P17. THE CLINIC AGRESIVITY AND EVOLUTION OF A PACIENT DISEASE DIAGNOSTICATED WITH BLASTIC PLASMOCYTOID DENDRITIC CELL NEOPLASM

Fetco Diana[2], Merticariu Amalia[2], Burcoveanu Cristina[2], Ghiorghiu Doramina[2], Dăscălescu Angelica[1]

1 UNIVERSITY MEDICAL AND PHARMACY „Gr. T. Popa" IASI

2 REGIONAL INSITUTE OF ONCOLOGY IASI

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare hematologic malignancy characterized by the clonal proliferation of precursors of plasmocytoid dendritic cells. The clinical features an evolution of BPDCN consist of two main patterns, one, 90% of cases characterized by an indolent onset dominated by cutaneous lesion followed by tumor dissemination the other, 10% cases, showing features of an acute leukemia with systemic involvement from the beginning. Also in these cases multiple skin nodules are frequently present.

On flow cytometry the expression of CD4, CD45RA,CD56 and CD123 is considered to represent the pathognomic phenotype.

No specific chromosomal aberration have been identified until now.

Case is of a man who shows multiple tumor located the anterior and posterior thorax, scalp, legs and hands, ranging in size from 5/6 cm to 10/15 cm, with aspect of ulcerative necrotic burjonate some containing worms, painless. BPDCN was diagnosed based on AP and IHC examination of skin biopsy and chemotherapy was initiated with initial favorable trend but subsequently encumbered impaired consciousness due to solid tumor localized parieto-occipital right and left cerebellum.

Despite the apparently indolent clinical presentation, the course is aggressive and the median survival is approximately 12-14 months.

At present, there is no consensees for optimal treatment of BPDCN. With intensive therapy for acute leukemia the rate of remission increases, but only allogeneic bone marrow the first remission is a chance of long term survival.