ALDER-REILLY ANOMALY IN PATIENTS WITH MUCOPOLYSACCHARIDOSIS VI

GRANULAÇÕES DE ALDER-REILLY EM PACIENTES COM MUCOPOLISSACARIDOSE VI

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Several authors have described the occurrence of coarse granular cytoplasmic inclusions (the "Alder-Reilly Bodies"; ARB) in leukocytes of patients with Mucopolysaccharidosis VI (MPS VI; OMIM +253200). Aiming at estimating the frequency of ARB in MPS VI, we analyzed the reports of the peripheral blood counts routinely performed in the MPS VI patients at our hospital from 2002 to 2003 (35 reports of 17 patients; 9/17 patients had two or more evaluations performed in this period). No patient was on enzyme replacement therapy at the time of the blood sampling. The analysis of blood cells was first performed with automation in haematology (Pentra 120-ABX). Whenever automation was suggestive of abnormalities in the white cells, a blood smear of the same sample, stained with May-Grünwald Giemsa, was therefore analyzed by optic microscopy (OM); if ARB were found by OM, their presence were referred in the reports. ARB were reported at least once in 13/17 patients, mainly in neutrophils. No other abnormality was reported in the tests. Out of the 9 patients that had more than one test performed, only 3 showed similar results on both tests (e.g., presence or absence of ARB). ARB seems to be a frequent finding in patients with MPS VI.

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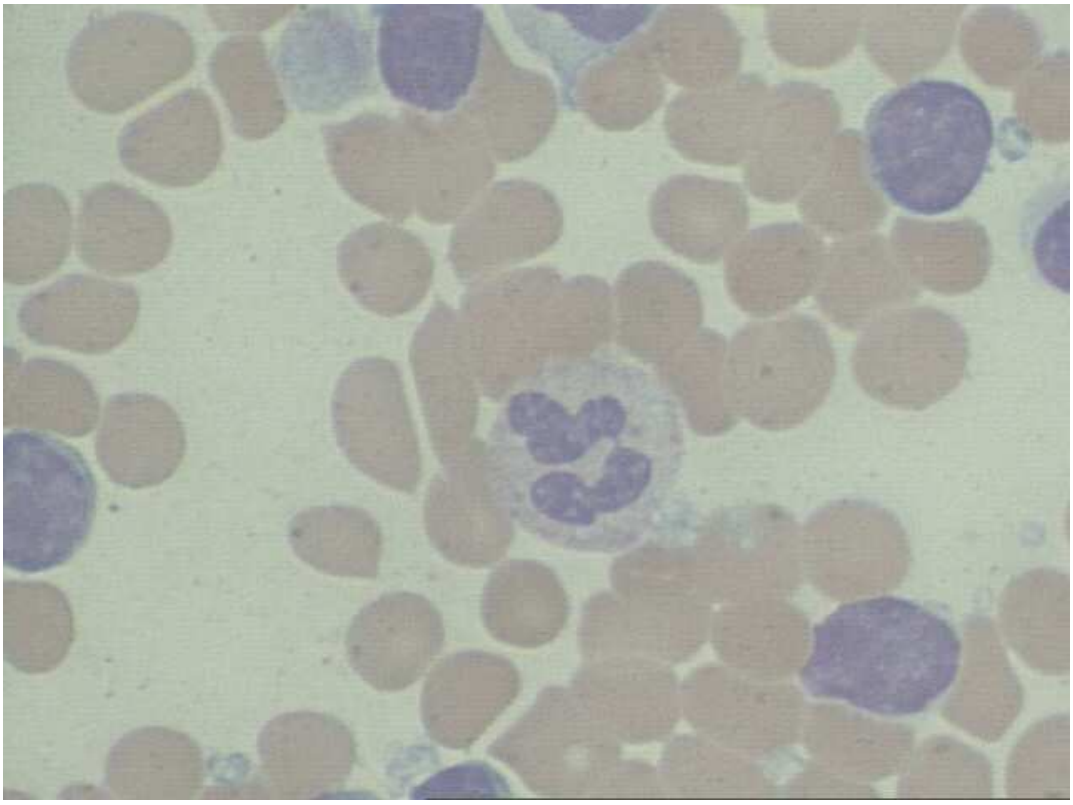

Figure 1 - Peripheral blood, May-Grünwald Giemsa staining, optic microscopy x1000.
Neutrophil showing no abnormalities in morphology (normal child)

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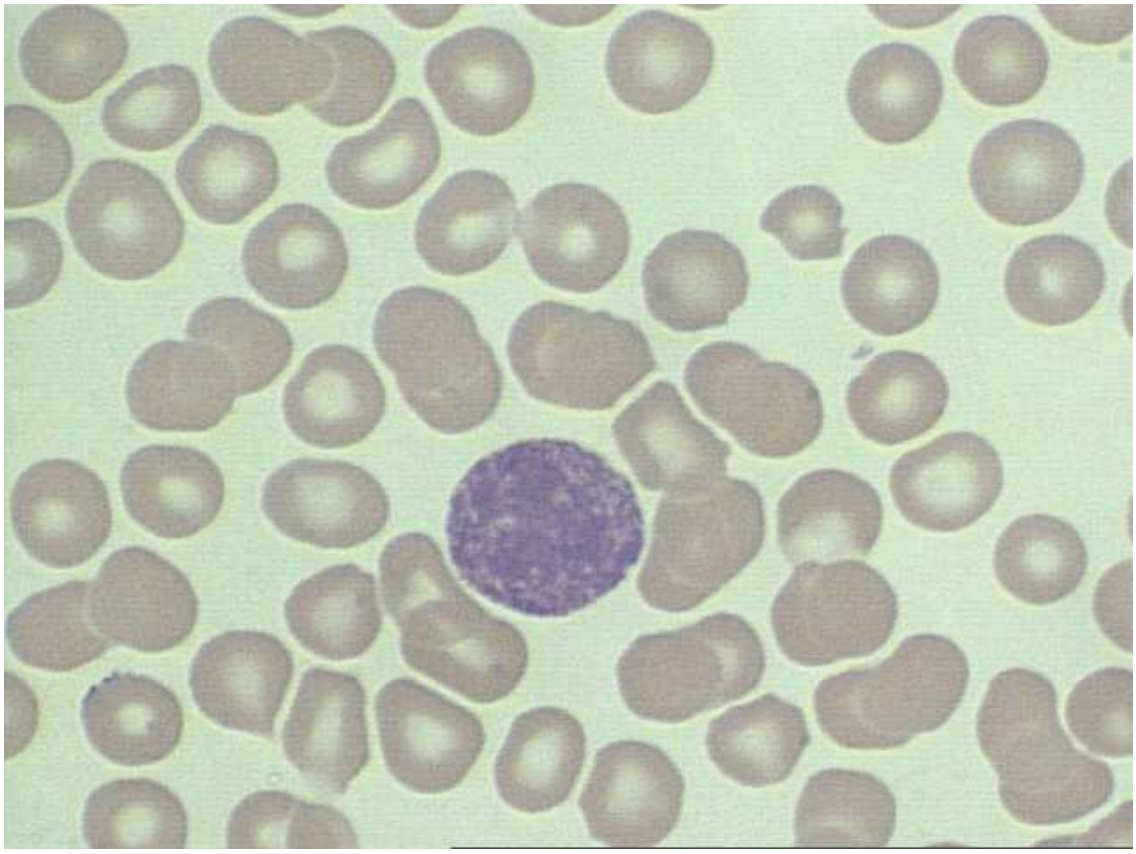

Figure 2 - Peripheral blood, May-Grünwald Giemsa staining, optic microscopy x1000. Neutrophil showing the Alder-Reilly Bodies (Brazilian MPS VI patient).

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