RELATO DE CASO

Tricoleucemia: opções de tratamento, ênfase no regime ambulatorial

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Tricoleucemia (TL) é uma doença linfoproliferativa de células B que ocorre em adultos. TL aparece geralmente em pacientes de meia idade, em uma proporção de homem: mulher de 4,5: 1. TL caracteriza-se clinicamente por citopenias, esplenomegalia e infecções. As células expressam imunoglobulinas e marcadores de células B (CD19, CD20). O diagnóstico quase sempre requer uma biópsia de medula óssea, imunofenotipagem ou imunohistoquímica e mielograma. Confirmação da origem da infiltração é obtida por imunohistoquímica usando CD22 e/ou DBA 44 e um anticorpo para fosfatase ácida tartarato resistente(TRAP). Imunofenótipo típico requer a presença de quatro antígenos: CD11c, CD25, CD103 e HC2. Quando três ou quatro destes marcadores estão presentes, permite- se a distinção entre TL e outras doenças de células B. Reticulina está, via de regra, aumentada e isto explica a dificuldade de obter amostra de aspirado da medula óssea, que freqüentemente é seco ("dry tap") (2,4). As drogas utilizadas na TL compreendem; pentostatina e cladribina. Ambas induzem uma alta taxa de remissão completa (>80%), sendo duradoura na maioria dos pacientes. A sobrevida global da TL é de 95% em 5 anos. A maioria dos pacientes que recaem entram em segunda remissão com retratamento. Tanto a pentostatina quanto a cladribina são bem toleradas e o único efeito por longo tempo é linfopenia, por pelo menos um ano (4). O esquema clássico de administração da Cladribina requer hospitalização, devido a infusão contínua por sete dias, sendo este esquema tão eficaz e eficiente quanto os esquemas ambulatoriais (3,4).

Em abril/2002, paciente de 68 anos foi avaliada por apresentar astenia, palidez cutâneo-mucosa e hepatoesplenomegalia vultuosa, com hemograma mostrando pancitopenia (Hb:8,7g%; leucócitos: 600/mm^3; plaquetas:73.000/mm^3). Realizado mielograma e biópsia de medula óssea, que demonstraram células linfóides atípicas compatíveis com TL (fig. 1). A imunofenotipagem da medula óssea confirmou o diagnóstico. Iniciado tratamento ambulatorial com Cladribina semanal, durante seis semanas. Após sete semanas do início da quimioterapia, o hemograma mostrava melhora da pancitopenia (Hb:9,5g%; leucócitos:2000/mm^3 sendo 1760 neutrófilos e 240 linfócitos; plaquetas: 110.000/mm^3). Oito meses após tratamento a paciente encontrava-se em remissão hematológica (Hb: 12,2g%; leucócitos: 4500/mm3; plaquetas: 105.000/mm^3), não apresentando visceromegalias. Nesta paciente foi administrada cladribina pela facilidade do esquema ambulatorial, sem necessidade de internação da mesma. A droga induziu remissão clínica com mínima toxicidade e com comodidade para a paciente.

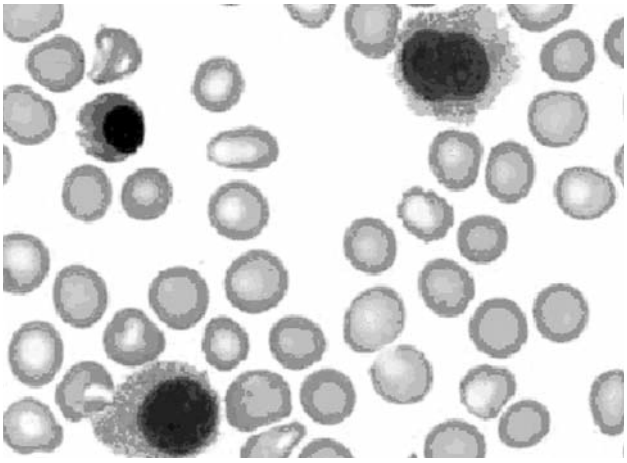

Figura 1

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5. Wooo, Alanstair J.J. Newer Purine Analogues for treatmente of hairy cell leukemia. N Engl J Med; vol 330:691-697.

Case Report

Hairy Cell Leukemia: therapeutic options, emphasis on ambulatorial regimen

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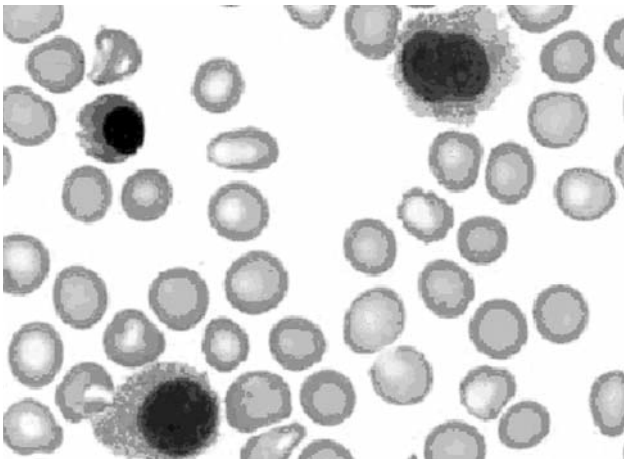
HCL is a rare lymphoproliferative disorder of B cells that occurs on adults. The incidence is about 2% of leukemias in general and 8% considering only mature leukemias of B and T cells, including non-Hodgkin lymphoma with lymphocytosis (5000 cells/ mm^3). HCL is 6 to 10 times less common than LLC. HCL happens usually in middle-aged patients, in proportion men: women of 4,5: 1. A big interest about this disease studying was concomitant to the discovering of powerful therapeutic agents: alpha-interferon and pentostatin in the 80's and cladribine in the 90's. (1-5)

HCL is clinically characterized by cytopenias, enlarged spleen and infections. The cells express immunoglobulins and B cell markers, such as CD19 and CD20. The definitive diagnosis almost always requires a bone marrow biopsy, immunophenotyping or immunohistochemical and mielogram. The confirmation of the source of infiltration is obtained by immunohistochemical using CD22 and/ or DBA 44 and an antibody to tartrate-resistant acid phosphatase (TRAP). Typical immunophenotype requires the presence of four antigens: CD11c, CD25, CD103 e HC2. When three or four of them are present, the distinction between HCL or other B cells illness is allowed. Reticulin is usually increased, and that explains the difficulty to obtain bone marrow aspirated samples, that are often dry ("dry tap") (2, 4). The drugs used on hairy cell leukemia are ; pentostatin and cladribine. Both agents induce a high complete remission rate (> 80%), being stable on most patients. Actual overall surviving rate of hairy cell leukemia it's in about 95 to 98% in 5 years. Most of the patients with falling back go into a second remission with re-treatment. Patients that do not answer this therapy are rare. Those with abdominal lymphadenopathy or falling back have a smaller answer to both agents. The two drugs are well-tolerated and the only long-term effect is lymphopenia, for at least one year (4). Once the diagnosis of hairy cell leukemia is done and the intervention indicated, the therapy must be initiated. The standard schedule for cladribine requires hospitalization due to it's continuous infusion for seven days, being such efficient as out-patient managements (3, 4).

In April 2002, a 68 year-old patient was with asthenia, paleness and hepatosplenomegaly with blood counting showing pancytopenia (Hb: 8,7g%; leukocytes: 600/ mm^3; platelets: 73000/ mm^3). Mielogram and bone marrow biopsy were done, and they demonstrated atypical lymphoid cells compatible with hairy cell leukemia at both (figure 1); bone marrow immunophenotyping was also done and confirmed diagnosis.

The patient started treatment with cladribine on out-patient protocol of chemotherapy, on week sessions, for six weeks. After seven weeks from the beginning of chemotherapy, blood counting showed improvement on pancytopenia (Hb: 9,5g/dl; leukocytes: 2000/ mm^3, with 1760 neutrophiles and 240 linfocits; platelets: 110000/ mm^3). Eight months after therapy the patient was on hematological remission (Hb: 12,2 g/dl; leukocytes: 4500/ mm^3; plaquets: 105000/ mm^3), without visceromegalies.

For this patient cladribine was dispensed, because of the facility on out-patient management, without necessity of hospitalization. The drug leads to clinical remission with minimum toxicity and with the patient's comfortableness.



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