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Primary Chronic Immune Hrombocytopenic Purpura with Large Spleen- A Rare Presentation

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ABSTRACT

Chronic immune thrombocytopenic purpura (ITP) is defined as immune thrombocytopenic purpura with platelets less than 1 lakh/cu.mm lasting for more than 12 months. 20 % of the children presenting with acute ITP evolves into chronic ITP. Usually, patients with ITP will not be having any hepato/splenomegaly or only tip of the spleen would be palpable in around 5% of the cases. In this case the size of the spleen is bigger than usual presentations of ITP.

Key words: ITP, CBC, Bone marrow

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CASE REPORT

A case of 11 years old female child presented with complaints of bleeding from left nostril. It was associated with abdominal pain, no history of fever, history of recurrent epistaxis was present for 1year [1]. Past history of fever with thrombocytopenia was present before 2 years. On Examination: child is conscious, oriented, afebrile, epistaxis was present [2]. Abdomen - Spleen was palpable 6 cm below right costal margin towards the umbilicus. No other organomegaly Investigations were as follows:

CBC - showed thrombocytopenia with other cell lines being normal.
Peripheral Smear - RBC, WBC - Normal, Platelets - Decreased.
RFT, LFT - Normal.
DCT, HIV, HBsAG, HCV, ANA - Negative.
Bone Marrow Aspiration study - Hyper cellular marrow, Myeloid and Erythroid series - Normal and Megakaryocyte – Increased (Figure 1).

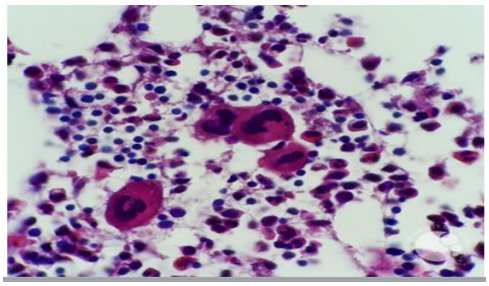

Figure 1: Bone marrow showing hypercellularity with increased megakaryocytes.

USG Abdomen: Spleen measuring 6 cm in size and other organs were normal [3]. Diagnosis of Primary Chronic Immune Trombocytopenic Purpura was made and was treated with Inj. Methyl Prednisolone I.V @ 20 mg/kg/day for 3 days followed by T. Prednisolone @ 2mg/kg/day for two to three weeks then was tapered down. Bleeding tendency improved and platelet count also improved. She was planned for splenectomy [4].

DISCUSSION

Chronic ITP is caused by auto antibody mediated destruction of platelets [5]. The auto antibodies are most often directed against platelet membrane glycoproteins IIb-IIIa or Ib-IX. Large spleen - a variant presentation makes it difficult to diagnose and adds to rule out other secondary causes.

CONCLUSION

Presence of significant splenomegaly is an unusual presentation for Chronic ITP and high index of suspicion is needed to rule out secondary causes of chronic ITP.

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