

Demographic and clinical characteristics of immune Thrombocytopenia in Sudanese Children

Fathelrahman Elawad

Al-Neelain University, Sudan

Background

The demographic and clinical profile of immune thrombocytopenia is well reported in the literature but was not reported before from Sudan.

Method

A retrospective chart review was performed for all children diagnosed as immune thrombocytopenia (ITP) in three major hospitals over six and half year period. As of 47 patients are identified and their median age was 6.5 years. Males and females were equally affected. A preceding upper respiratory infection was present in one third of patients. Introducing Epistaxis was the commonest highlights like (87.2%), where gastrointestinal bleeding net haematuria and sub-conjunctival draining were the introducing highlights in 36.2%, 19.1% and 4.3% respectively. Ecchymosis and petechial were the most typical clinical signs, seen in 46.8% and 29.8% respectively. Chronic ITP constituted one third of patients. Steroids were the first line of treatment and no death was encountered.

The haematological disorder ITP acquired that's developed secondary to the assembly of auto-antibodies against platelets resulting in isolated thrombocytopenia, as the absence within that causes of thrombocytopenia such as drugs, infections, malignancy, or other autoimmune diseases .

A report on standardization of terminology, definition and outcome of ITP was published recently in Blood by International working party (IWG) [6]. The group found that the term purpura isn't accurate, because many patients don't present with bleeding symptoms and even thrombocytopenia might be discovered incidentally during routine clinic visit. However, the Acronym ITP will continue to be used and will stand for the new proposed name (Immune Thrombocytopenia). The platelet count threshold to establish the diagnosis was set at a new level (100×10^3), instead of the previous limit of (150×10^3). The definitions of the further reports to the phases of the disease as follows: Newly treated ITP (within 3 months from diagnosis), consistent ITP (between 3 to 12 months from diagnosis; this includes patients that don't reach spontaneous remission or don't maintain complete response off therapy), and chronic ITP (lasting for quite 12 months). The presence of bleeding symptoms with severe ITP at presentation that mandate diagnosis or occurrence of latest bleeding introducing new therapies like platelet enhancing agents or increasing the doses of previously used medication for treatment.

The reddish-purple lesions denote Purpura caused by bleeding under the skin. Purpura may be a word of Latin origin meaning

“purple”. As a clinical symptom, purpura was recognized by the traditional Greek and Roman (Hippocrates and Galen) who described it as red “eminence” related to plaque (pestilential fevers). In the 10th century the Muslim philosopher and physician Abu Ali Al-Hussain, Ibn Abdullah Ibrnsina (Avicenna) (980–1037) described a chronic form of purpura that matches the diagnosis of ITP. As the German Poet in 1735 and physician Paul Gottlieb Werlhof (1699–1767) brief description of ITP. He described a disease during a 16 years old girl who had cutaneous and mucosal bleeding and called it “morbus maculosus haemorrhagicus”. After him this condition was called as Werlhof disease. As the name Werlhof disease or condition termed as the (purpura simplex). Many milestones followed during the years including the discovery of platelets and megakaryocytes by the end of the 19th century. In 1916, Paul Kaznelson, who was still a medico in Prague, assumed that the spleen was the location of platelet destruction and convinced his tutor, Professor Doktor Schloffer, to perform the primary splenectomy ever in ITP for a 36- year wife , who had a disease which inserts our definition of chronic ITP. Splenectomy was followed by significant rise in platelets count. As the first immune nature of ITP was first demonstrated by William J Harrington in 1951, who infused normal volunteers with plasma extracted from ITP patients. This resulted in severe drop in platelet counts. In 1951, Evans was ready to identify the plasma factor as antiplatelet antibody. In the same year, Wintrobe was the primary to use steroids for the treatment of ITP. In 1981, Imbach et al, reported the successful use of intravenous immunoglobulin (IVIg) in treating ITP and in 1983, the use of anti-D in ITP was described by Salama et al. The use of rituximab was introduced by the top of 20th century for the treatment of patients with chronic and refractory ITP. Recently the use of thrombopoietin agonists to enhance platelets production showed promising results in severe refractory chronic ITP.

ITP in children typically affects a previously healthy young child who is between two to seven years aged. Males and females are equally affected. However, recent studies reported a higher male/female ratio during infancy with a decreasing trend toward older age. The disease onset is abrupt with bruises and petechial rashes affecting most patients. Epistaxis may occur in about one third of patients and hematuria is uncommon. In about two thirds of patients, the disease onset is preceded by an infection in the previous few days to several weeks. The infection is most often an upper respiratory tract viral infection and the interval between the infection and the ITP onset is in the range of two weeks. Clinical examination reveals a healthy child who only has bruises and petechiae as manifestation of the low platelet count. There should be no organomegaly and no lymphadenopathy. In very rare occasions, the tip of the spleen may be felt. The

diagnosis of ITP in children is essentially one of exclusion. The CBC shows isolated thrombocytopenia with normal WBC and normal Hb levels. The peripheral blood smear shows no evidence of abnormal cells. The diagnosis of ITP in children is essentially one of exclusion. In order to differentiate ITP from other conditions, medical record should include type and severity of bleeding, systemic symptoms, history of respiratory infections, recent live viral vaccine, medications, presence of bone pain, and case history of bleeding disorders. Clinical examination should include observation for body dimorphic features, especially skeletal

anomalies, and the presence or absence of hepatosplenomegaly and/ or lymphadenopathy. The production of antiplatelet autoantibodies was explained by the idea of molecular mimicry.

Conclusion

ITP in Sudanese children has similar features as those reported before; however, gross haematuria and gastrointestinal bleeding were seen more frequent in them.