MALIGNANCIES IN CHILDREN WITH IMMUNODEFICIENCIES

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Background. Primary immunodeficiency disorders (PIDs) are a heterogeneous group of genetic conditions characterized by impaired immune function. Recent studies have highlighted a complex connection between PIDs and an increased risk of malignancies. However, the underlying mechanisms remain poorly understood. **Objective of the study.** This study aims to understand the relationship between immunodeficiency and susceptibility to malignancy by reviewing literature sources. **Material and methods.** Data from online databases such as Pubmed, UpToDate, and the National Library of Medicine were used to review the literature examining factors associated with malignancy in patients with PID. **Results.** Cancer is the second most common cause of death in PID patients. The cancer risk in PID patients is estimated to be 4 - 25%, and some studies have reported a notable increase in standardized incidence ratios of various cancers. Non-Hodgkin lymphoma (48%) and Hodgkin lymphoma (10% of cases) are the most common cancers in PIDs, diagnosed often at younger ages. PID-related malignancies affect mainly ataxia-telangiectasia and common variable immunodeficiency patients over 50% of cases, another 30% linked to Wiskott-Aldrich syndrome, severe combined immunodeficiency and selective IgA deficiency. **Conclusions.** PID patients have a higher risk of many diverse cancers for genetic, immunological, and environmental reasons. Management is challenging, requiring continued surveillance and individualized treatment based on the underlying immunodeficiency and associated complications. **Keywords:** primary immunodeficiency, malignancy, tumor predisposition.

CLINICAL, PARACLINICAL AND EVOLUTIONARY FEATURES OF JUVENILE IDIOPATHIC ARTHRITIS, SYSTEMIC FORM

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Background. Systemic juvenile idiopathic arthritis (SJIA) is an inflammatory disease that can cause fever, arthritis, and sometimes rash. It can also cause extensive lymphadenopathy, hepatosplenomegaly, and serositis. The prevalence of SJIA ranges from 0 to 8.6/100 000, with incidence maxima occurring in children ages 1 to 5 years with both men and women equally. **Objective of the study.** To evaluate the clinical, paraclinical and evolutionary features of juvenile idiopathic arthritis, systemic form. **Material and methods.** The bibliographic sources for this study were analyzed using PubMed, Google Scholar, MedScape, Oxford Academic and were released between 2013-2023. **Results.** The typical laboratory findings of granulocyte predominant leukocytosis, elevated acute-phase reactants including thrombocytosis and hyper-ferritinemia, C-reactive protein (CRP) and very high ESR levels. The presence of intermittent, daily, high, spiking fevers (in a quotidian fever pattern), typical evanescent rash, and arthritis are used to make the clinical diagnosis. Additionally, alarmin proteins S100A8/A9 (calprotectin or MRP8/14) and S100A12 (calgranulin C), which are significantly raised in SJIA, are demonstrated to be significantly higher in serum markers of innate immune activation. Macrophage activation syndrome (MAS), a consequence of SJIA, should be immediately suspected in cases of mild increases in AST, ALT, hypoalbuminemia, elevated globulin level, and low-grade D-dimer positive. Imaging tests such as X-rays are rarely useful in the diagnosis of juvenile arthritis; however, an MRI or, on occasion, an ultrasound, can be used to screen for problems or to identify early joint inflammation. Inhibitors of IL-1 and IL-6 have been proven to be quite successful in treating SJIA, as these two factors are important in the disease's pathophysiology. A possible "window of opportunity" in the treatment of children with this rare illness appears to be represented by recent results suggesting that early cytokine blockade may abrogate chronic, destructive, therapy-resistant arthritic phase. **Conclusion.** Systemic Juvenile Idiopathic Arthritis (SJIA) presents several challenges because of its complexity and variability. Specialized care is necessary for patients with erosive polyarthritis, long-term systemic disease, those who are not responding to standard therapies, and those who have remitting-relapsing Macrophage Activation Syndrome (MAS). To effectively manage a health issue, monitor its progression, and address its long-term effects to improve patient outcomes and lower morbidity and mortality, a multidisciplinary approach is essential. **Keywords:** systemic juvenile idiopathic arthritis, macrophage activation syndrome, quotidian fever.