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Clinical case of neutropenia, juvenile idiopathic arthritis in infant

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Abstract

Neutropenia is known to result from decreased production, ineffective granulopoiesis, shift of circulating polymorphonuclear cells (PMN) into the vascular endothelium or tissue pools, or increased peripheral destruction. Juvenile idiopathic arthritis (JIA) is a chronic idiopathic inflammatory disease that predominantly affects the joints. The article presents a clinical case of a patient 13 month old. The condition was evaluated as JIA, and glucocorticosteroid therapy was with high doses of intra venous. There was no exacerbation of the disease.

KEY WORDS: juvenile idiopathic arthritis; neitropenia; inflammation

Background

Neutropenia is a challenging issue for pediatricians. The absolute neutrophil count (ANC) is equal to the product of the white blood cell (WBC) count and the fraction of polymorphonuclear cells (PMNs) and band forms noted on the differential analysis. The absolute neutrophil count (ANC) is equal to the product of the white blood cell (WBC) count and the fraction of polymorphonuclear cells (PMNs) and band forms noted on the differential analysis. The causes of isolated neutropenia can be classified

by mechanism or by etiologic agent. Neutropenia results from four basic mechanisms: decreased production, ineffective granulopoiesis, shift of circulating polymorphonuclear cells (PMNs) to vascular endothelium or tissue pools, or enhanced peripheral destruction. Juvenile idiopathic arthritis (JIA) is a chronic idiopathic inflammatory disorder primarily involving joints. The pathogenesis and etiology of JIA are unclear. As with most autoimmune disorders, interactions among genetic factors, immune mechanisms, and environmental exposures are thought to contribute in most cases.

Most of the genetic predisposition to JIA is determined by the major histocompatibility complex (MHC) loci. Although at least some genetic component is evident in all clinical forms of JIA, the environmental component appears to be stronger for some forms [1,2]. Potential environmental influences that may improve or worsen disease include infection, antibiotic use, breastfeeding, maternal smoking, and vitamin D/sun exposure [3].

In classical manifestation sJIA White blood cell (WBC) counts are almost always elevated, and counts in the 20,000 to 30,000/mm 3 range are not uncommon, Antinuclear antibodies (ANA) and rheumatoid factor (RF) are almost always negative in sJIA.

Clinical case

Here we are presenting the case of the infant with severe neutropenia, anemia and arthritis. Patient is 13 months old. Second child in the family, born full term, weight 3300gr. Mother's pregnancy was complicated by severe gestosis of early pregnancy (NVP); COVID-19; at 12 weeks of gestation, hypercoagulation, with short term use of anticoagulation therapy. At the age of 6 months (10/2021) after vaccination, the patient experienced high fever which lasted for 1 month, later she developed moderate anemia, agranulocytosis. Child was hospitalized in December 2021 in the hematology department for anemia (Hg – 6.8 mg/dl). Numerous investigations were performed, including bone marrow aspirate which showed no abnormalities.

Treatment included antibiotic therapy with ceftriaxone IV, and according to protocol for neutropenia, cefixime p.o was prescribed for following 4 months. From March 2022, the patient expressed pain and swelling in both knees, after short period of time the both ankles were involved. Immunodeficiency was ruled out by the conducted labs. Peripheral blood analysis revealed severe anemia and agranulocytosis, Hg – 7.0 mg/dl, ANC – 0.42, Ferum – 2.47 mcm/l, C-reactive protein (CRP) – 76 mg/l, erythrocyte sedimentation rate (ESR) – 70 mm/hr. Intravenous antibiotic therapy (tazobactam/piperacillin 80mg/kg/d) was initiated, hemotransfusion was performed. During the last

hospitalization there were episodes of fever, severe malaise, swelling, pain, limitation of movement in both knees, both ankles, right hip, elbow, wrist joint, PIP joint of the third finger on the right hand. A mass deficit; – 2SD was observed. Laboratory and instrumental studies were conducted – WBC – 5.5 10.6, RBC – 4.0 – 10 12 /l, HGB – 9.2 g/l (after hemo transfusion), HCT – 29.5, MCV – 60.0, MCH – 18.8; MCHC – 32,2; bands – 2, Seg – 15 (ANC-0.93); ESR – 40mm/hr, CRP – 43.9g/dl. Antinuclear factor 1:2560, homogeneous type fluorescenece in high titer.

The condition was evaluated as sJIA, and glucocorticosteroid therapy with high doses of i.v. was prescribed. Intensity of arthritis decreased in dynamics, inflammatory markers – positive dynamics, fever was no longer observed. The patient is currently 1 year and 5 months on 7.5 mg prednisolone daily. There is no exacerbation of the disease.

Conclussion

As resume we can say, there are two very important questions about this patient: Is neutropenia lasting for more then 5 months the beginning of the Juvenile Idiopathic Arthritis (JIA), and could the maternal COVID-19 infection lead to innate immune dysregulation and as the result an unusual presentation of JIA and neutropenia.

References

1. Dong Y, Mo X, Hu Y, Qi X, Jiang F, Jiang Z, Tong S. Epidemiology of COVID-19 Among Children in China. Pediatrics. 2020;145(6). doi: 10.1542/peds. 2020-0702
2. Ellis JA, Munro JE, Ponsonby AL. Possible environmental determinants of juvenile idiopathic arthritis. Rheumatology (Oxford) 2010; 49(3):411-425. doi: 10.1093/rheumatology/kep383
3. Hinks A, Marion MC, Cobb J. Brief Report: The Genetic Profile of Rheumatoid Factor-Positive Polyarticular Juvenile Idiopathic Arthritis Resembles That of Adult Rheumatoid Arthritis. Arthritis Rheumatol 2018; 70:957
4. Gracia-Ramos AE, Martin-Nares E, Hernández-Molina G. New Onset of Autoimmune Diseases Following COVID-19 Diagnosis. Cells. 2021; 10(12):3592. doi: 10.3390/cells10123592