



HODGKIN'S LYMPHOMA MANIFESTED WITH BICYTOPENIA WITHOUT PALPABLE LYMPHADENOMEGALY

Linfoma de Hodgkin Manifesto Com Bicitopenia sem Linfadenomegalia Palpável

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Abstract: Introduction: Hodgkin's disease (DH) is a neoplasm of the lymphatic and reticuloendothelial system, responsible for about 30% of lymphomas and can be divided into two subgroups: (1) predominantly lymphocytic DH and (2) classical DH. The main clinical manifestation is adenomegaly mainly in the cervical and supraclavicular region, which may or may not be associated with systemic symptoms. The definitive diagnosis is made by biopsy followed by an analysis study of the lymph nodes involved. **Objective:** To report a case of manifest Hodgkin's lymphoma with anemia and thrombocytopenia without palpable lymph adenomegaly. **Methodology:** This is a cross-sectional, documentary, retrospective and descriptive study, case report type. The findings described in the medical record will be used as instruments. It is a young female patient with anemia refractory to clinical treatment, intermittent fever and severe thrombocytopenia requiring transfusion of packed red blood cells and platelets. At clinical examination there were no palpable lymph nodes. During the propaedeutic an abdominal tomography was performed that revealed retroperitoneal masses, from which the biopsy of these lesions was performed. The result of the immunohistochemical study of the parts confirmed Hodgkin's lymphoma. **Conclusion:** Hodgkin's lymphoma should be suspected in patients with thrombocytopenia and anemia, refractory to clinical treatment, without palpable lymph node.

Keywords: Neoplasms; Hodgkin Disease; Risk Factors.

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Resumo: Introdução: A Doença de Hodgkin (DH) é uma neoplasia do sistema linfático e reticuloendotelial, responsável por cerca de 30% dos linfomas. Sua principal manifestação clínica é a adenomegalia, principalmente em região cervical e supraclavicular. O diagnóstico definitivo é feito por biópsia seguida de estudo imuno-histoquímico dos linfonodos acometidos. **Objetivo:** Relatar um caso de Linfoma de Hodgkin manifesto de forma atípica com anemia e plaquetopenia sem linfadenomegalia palpável. **Metodologia:** Trata-se de um estudo com caráter documental, retrospectivo e descritivo, tipo relato de caso. Paciente sexo feminino, 31 anos, com anemia refratária ao tratamento clínico, febre intermitente e trombocitopenia grave, com necessidade de transfusão de concentrado de hemácias e plaquetas. Ao exame clínico não havia cadeias linfáticas palpáveis. A tomografia abdominal revelou massas retroperitoneais, a partir disso foi feita a biópsia dessas lesões. O resultado do estudo imuno-histoquímico das peças confirmou o Linfoma de Hodgkin. **Conclusão:** O Linfoma de Hodgkin deve ser um diagnóstico suspeitado em pacientes com trombocitopenia e anemia, refratárias ao tratamento clínico, sem linfonodomegalia palpável.

Palavras-chave: Neoplasias; Trombocitopenia; Linfoma de Hodgkin.

INTRODUCTION

Hodgkin's disease (DH) is a neoplasia of the lymphatic system and the reticuloendothelial initially described by Thomas Hodgkin in 1832. According to the classification described by the World Health Organization (WHO), t DH is divided into two subgroups: (1) DH lymphocyte predominance and (2) classical DH, being this subgroup divided into four subtypes: (a) nodular sclerosis; (b) rich in lymphocytes; (c) mixed cellularity; and (d) lymphocyte depletion.¹⁻³ Epidemiologically, DH is responsible for approximately 30% of the lymphomas, being that 95% belong to the subgroup of DH and the remaining 5% are DH lymphocyte predominance.¹

The main clinical manifestation of DH is the adenomegaly, characterized by lymph nodes usually hardened by associated fibrotic process. Among the main affected chains are, the cervical and supraclavicular (70%).^{2,3} The itching, pain in the lymph nodes after use of alcoholic drinks and the Pel-Ebstein fever (days of high fever alternated with days without fever) are some of the systemic symptoms observed.³⁻⁷

The definitive diagnosis of DH happens through the immunohistochemical study of the affected lymph nodes, with the presence of markers CD30 and CD15. For staging of the disease, it is used and internationally accepted the system of Ann Arbor modified by Costwolds conference in 1989. The most appropriate treatment today for DH is chemotherapy associated or not with radiotherapy.⁸⁻¹⁰

The objective of this study is to report a case of manifest Hodgkin's lymphoma with anemia and thrombocytopenia without palpable lymph adenomegaly. The design of this study was

approved by the Ethics and Research Committee, with the opinion of approval number 2.526.746. The research was conducted within the standards required by the Declaration of Helsinki.

CASE REPORT

Female patient, 31 years old, was admitted at a hospital because of severe anemia refractory to drug treatment and blood transfusion, associated with intermittent fever for twenty days - axillary temperature between 38°C to 39°C. Denied comorbidities, use of medications and allergies. Upon examination, the patient was conscious, regular general state, pale 3+/4+, LETIFICADA???? tissue perfusion. Heart rate: 110bpm, oxygen saturation: 98%, blood pressure: 90/70mmHg, Axillary temperature: 35.4°C. Evaluation of cardiovascular, respiratory and digestive systems without changes.

Requested laboratory examinations on admission: Hemoglobin: (Hb) 8.1; hematocrit (Ht): 27.1; VCM: 69; HCM: 21; RDW: 27; total leukocyte count: 5,000; platelets: 206,000. LDH: 575; Iron (Fe): 46; Ferritin: 766.7; Transferrin Saturation: 16%; PCR: Positive; total proteins: 7.3; Albumin: 2.8; Globulin: 4.5.

In the following days she remained pale, with no palpable lymph node. She presented hepatomegaly (palpable liver 2 cm away from the right upper quadrant) and extensive MACICEZ upon percussion in left hypochondrium. It has been suggested the hypothesis of visceral leishmaniasis, whose rapid test result of Leishmaniasis was not reactive. Total abdominal ultrasonography was also requested which evidenced hepatosplenomegaly. After hematology investigation was carried out with a myelogram to inves-

tigate the bicitopenia and assess the possibility of false-negative test for leishmaniasis. The patient evolved with gingival bleeding while performed the dental hygiene, in addition to petechiae in the upper limbs. On that occasion Prednisone was prescribed and platelet count and new laboratory examinations were performed (Hb: 8,9; Ht: 28.7; VCM: 72; HCM: 22; RDW: 23; total leukocyte count: 4710; platelets: 18,500; Prothrombin Time: 13.8 seconds; Prothrombin activity: 63%; RNI: 1.31). Thrombocytopenia propaedeutic was initiated and a computed tomography of the abdomen (TC) was requested to investigate the source of the fever. Afterwards, new blood transfusions of platelets concentrate were necessary because of persistent severe thrombocytopenia, being also required transfusion of red blood cells concentrate.

The result of TC showed two retroperitoneal masses, due to which the patient was referred to the surgical center for performing laparotomy. During the procedure hepatosplenomegaly and lymph adenomegalies in hepatoduodenal hepatogastric and para-aortic ligaments, being performed excision of two of them and anatomopathological analysis. Procedure without intercurrents and patient was referred to ICU.

The result of the immunohistochemistry revealed atypical lymphoid proliferation in lymph nodes consistent with Classic Hodgkin lymphoma type Mixed Cellularity. After improvement of thrombocytopenia and anemia she was discharged from the hospital with referral to start of treatment of Hodgkin lymphoma in specialized service and hematological monitoring series.

DISCUSSION

As it is described in the literature, t DH has

two peaks of incidence,¹⁻³ the patient in question does not fall within the described epidemiological age range and has the second most common subtype, Mixed cellularity (25%), which is characterized by the presence of numerous Reed-Sternberg cells dispersed in an inflammatory infiltrate (reactive histiocytes, plasmocytes, small lymphocytes, eosinophils, besides some foci of necrosis).³⁻⁷

Contrary to what is seen in most patients with DH,²⁻⁵ the patient from the study showed no palpable lymph node chains in cervical and supraclavicular regions, but it was observed mediastinal and subdiaphragmatic involvement that appears in less than 3% of cases and is more observed in middle-aged men.³⁻⁸

According to the literature, in one third of the patients the “B symptoms” occur - fever greater than 38 degrees °C, night sweats and weight loss greater than 10% - that are associated with a poor prognosis. Only t fever was observed in the patient, this being of Pel-Ebstein, which in turn is more common in patients of advanced age.⁷⁻⁹

Laboratory changes as normocytic and normochromic anemia is commonly found in patients with DH as well as leukocytosis, lymphopenia and thrombocytosis, manifestations which are immune-mediated that have connection with poor prognosis.⁷⁻⁹ In some cases, HFD evolves with unusual symptoms, such as cutaneous disorders (ichthyosiform atrophy and erythema nodosum), effects on the central nervous system, intense and unexplained pruritus, nephrotic syndrome, hypercalcemia, autoimmune hemolytic anemia, pain in the lymph nodes with the consumption of alcohol and thrombocytopenia.⁸⁻⁹ Among these changes, it was observed in the patient bicytopenia, due to hypersplenism and the medullar infiltrate.

Because of the provenance of the patient, as well as the signs and symptoms presented

by her and results of laboratory examinations, it was possible to suggest new diagnostic hypotheses, among them, parasitic diseases such as leishmaniasis and hematologic diseases.

Hematological Leishmaniasis, due to its high incidence, wide distribution in Brazil, the possibility of severe clinical forms and highly compatible with the symptoms presented by the patient, was the main diagnostic hypothesis. One of the techniques capable of diagnosing the disease is through the myelogram, performed by the patient in the study, whose result was possible to discard not only the Leishmaniasis (absence of hypergammaglobulinemia) as well as other hematological causes diseases, such as leukemia, through the analysis of the morphology of the cells and use of cytochemical evidences.¹⁰

According to the system of Ann Arbor for staging,^{2,3,11} The patient was in stage IVB, which denotes the severity of the disease. In studies carried out, adverse prognostic factors were identified, related closely with decreased survival of patients at this stage. Among those presented by the patient, we can mention: stage IV disease; hemoglobin level below 10.5 g/dL; and albumin level below 4 g/dL.^{11,12}

The thick needle biopsy can be used as a diagnostic method, but the recommendation is, in general, that be performed excisional lymph node biopsy, as happened with the patient. Fine-needle biopsy, although is widely used in the diagnosis of malignant tumors, its role in the diagnosis of lymphoma is controversial. As also recommended by the literature^{9,11} the patient underwent an immunohistochemistry evaluation which confirmed the diagnosis.

CONCLUSION

The case report presented a patient diagnosed with Hodgkin lymphoma from atypical symptoms, after exclusion of other differential diagnoses.

Hodgkin's lymphoma should be suspected in patients with thrombocytopenia and anemia, refractory to clinical treatment, without palpable lymph node. Because this is a neoplasm with a high rate of cure, it is of utmost importance the knowledge about the signs and symptoms of the disease, including more rare clinical manifestations, which will contribute to an appropriate clinical behavior, as well as deepening in the investigation of differential diagnoses.

CONTRIBUTIONS

All authors participated in the conception and design of the study, analysis and interpretation of data, critical writing or revision of intellectual content of the manuscript, final approval of the version to be published, and are responsible for all aspects of the work including the guarantee of its accuracy and integrity.

Declaration of Conflict of Interests: Nothing to declare.

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