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Acute Plasmacytoid Dendritic Cell Leukemia: A Morphological Diagnosis Is Often Difficult

M. SEXTIUS[1,2], M. BENSALAH[1,2] , C. FLEURY[3], P. LEMAIRE[4], M. CHAQUIN[4], L. CHEMALI[4], D. LUSINA[4], J.M. MARTELLI[4], and V. LEYMARIE[3]

1. Avicenne Hospital, Hematology Laboratory, GHUPSSD, Bobigny, France
2. Saint-Louis Hospital, Hematology Laboratory, Paris, France
3. Saint-Antoine Hospital, Hematology Laboratory, Paris, France

INTRODUCTION

Acute Plasmacytoid Dendritic Cell Leukemia (APDCL) is a rare neoplasm characterized by the malignant proliferation of plasmacytoid dendritic cells, with a very poor prognosis. The 4th revision of the WHO (World Health Organization) highlights the typical cytological form, and the 5th revision confirms the phenotypic profile. In about 80% of cases, the disease presents with localized or extensive, sometimes generalized cutaneous manifestations. Blood or bone marrow infiltration (in 60 to 90% of cases) appears at an early stage (sometimes later) with lymph node involvement in approximately 40% to 50% of patients. The typical cytological form is well known but there are different morphological variants that can mimic other hematological diseases, creating an initial diagnostic challenge.

METHOD

We grouped 16 patients (M/F = 14/2, aged 14 to 87 years), for whom a definitive diagnosis of APDCL was confirmed based on their immunophenotypic profile in flow cytometry : CD4+, CD56+, CD123+, sometimes with the expression of one or more aberrant markers of T lineage (CD2 or CD7) or myeloid lineage (CD33).
All patients in our cohort presented with cutaneous involvement at the time of diagnosis, sometimes associated with a tumor syndrome.
We separated the patients into different subgroups based on their cytological presentations.

RESULTS

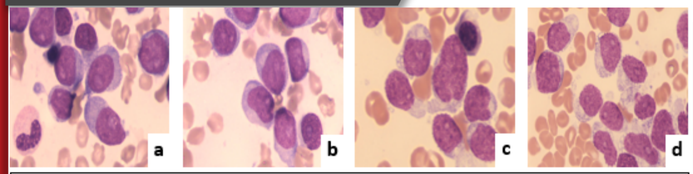


Figure 1 : Typical forms

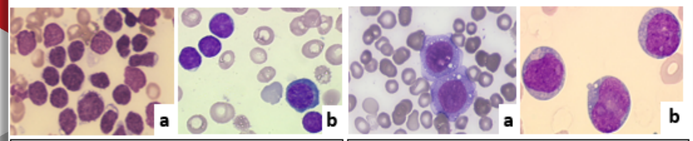


Figure 2 : « Lymphoblastic acute Leukemia Like »

Figure 3 : « Acute Monoblastic Leukemia Like »

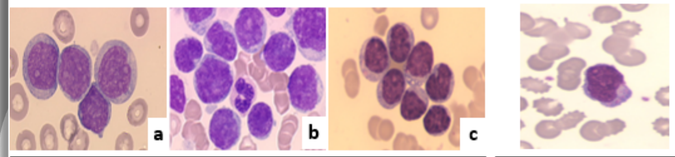


Figure 4 : « Lymphoma-Like »

Figure 5 : « AML Like »

	Size	Cytoplasm	Nucleus	Chromatin
Typical Fig. 1 (a, b, c, d)	Medium to large Average nucleus/ cytoplasm ratio	Moderately basophilic Agranular Fine "string of pearls" vacuoles Plasmacytoid expansions with a "hand mirror aspect"	Regular and eccentric	Fine and nucleolated
« Lymphoblastic acute Leukemia Like » Fig. 2 a et b	Small to medium High nucleus/cytoplasm ratio	Sparse Poorly basophilic Rarely vacuolated	Regular	Intermediate to dense chromatin
« Acute Monoblastic Leukemia Like » Fig. 3 a et b	Medium to large Average nucleus/ cytoplasm ratio	Grizzled or basophilic Vacuolated Sparse granulations	Regular	Fine and nucleolated Sometimes folded
« Lymphoma Like » Fig. 4 a, b, c	Medium to large Average to large nucleus/cytoplasm ratio	Grizzled Vacuolated, poorly basophilic Sometimes sparse granulations	Heterogenoeous, possibly budding	Heterogenoeous
« AML Like » Fig. 5 2 a et b	Medium High nucleus/cytoplasm ratio	Poorly basophilic Often granular	Regular	Fine and nucleolated

CONCLUSIONS

- Many morphological variants are misleading and can mimic other hematological disorders.
- In atypical cytological forms : a minority of cells with typical morphology may be present, which can help the initial hypothesis.
- Understanding atypical situations is important in order to effectively guide the etiological workup from the beginning of patient care.

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Saint-Louis Hospital, Hematology Laboratory
Saint-Antoine Hospital, Hematology Laboratory

CONTACT INFORMATION

mellody.sextius@aphp.fr
vincent.leymarie@aphp.fr