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PSEUDO-GAUCHER CELLS IN THE BONE MARROW OF A PATIENT WITH CENTROCYTIC NODULAR NON-HODGKIN'S LYMPHOMA

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Storage histiocyte disorders are inborn errors of metabolism caused by enzyme deficiencies. Gaucher's disease is the most prevalent lysosomal storage disorder, but cells indistinguishable by light microscope examination from typical Gaucher cells (pseudo-Gaucher cells) have been observed in many hematologic and non hematologic disorders. We report a case of centrocytic nodular non Hodgkin's lymphoma (NHL) in which pseudo-Gaucher cells were seen in bone marrow biopsy after systemic relapse.

Clinical and laboratory findings

A 28-year-old man with centrocytic nodular NHL according to the updated Kiel classification,[1] stage III A, underwent complete remission after 6 cycles of a CHOP-like regimen. After three years the patient suffered lymph node relapse. During the staging examinations for relapse, bone marrow biopsy showed a paratrabecular lymphomatous infiltration surrounded by numerous abnormal storage cells that strikingly resembled Gaucher's cells (Figure 1). The patient was treated with the VACOP-B protocol and then submitted to consolidation therapy with autologous bone marrow transplantation (ABMT). Bone marrow biopsy performed after ABMT detected no lymphoma and the pseudo-Gaucher cells had also disappeared from the marrow specimen.

Morphology and histochemistry

We evaluated the morphology of a bone marrow biopsy after relapse. The bone marrow trephine specimen was embedded in glycol methacrylate resin (JB-4 Kit, Polysciences, Inc) and cut into 1.5 μm sections which were stained with Giemsa; PAS, silver impregnation and the Prussian blue reactions were carried out. The bone marrow trephine sections were normocellular with a paratrabecular lymphocyte infiltration. Around the lymphoma proliferation there were abnormal pseudo-Gaucher-like storage cells (Figure 1). Light microscope examination revealed that these cells had a diameter of approximately 20-30 μm, pale cytoplasm with dense round deposits inside and a single eccentric nucleus (Figure 2).

The pseudo-Gaucher cells were positive for acid phosphatase after tartrate treatment (Figure 3); reticulin fibrosis was not prominent in the areas of the pseudo-Gaucher cells and the PAS, naphthol AS-D chloroacetate esterase and Prussian blue reactions were negative in these cells.

Conclusions

Pseudo-Gaucher cells have been described in several diseases, including hematologic disor-

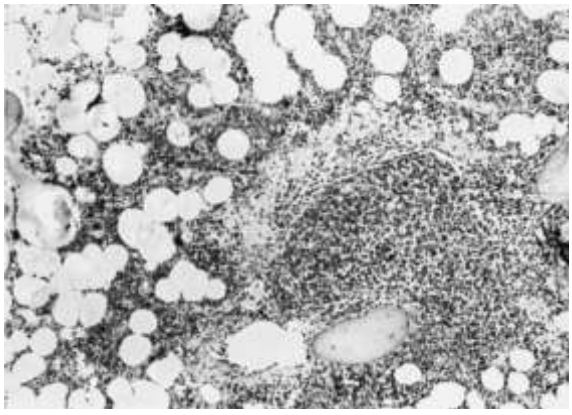

Figure 1. Bone marrow trephine section. Paratrabecular lymphoid proliferation surrounded by large storage cells. Giemsa 10×.

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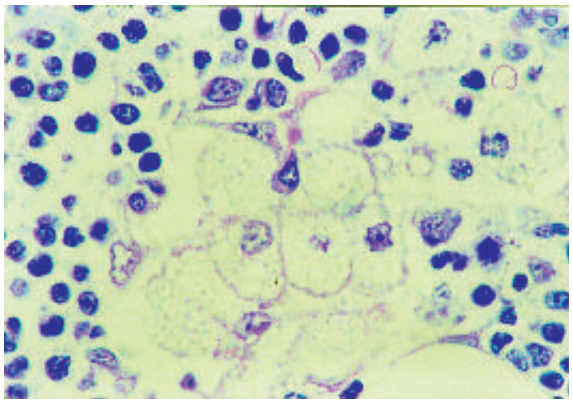

Figure 2. Bone marrow trephine section. Large pseudo-Gaucher cells with dense round deposits and a single eccentric nucleus. Giemsa 100×.

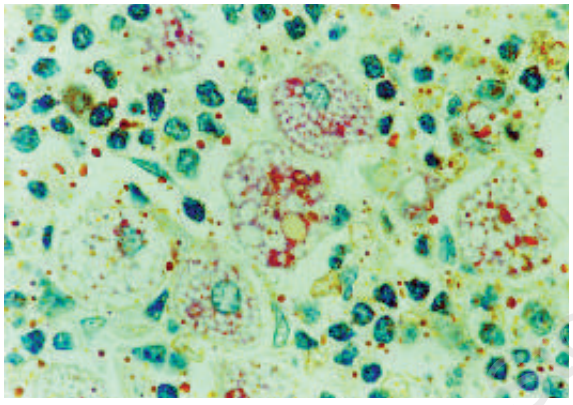

Figure 3. Bone marrow trephine section. Weak positivity with acid phosphatase after tartrate treatment. Leukocyte acid phosphatase 100 X.

ders (chronic myelogenous leukemia, acute myeloid leukemia, idiopathic thrombocytopenic purpura, aplastic anemia, plasma cell myeloma, chronic lymphocytic leukemia, thalassemia major, Hodgkin's disease and immunoblastic non-Hodgkin's lymphoma),[2-5] bacterial infections (*Mycobacterium avium* in AIDS),[6] and autoimmune disease (rheumatoid arthritis).[7]

These cells had never been described in centrocytic nodular lymphoma. In our patient, there was no family history or evidence of inherited Gaucher's disease, and serum β-glucosidase activity was elevated. Furthermore, the patient showed no signs of infectious disease. The disappearance of the pseudo-Gaucher cells after successful induction of complete remission indicates that these cells and the lymphomatous bone marrow proliferation were linked. As postulated by Zidar *et al.*[8] and by Papadimitriou *et al.*,[9] these storage cells probably result from excessive cell breakdown due to malignant lymphoma cells or tumor necrosis. Thus in these cases there is a relative rather than a true deficiency of β-glucosidase, as occurs in inherited Gaucher's disease.

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