

Childhood idiopathic myelofibrosis : a case report and review of literature.

Abstract

Idiopathic myelofibrosis occurs predominantly in older adults. It is very rarely seen in children. We describe a 3-year-old girl with Down's syndrome who presented with recurrent chest infections associated with anaemia and easy bruising. There was mild hepatosplenomegaly. Full blood picture revealed pancytopenia with leucoerythroblastosis with absence of circulating blast cells. Repeated attempts at bone marrow aspiration and trephine biopsy were unsuccessful. A trephine biopsy from the tibia showed depressed myelopoiesis and erythropoiesis, megakaryocytes with atypical morphology and increased bone marrow reticulin fibres, findings compatible with idiopathic myelofibrosis. She was treated symptomatically as she was clinically stable. Review of the English literature online yielded 46 reported cases of childhood idiopathic myelofibrosis with variable outcome from spontaneous remission to an indolent course with shortened survival. 6 cases evolved to another malignancy. 5 cases were associated with Down's syndrome.

Keyword: Idiopathic myelofibrosis; Childhood; Down's syndrome; Pancytopenia; Leucoerythroblastic picture.