EBV-Associated Cytotoxic Peripheral T-cell Primary Pulmonary Lymphoma

Linfoma Primário Pulmonar de Células Citotóxicas Periféricas T Associado ao EBV

Keywords: Epstein-Barr Virus Infections/complications; Lung Neoplasms; Lymphoma, T-Cell; T-Lymphocytes, Cytotoxic
Palavras-chave: Infecções pelo Vírus Epstein-Barr/complicações; Linfoma de Células T; Linfócitos T Citotóxicos; Neoplasias dos Pulmões

Dear Editor,

Primary pulmonary lymphomas (PPL) are extremely rare, comprising less than 1% of all lymphomas and less than 0.5% of all primary lung cancers.[1] When they occur, they are usually B-cell lymphomas.[1,2] Primary T-cell lymphomas of the lung are exceedingly rare, with only a few case reports described in the literature. We present a case of a primary pulmonary T-cell lymphoma to highlight its clinical and radiological characteristics and the challenges in its diagnosis and treatment.

A 43-year-old white woman, smoker and with a history of mild asthma and Behçet's disease under azathioprine, presented with left pleuritic chest pain. Initial chest radiography was unremarkable. Two weeks later, she developed a fever, productive cough and exertional dyspnea. Reevaluation with chest CT showed multiple bilateral consolidations and ground-glass opacities (Fig. 1A). Lab results revealed elevation of C-reactive protein and lactate dehydrogenase. Despite empirical antibiotics and systemic corticosteroids, there was no improvement. Bronchoscopy and bronchoalveolar lavage were unremarkable. A CT-guided transthoracic biopsy showed small lymphoid cells with scant cytoplasm and slightly irregular basophilic nuclei, and necrosis (Fig. 1B), which, combined with the immunohistochemical staining results, allowed for the diagnosis of an Epstein-Barr Virus (EBV)-associated peripheral cytotoxic T-cell lymphoma by a pathologist specialized in lymphomas. The patient's condition deteriorated rapidly, and she passed away shortly after being transferred to the hematology department.

Primary pulmonary lymphoma is defined as a lymphoma confined to the lung, with or without lymphatic hilar involvement at the time of the diagnosis or up to three months after.[1,2] This type of lymphoma, particularly of T-cell origin, poses a diagnostic challenge due to its rarity (either as PPL or through secondary involvement of the lung) and overlapping clinical presenting features with more common respiratory infections.[1] Clinically, men seem to be slightly more affected than women[1,2] and the mean age at diagnosis is around 60 years.[1] Radiologically, it may present as a solitary or as multiple nodules, ground-glass opacities, or consolidation with air bronchogram.[2] Unilateral involvement is more common, although bilateral cases can occur in about 10% of cases.[1] The prognosis is generally poor, with a rapid clinical deterioration.[3] Immunosuppression and EBV infection may play a role in its pathogenesis.[4] There is no established treatment protocol, although cyclophosphamide, doxorubicin, vincristine, and prednisolone (CHOP)-based chemotherapy is generally used.[4] Early diagnosis and treatment initiation are crucial.

The aim of this case is to raise awareness of this entity which requires high clinical suspicion, emphasizing the importance of a timely and accurate diagnosis to increase the chances of better outcomes.

AUTHOR CONTRIBUTIONS

AN, LM, MJC: Study conception and design, data acquisition, analysis and interpretation, writing and critical review of the manuscript.

JP: Study design, data acquisition, analysis and interpretation, writing and critical review of the manuscript.

CC: Study conception and design, data interpretation, critical review of the manuscript.

All authors approved the final version to be published.

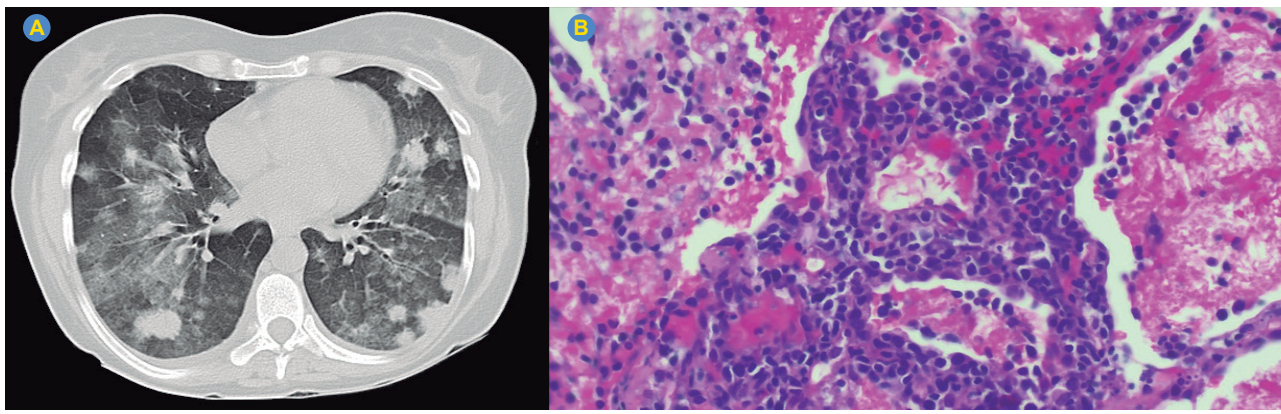

Figure 1 – Chest CT revealing multiple bilateral foci of consolidation, some of which with a nodular pattern and with air bronchogram, associated with areas of ground glass and mosaic pattern, with predominant distribution on the lower thirds of both lungs (A). Lung tissue with infiltration by small lymphoid cells with scant cytoplasm and slightly irregular basophilic nuclei, and necrosis (H&E, 400x) (B). Immunohistochemical staining revealed positivity for CD3, CD8, CD2, TIA-1, and BCL-2 and negativity for CD20, CD5, CD7, CD30, TdT, MUM-1, CD10 and BCL6. EBER was also positive (not displayed).

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in October 2024.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Consent obtained from the patient's legal representative.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

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Anemia Sideroblástica Congénita e Sobrecarga de Ferro em Idade Avançada

Congenital Sideroblastic Anemia and Iron Overload in Older Age

Palavras-chave: Anemia Sideroblástica/congénita; Hiperferritinemia; Sobrecarga de Ferro
Keywords: Anemia, Sideroblastic/congenital; Hyperferritinemia; Iron Overload

As anemias sideroblásticas (AS) enquadram-se num grupo de distúrbios da eritropoiese, caracterizado pela presença de sideroblastos em anel na medula óssea. As causas adquiridas incluem síndrome mielodisplásica, neoplasias mieloproliferativas, consumo de álcool, entre outras. Das causas congénitas, a anemia sideroblástica ligada ao X (ASLX) é a mais comum. Tipicamente resulta de mutações do gene *ALAS2* (envolvido na síntese do heme), cursa com anemia microcítica e afeta jovens ou adultos do sexo masculino. Alguns doentes respondem ao tratamento com vitamina B6.[1,2]

Descrevemos o caso de um doente do sexo masculino, com 67 anos, referenciado ao seviço de hematologia por hiperferritinemia, anemia microcítica e alterações no esfregaço de sangue periférico (ESP). O doente não tinha antecedentes transfusionais ou antecedentes familiares de anemia conhecidos e era assintomático. Do estudo efetuado, destacava-se: anemia microcítica hipocrómica [Hb 9,5 g/dL (VR: 13 - 18), HTC 33,8% (VR: 43 - 55), VGM 56,9 fL (VR: 87 - 103), MCHC 28,1 g/dL (VR: 28 - 36)], RDW 25,4% (VR: 11 - 16), eritrócitos 5,94 x 10^{12}/L (VR: 4,4 - 6,0), reticulócitos 0,96% (VR: 0,5 - 2,5), índice reticulocitário 0,5 (VR: > 2), sugestivo de distúrbios na maturação eritroide; ESP: dimorfismo eritrocitário; elevação das transamínases (3-4xLSN) e gama GT (2xLSN), ferro sérico 214 ug/dL (VR: 53 - 167), transferrina 172 mg/dL (VR: 200 - 360), ferritina 4665 ng/mL (VR: 20 - 250), saturação de transferrina (ST) 89% (VR: 20 - 50); ecografia abdominal: discreta esplenomegalia e esteatose hepática; eletroforese Hb normal; pesquisa de mutações genéticas associadas a hemoglobinopatias ou hemocromatose negativa. A ressonância magnética abdominal mostrou sobrecarga de ferro hepática [25,2 mg/g de peso seco (N < 2,0 mg/g)].

Foi instituída quelação de ferro com desferasirox 360 mg bid/PO. Após dez meses de tratamento, mantinha ferritina > 3000 ng/mL, ST 149%, Hb 9,5 g/dL e hepatoesplenomegalia, optando-se por introduzir desferoxamina 3x/semana/SC. Após três meses, verificou-se descida da ferritina para 1700 ng/mL, ST 92%, Hb e transamínases estáveis. O estudo genético, com painel do metabolismo do ferro e porfiria, detetou a variante *ALAS2.c.1447G>T;p.*