very rarely to convert into a lymphoma. The diagnosis of cHL of the elderly is favored, based on the features displayed in Table 1.

1. Female patient.
2. Immune senescence is not excluded.
3. Large atypical HRS-like cells are dispersed.
4. Large cell markers: CD20 -/+, CD30+, CD15+, MUMl+, EBER+.
5. No extensive necrosis.

Table 1: Features in favor of classic Hodgkin lymphoma in the elderly (the patient reported).

Conflicts Of Interest

The authors declare 'No conflicts of interest exist'.

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