Case Report

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Intracranial Haemorrhage in Childhood ITP: An Unfortunate Twist of Fate

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Abstract

Immune thrombocytopenic purpura (ITP) is a clinical syndrome in which a decreased number of circulating platelets (thrombocytopenia) manifests as a bleeding tendency, easy bruising (purpura), or extravasation of blood from capillaries into skin and mucous membranes (petechiae). Although most cases of acute ITP, particularly in children, are mild and self-limited, intracranial hemorrhage may occur when the platelet count drops below $10 \times 10^9/L$ ($<10 \times 10^3/\mu L$); this occurs in 0.5-1% of children, and half of these cases are fatal. Intracranial haemorrhage is a rare but life threatening complication of childhood Immune Thrombocytopenic Purpura. Aggressive multi – modality treatment is warranted in such cases.

Keyword: Immune thrombocytopenic purpura (ITP); Intracranial Haemorrhage.

Case Summary

We report a 6 yrs old boy who presented to our centre with complaints of bleeding from nose and mouth 3 days prior to admission –1 to 2 episodes with malena on the day of admission and bluish discolouration of skin with trivial injury. On enquiry there was no history of fever, no significant history of spontaneous bleeding episodes in the past , no history of blood transfusion or family history suggestive of bleeding diathesis. On general examination, patient had pallor, bilateral subconjunctival bleeds, nasal crusting, clot on the lower lip andpetechiae on the hard palate. Multiple ecchymotic patches were present over trunk and bilateral lower limb(3*3 cm). No lymphadenopathy or sternal tenderness was present. On Systemic examination, there was no organomegaly. A Bleeding screen panel revealed platelet count of 7000 per cu. mm with normal PT, INR, PTT with occasional giant platelets on Peripheral Smear examination. Fundus was normal. A single dose of Anti D(70 microgram per kg) was given on the day of admission. Despite this, Platelet counton Day 3 of admission was 8000 per cu. mm. On day 5 of admission, patient had one episode of Generalised tonic clonic convulsion with altered sensorium (Glasgow Coma Score – 7) and

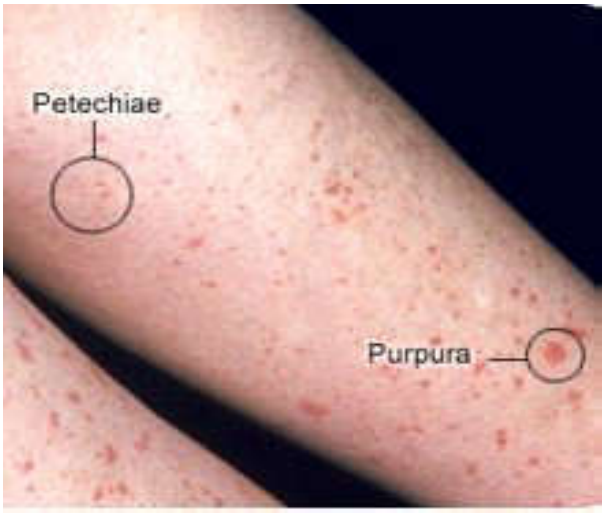

Fig. 1:

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required mechanical ventilation with anticonvulsants. Repeat Fundus examination showed multiple retinal hemorrhages. Urgent CT Brain revealed Intraparenchymal bleed with midline shift. Patient was given Inj Intravenous Immunoglobulin and Pulse Methylprednisolone along with platelet transfusion. Platelet count marginally increased to 47,000. However, before Neurosurgical intervention could be done, patient developed hypernatremia and went into acute renal failure following which he had cardiorespiratory arrest and succumbed.

Discussion

We report this case to emphasize that any child with Immune Thrombocytopenic Purpura who presents with severe thrombocytopenia (platelet count less than 10,000 per cu. mm) and severe bleeding manifestations (beyond petechiae and purpura) with refractoriness to initial therapy requires aggressive multimodality treatment to prevent and manage Intracranial Hemorrhage [1].

Immune thrombocytopenic purpura (ITP) is a clinical syndrome in which a decreased number of circulating platelets (thrombocytopenia) manifests as a bleeding tendency, easy bruising (purpura), or extravasation of blood from capillaries into skin and mucous membranes (petechiae). Intracranial hemorrhage (ICH) is the most devastating complication of immune thrombocytopenic purpura (ITP) in children, and prevention of ICH is the primary goal of ITP treatment. However, the great majority of patients with ITP, even those with very low platelet counts, do not experience severe bleeding and ICH occurs in less than 1 in 100 children with ITP. Potential risk factors include platelet counts below 10 to 20 × 10^9/L, nonsteroidal anti-inflammatory drugs (NSAIDs), head trauma, vasculitis associated with systemic lupus erythematosis (SLE), and cerebral arteriovenous malformations (AVMs) [2] .

Findings Suggestive of Intracranial Hemorrhage Include the Following:

- Headache, blurred vision, somnolence, or loss of consciousness.
- Hypertension and bradycardia, which may be signs of increased intracranial pressure.
- On neurologic examination, any asymmetrical finding of recent onset.
- On fundoscopic examination, blurring of the optic disc margins or retinal hemorrhage [3].

Bone marrow examination is unnecessary in children and adolescents with the typical features of ITP.

Bone marrow examination is not necessary in children who fail IVIg therapy.Bone marrow examination is also not necessary in similar patients prior to initiation of treatment with corticosteroids or before splenectomy. Platelet transfusion is not indicated in all cases of ITP.

Children with no bleeding or mild bleeding (defined as skin manifestations only, such as bruising and petechiae) should be managed with observation alone regardless of platelet count. For pediatric patients requiring treatment, a single dose of IVIg (0.8-1 g/kg) or a short course of corticosteroids should be used as first-line treatment. IVIg leads to more rapid increase in the platelet count [4].

Severe bleeding is distinctly uncommon when the platelet count is >30 × 10^9/L and usually only occurs when the platelet count falls <10 × 10^9/L.

In children who have acute ITP, with a self-limiting period of thrombocytopenia and with no or little bleeding, the UK registry data has shown that ascribing no treatment has not resulted in any immediate increase in ICH. The risks of bleeding remain low and the adverse effects of steroids or immunoglobulins may have a greater risk.However, a small number of patients do suffer from bleeding and require treatment. Not treating relies on avoidance of risks, and an understanding of when intervention is required,such as head injury and degree of bleeding [5].

Conclusion

In cases with Intracranial haemorrhage multimodality treatment should be given. Intravenous Immunoglobulin with Platelet transfusion with Anti D should be given and aggressive management should be done in such cases.

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