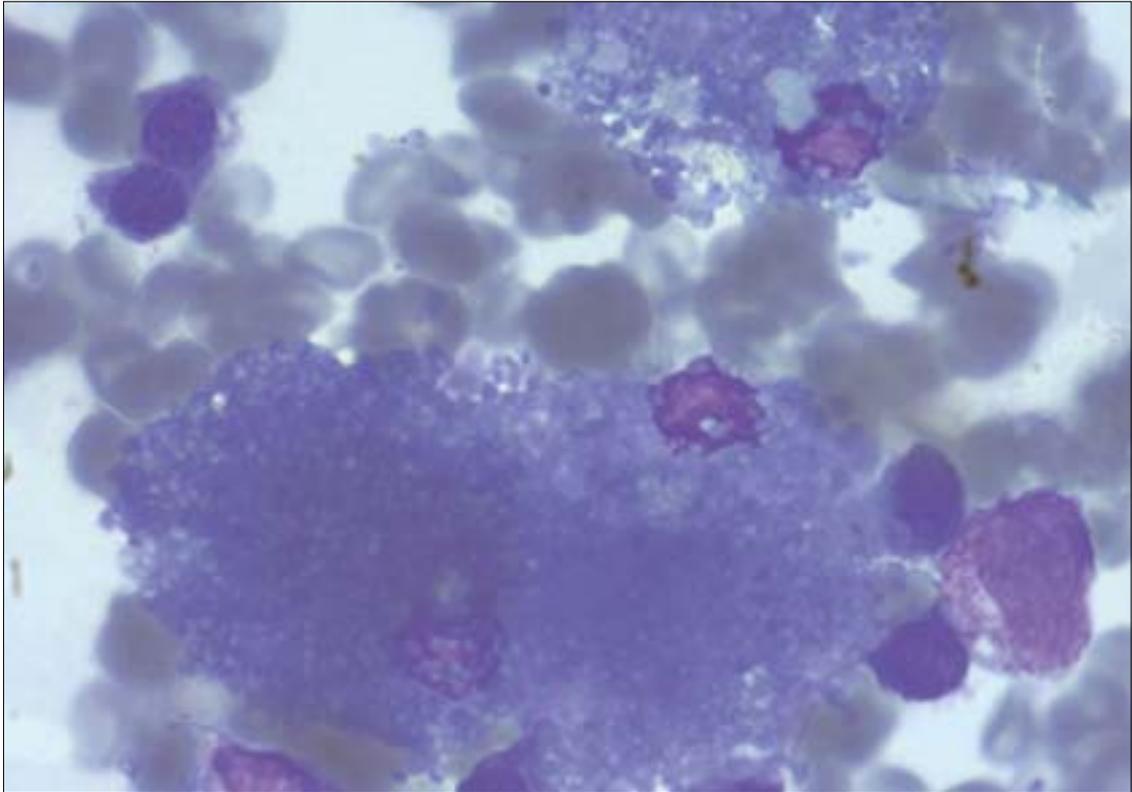


## Sea-blue histiocytes in bone marrow aspiration sample: what is the importance?

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**Figure.** Sea-blue histiocytes in bone marrow aspiration sample.

A 45-year-old woman was investigated in our hospital because of hepatosplenomegaly and mild leukopenia ( $2820/\text{mm}^3$ ) and thrombocytopenia ( $117,000/\text{mm}^3$ ). There were no signs of hepatic viral, autoimmune or other etiologies which could explain hepatosplenomegaly. Bone marrow aspiration and biopsy showed normocel-

lular marrow with no maturation arrest of myeloid cells. There were atypical macrophages, the cytoplasm of which were distended and closely packed with fine granules staining blue with Giemsa. The nuclei were centrally located in some cells and displaced towards the periphery. The features were consistent with sea-blue his-

tiocytes (Figure). Liver biopsy showed foamy cell infiltration. Splenic sequestration was shown by performing a radionuclide imaging test. On the basis of these findings, diagnosis of Niemann-Pick disease type B was highly suspected, so the activity of acid lysosomal sphingomyelinase in the leukocytes was measured, and was determined as 7.34 nmols/mgprotein, which was below the normal range (34–78 nmols/mgprotein). These findings suggested sea-blue histiocytosis in bone marrow due to Niemann-Pick disease type B. Due to storage disease, splenic enlargement and hypersplenism occurred and cytopenias developed.

The sea-blue histiocytes are macrophages that have large cytoplasm, empty vacuoles, and sea-blue granules containing lipofuscin or ceroid. The term “primary sea-blue histiocyte syndrome” has been reserved for those conditions in which there is a significant accumulation of these cells in multiple organs with no predisposing disease. This syndrome is a chronic benign illness, which

is characterized by splenomegaly and thrombocytopenia. Sea-blue histiocytes can be seen in various diseases in which there is an increased hemopoietic cellular turnover, like chronic myeloid leukemia and other myeloproliferative disorders, myelodysplastic syndromes, severe autoimmune neutropenia and immune thrombocytopenic purpura. They can also be seen in lysosomal and lipid metabolic defect disorders.

Niemann-Pick disease is an autosomal recessively transmitted congenital metabolic disorder that includes three types (A,B,C). Type B is a benign disorder characterized by splenomegaly, thrombocytopenia, and sea-blue histiocytes in bone marrow. Rarely, liver and lungs are involved. The nervous system is spared. When sea-blue histiocytes are seen in bone marrow of an adult patient with splenomegaly and thrombocytopenia, Niemann-Pick disease type B should be considered as the most possible diagnosis and sphingomyelinase activity in the leukocytes must be measured.