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Acta Medica Iranica
2009;47(4) : 155-158

Griscelli Syndrome Type 2; A Pediatric Case with Immunodeficiency

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Abstract:

A 3.5 month-old girl was admitted with silvery gray hair, light colored skin, recurrent diarrhea, chest infections, hepatosplenomegaly, episodes of pancytopenia, and hemophagocytosis in the bone marrow. Light microscopy of hair showed characteristic large and irregular clumps of melanin in the middle of hair shaft. Peripheral blood smear examination did not show giant granules in granulocytes. On the basis of these clinical and laboratory findings, Griscelli syndrome was diagnosed. The child succumbed to infection during an accelerated phase of the disease.

Keywords:

Griscelli Syndrome ، Phagocyte Disorders

TUMS ID: 3876

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