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# Acute Myeloblastic Leukemia with Pseudo Chediak Higashi Granules: A Case Report

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***Abstract*—** *A 62-year-old man presented with acute leukemia secondary to chemotherapy had giant inclusions in the cytoplasm of the granular lineage at the medullar level, These granules resembled those observed in the hereditary Chediak Higashi syndrome.*

***Keywords*—** *Acute myeloblastic leukemia , granules, pseudo Chediak Higashi, MDS.*

## I. Introduction

Pseudo-Chediak–Higashi (PCH) granules are giant cytoplasmic inclusions in myeloblasts or myeloid precursors, resemble those observed in hereditary Chediak-Higashi syndrome characterized by partial oculocutaneous albinism, variable cellular immune deficiency and lethal lymphohistiocytic activation phases in the absence of a bone marrow transplant. They are commonly described in acute myeloblastic leukemia (AML), in chronic myeloid leukemia, myelodysplastic syndromes and some other cases of acute lymphoblastic leukemia [1,2].

We report the case of acute myeloblastic leukemia secondary to chemotherapy with Pseudo-Chediak-Higashi inclusions in the granular line diagnosed in a 62-year-old man.

## II. Case Report

62 year old man, with a history of chronic smoking, hypertension under monotherapy for 7 years and operated for an anal fistula in 1997; followed for 3 years for diffuse non-Hodgkin B lymphoma with large colonic cells, stage 1E, IPI 0, revealed by abdominal pain. Osteo-medulary biopsy is without tumor infiltration, progress was monitored on CT (computed tomography). Therapeutically, he received 6 RCHOP cures, achieving complete remission.

The patient presented progressive worsening cytopenias with a LDH (Lactate desydrogenase) level slightly higher than normal (378> VN = 320 IU / l), still in complete CT remission.

The hemogram detected pancytopenia with neutropenia (PNN: 0.45G / L) and anemia (Hb: 6.5 g / dL), macrocytic (VGM: 102 fl) and argenative (RETIC: 50 G / L), with discreet thrombocytosis (platelets: 478 G / L). The rest of the assessment was without particularities the exception of an inflammatory syndrome (CRP = 83g / L). The blood smear did not find any circulating blasts.

A myelogram is performed and finds a significant medullary invasion by (43%) medium to large blasts, with high cytoplasmic nucleo ratio; sometimes fine chromatin sometimes mottled, basophilic cytoplasm sometimes showing an archoplasm and often an Auer rods. Pseudo-Chediak-Higashi granules were noted in the granular line (fig. 1), Myelo Peroxidase negative. multilineage dysplasia suggesting a myelodysplastic syndrome has been noted. karyotype was carried out in favor of a complex karyotype AML.

He patient was put on induction chemotherapy (Doxorubicin) with clinical stability.

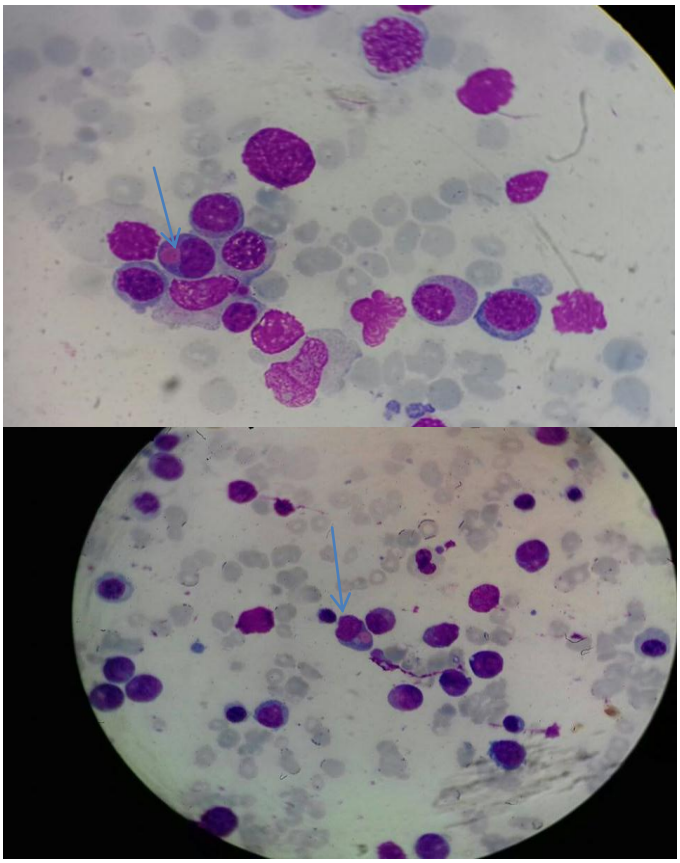

Figure 1: Image showing pseudo chediak higashi granule (Staining: May-Grunwald-Giemsa. Magnification: x 1000 ; Laboratory of Hematology – HMIMV)

## III. Discussion

Pseudo-chediak higashi granules were first described in 1964 by Didisheim and al in a patient with acute



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promyelocytic leukemia [3]. Later, VanSlyck and Rebuck [4] reported similar granules in the leukemia cells of two patients with acute myelomonocytic leukemia, and used the term, pseudonym Chediak-Higashi abnormality, due to the resemblance of the granules to those observed in patients with hereditary Chediak-Higashi syndrome.

The PCH anomaly is characterized by the presence of large cytoplasmic eosinophilic granules in blasts, promyelocytes and myelocytes. It is most often observed in patients with AML(Acute Myeloide Leukemia) M2, M3, M4 and M5, but it is also associated with chronic myeloid leukemia, myelodysplastic syndrome and mixed line leukemias [5]. Most of the reported cases are in adults; with a few cases reported in children.

Auers rods were also present in many cases, in addition to these granules while in others, only granules were present [6, 7,8].

By cytochemistry, these inclusions were positive for peroxidase in all the cases reported, with rare exceptions. It is believed that the origin of these inclusions is the result of the fusion of azurophilic granules, in electron microscopy, they have been found to contain many microcrystalline structures such as those of Auer rods. Tsai and al. thought that the anomaly may be a manifestation of an underlying abnormal granulogenesis, and that it may be associated with increased susceptibility to infections due to defective leukocyte function [6].

In children, the presence of these inclusions does not seem to be associated with specific clinical characteristics, with a prognosis similar to that of cases without abnormality. However, no studies have been done in adults [9].

## IV. Conclusion

The pseudo-Chediak-Higashi anomaly is rarely described, the significance of these granules remains largely unknown. It is interesting to note that these inclusions observed in the congenital Chediak-Higashi syndrome, are acquired in acute myeloblastic leukemias. Other molecular studies are necessary to establish the clinical, therapeutic and prognostic relevance of these granulations

*Conflicts of Interest:*
No.

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