Pediatric Rheumatology

Poster presentation

Idiopathic hypereosinophilic syndrome (HES) in a 15 year-old girl

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Case report

The hypereosinophilic syndrome (HES) is a group of diseases characterized by persistent and marked blood eosinophilia, with end-organ involvement and no recognized secondary cause.

We present a 15 year-old girl who was admitted to our Department in January 2008 with a four-week history of headache, arthralgias, myalgias, sore troath and angioedema. Laboratory test revealed significant leucocytosis (76 × 10ex9/L with 88% eosinophils), thrombocytosis (758 × 10ex9/L), elevated ESR (82 mm/h) and IgE 348.3 (n.v. < 114 g/L), and hypergamaglobulinemia. Extensive allergologic, immunologic, infectious, and toxicological studies were negative. Bone marrow biopsy showed increased cellularity with increased granulopoiesis predominated by cells of the eosinophilic lineage, with a normal karyotype. Patient was negative for the FIP1L1-PDGFRA fusion kinase and BCR-ABL gene fusion by RT-PCR: Abdomen CT showed diffuse small intestine wall thickness, and cardiac echocardiogram showed thickness of the left ventricular wall and interventricular septum. The biopsy of myocardium and small intestine was planned, but in a mean time patient's condition was worsened. She developed hypoproteinemia (46 g/L), generalised oedema, and diarrhoea. A diagnosis of idiopathic HES was made and methylprednisolone was introduced in the therapy. She had rapid response to methylprednisolone (within 12 hours), with normalisation of the blood counts, protein level and regression of oedema. Methylprednisolone was slowly tapered, and at present, HES is in complete clinical and laboratory remission.

Conclusion

Although HES is extremely rare in childhood, it has to be considered, when a patient is presented with significant leucocytosis and eosinophilia.