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Myelodysplasia and autoimmunity.

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PURPOSE OF REVIEW: Primary myelodysplastic syndromes (MDS) are heterogeneous clonal haemopoietic stem-cell disorders clinically presented with a varying degree of peripheral cytopenias and an increased probability of leukemic evolution. The natural history of MDS ranges from more indolent forms of disease spanning years to those rapidly progressing to overt leukemia. A distinct subset of MDS patients manifest overt autoimmune manifestations (AIMs), the pathogenesis and prognostic significance of which remain controversial. This review will briefly highlight aspects of immune-mediated myelosuppression and cytokine-induced cytopenias in MDS and further analyze MDS-associated AIMs clinically and pathogenetically. RECENT FINDINGS: Facts provided by advanced studies suggest that an immune reaction against the evolving clone, operated by macrophages, T, natural killer and other effectors contribute to ineffective and dysplastic MDS hemopoiesis. Despite the fact that several immunologic abnormalities have been described in MDS, the precise pathophysiologic mechanism underlying AIMs remains unclear. SUMMARY: The encouraging biological insights into the autoimmune component of MDS pathophysiology can lead to the development of novel forms of treatment for controlling MDS process. MDS with AIMs constitute an ideal model in the investigation of disordered immune function in preleukemic states.

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