

Sea-blue histiocytes in the bone marrow of a patient with Niemann-Pick disease type B

Sea-blue histiocytosis is a morphologic finding that can be associated with both hematologic conditions in which an increased cellular turnover is present (myelodysplastic syndromes, myeloproliferative disorders) or inborn errors of lipid metabolism. We report a case of sea-blue histiocytosis associated with a mild phenotype of Niemann-Pick disease type B (NPDB) in a 44-year old Caucasian man who presented with splenomegaly and mild thrombocytopenia without other signs or symptoms. A bone marrow aspirate (Figure 1) and biopsy (Figure 2) showed the presence of histiocytes with sea-blue cytoplasm. Two different measurements of acid liposomal sphingomyelinase activity were below normal (1.81 and 4.77 nmol/mg, normal values > 9.85 nmol/mg). These findings were pertinent with the diagnosis of NPDB, mild phenotype. This rare inborn error of metabolism becomes evident during adult age, classically with splenomegaly, thrombocytopenia and with evidence of sea-blue histiocytes in the bone marrow.¹

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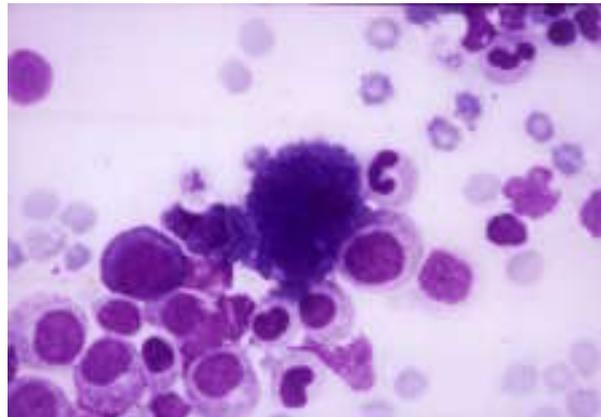


Figure 1. Bone marrow aspirate: sea-blue histiocyte among normal haemopoietic cells; typical sea blue color by Giemsa (May-Grünwald-Giemsa × 1,000).

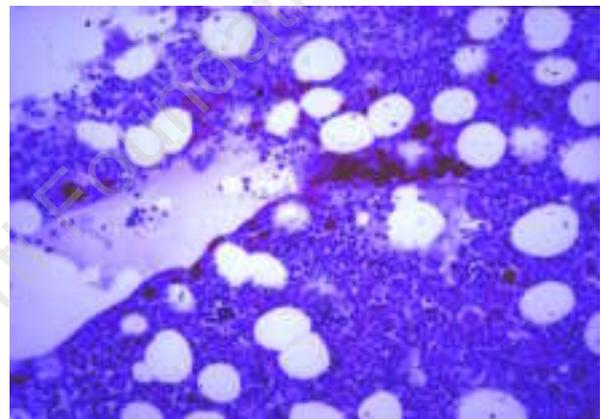


Figure 2. Bone marrow biopsy: sea-blue histiocytes by a blood vessel. Periodic acid-Schiff (PAS) positive (× 200).