Surgery Journal 4 (3): 35-37, 2009
ISSN: 1816-3211

Idiopathic Thrombocytopenic Purpura Presenting with Severe Epistaxis: A Report of 2 Cases

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Abstract: This is a study of two cases of Idiopathic Thrombocytopenic Purpura (ITP) (in a child and an adult) presenting in the Accident and Emergency department of the hospital with severe epistaxis and managed by the Otolaryngologists. The criteria, management and out come of the two cases are discussed.

Key words: Idiopathic thrombocytopenic purpura, presentation, epistaxis, management, disorder

INTRODUCTION

Immune or Idiopathic Thrombocytopenic Purpura (ITP) is an autoimmune disorder characterized by a low platelet count and mucosal bleeding. The thrombocytopenia is a result of both increased platelet destruction and insufficient platelet production. It is aptly described as an autoimmune disorder characterized by persistent thrombocytopenia due to auto-antibody binding to platelet antigen (s) causing their premature destruction by the reticuloendothelial system and in particular the spleen (Woods *et al*., 1984a, b). The estimated incidence is 100 cases per 1 million persons per year and about half of these cases occur in children (Cines, 2002; Prederiksen and Schmidt, 1999; Lilleyman, 1994; George *et al*., 1995, 1996). ITP is a common hematologic disorder manifested by immune-mediated thrombocytopenia (Cines and McMillan, 2005; McMillan and Imbach, 2003). ITP is classified as primary or as secondary to an underlying disorder and as acute (of 6 months or less in duration) or chronic. Adult-onset and childhood-onset thrombocytopenic purpura are strikingly different. Affected children are young (peak age, approximately 5 years) and previously healthy and they typically present with the sudden onset of petechiae or purpura a few days or weeks after an infectious illness.

Boys and girls are equally affected. In >70% of children, the illness resolves within 6 months, irrespective of whether they receive therapy. By contrast, ITP in adults is generally chronic, the onset is insidious and approximately twice as many women as men are affected (Mareaccio, 2000). Thrombocytopenic purpura is characterized by prolonged bleeding time, poor clot retraction, an increase in capillary fragility and a decrease in circulating platelets. Some patients may present with haemmorrhagic bullae on mucous membranes, GI bleeding, menorrhagia, retinal haemorrhages or evidence of intra-cranial haemorrhages with neurologic symptoms. It may be classified as; the essential or primary type and the symptomatic or secondary type (Nachlas, 1995). This study presents two cases (an adult and a child) with ITP presenting with severe epistaxis and focuses on the management of ITP as well as discussing criteria and roles of different forms and types of treatment.

MATERIALS AND METHODS

Case 1: Master S.I, an 8 years old boy who was referred from a private hospital presented at the accident and emergency department of the hospital on 14th of August 2007, with severe epistaxis as evidenced by massive oozing of blood from the nasal cavities. A week earlier, he had presented to the referral hospital with complaints of bleeding on and off for about 1 week which got worse in the past 2 days prior to his admission. He was said to be febrile at presentation and had received six pints of blood at the private hospital. He did not respond to medication with dicynone and vitamin K injections given at the hospital. At the accident and emergency department he presented with severe epistaxis as stated, haemoptysis and difficulty in breathing. There was no history of recent drug in take or vaccination prior to on set of the symptoms. There was no history of prolonged bleeding at circumcision, injection or wound site and no positive family history of prolonged bleeding or similar illness. On examination at presentation, he was pale with

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blood coming out of the mouth and oozing from the nasal cavities. He was afebrile to touch and his breathing was shallow and pulse was weak. There was no palpable peripheral lymphadenopathy or pedal oedema. Chest was clear and heart sounds 1 and 2 only were heard. Abdomen was flat, moved with respiration and no palpable intra abdominal organs. Nose was cleared of blood and clot by suctioning. Attempted cauterization was unsuccessful and the nose was packed with vaseline gauze impregnated with genticin cream.

Results of blood investigations showed a genotype of AS, Haemoglobin of 3.2 gm dL^{-1}, platelet count of <20,000 cu mm^{-1}, White blood count of 6,500 cu mm^{-1}. Human Immunedeficiency Virus (HIV) screening test was negative. Sickling test was mildly positive. Urine analysis was normal. He was grouped and cross matched and given fresh whole blood immediately. Oozing continued from the nose despite packing and transfusion of fresh blood. A working diagnosis of Idiopathic Thrombocytopenic Purpura was made. Repeat haemoglobin after transfusion was 6.8 gm dL^{-1}. He was given vitamin K injection and vitamin C. He made slight improvement with his breathing stabilizing.

The next day blebs appeared in the mucous membrane in the mouth and the oozing increased. He developed haematuria. Additional two pints of fresh whole blood were transfused as there were no packs of fresh platelets. Repetition of tests showed that the platelet count had risen to 50,000 cu mm^{-1}, while the haemoglobin rose to 7.7 g dL^{-1}. Bone marrow biopsy was done, which was inconclusive. The patient was placed on Prednisolone 15 mg stat and 5 mg daily for 5 days. On the 3rd day of treatment with this regimen the blebs disappeared, the oozing reduced and the haematuria stopped. A repeat of blood tests showed Haemoglobin of 8.4 g dL^{-1}, platelet count of 120,000 cu mm^{-1} and white blood cell count of 5,900 cu mm^{-1} was recorded. He was discharged on the 5th day and was followed up in the out-patient ENT clinic. One month after discharge his blood tests showed Haemoglobin of 11.5 g dL^{-1}, Platelet count of 205,000 cu mm^{-1} and white cell blood count of 5,500 cu mm^{-1}.

Case 2: Miss O.O, a 25 years old female secretary/typist presented in the accident and emergency department with severe epistaxis and haemorrhagic blebs in the buccal cavity on the 29th of June 2008. This was preceded by fever, catarrh and cold and generalized weakness. Two weeks before, she had taken chloroquine and fesolate tablets. She noticed that she was loosing weight and having bouts of dizziness. There was no history of excessive or prolonged bleeding from previous surgery or trauma and no menorrhagia. She does not take alcohol or tobacco in any form and she is not on any regular drug. There is no family history of prolonged or excessive bleeding from wound site or trauma of any sort. She is not a known peptic ulcer patient.

Examination showed, a young anxious lady with blood oozing out from both nasal cavities and blebs in the bucal cavity. Her vital signs were maintained. She was warm, not pale and anicteric. Abdomen was full and moved with respiration. There were no palpable peripheral lymph nodes, pedal oedema or enlarged abdominal organs. Chest was clear with good air entry and heart sounds 1 and 2 only heard. Blood tests showed haemoglobin of 13.5 g dL^{-1}, white blood cell count of 8,600 cu mm^{-1}, platelet count of <10,000 cu mm^{-1}, genotype was AA. HIV screening test was negative and blood film was normal. Urine analysis was negative. A diagnosis of Idiopathic Thrombocytopenic Purpura was made. The nose was cleared of clot and packed. She was placed on Prednisolone 15 mg stat and 10 mg daily for 5 days on the 2nd day of admission. The blebs disappeared on the third day and nasal bleeding stopped completely on the 5th day of admission. Repeat platelet count after 1 week was 176,000 cu mm^{-1}. She was discharged after 1 week and followed up in the clinic. The platelet count after 1 month was 195,000 cu mm^{-1}.

RESULTS AND DISCUSSION

Thrombocytopenic purpura is characterized by multiple hemorrhages into the skin and mucous membranes. One of the earliest indications of hemorrhage may be epistaxis and the otolaryngologist may be the first to suspect the diagnosis and be instrumental in the institution of early treatment (Nachlas, 1995). The diagnosis remains one of exclusion, after other thrombocytopenic disorders are ruled out based on history, physical examination and laboratory evaluation. The goal of treatment is to raise the platelet count into a haemostatically safe range (Cines and McMillan, 2005). Childhood ITP differs from adult ITP in pathogenesis, differential diagnosis and management. Patients typically present with petechiae or purpura that develop over several days, accompanied by platelet counts of 10-20,000 cu mm^{-1}, although onset can be insidious. Severe cutaneous bleeding, epistaxis (like in the two cases presented), gingival bleeding, haematuria or menorrhagia may develop at platelet counts below 10,000 cu mm^{-1} (Nachlas, 1995). In adults, the course is commonly chronic, but most patients never experience serious bleeding even with severe thrombocytopenia. The frequency of death from bleeding is low, <1%. The major cause of fatal bleeding in patients with ITP is

intracranial haemorrhage (Prederiksen and Schmidt, 1999; Mareaccio, 2000; George, 2006). All current treatments are designed to diminish the increased platelet destruction, either by immunosupression or splenectomy. However, treatment is necessary for patients with severe and symptomatic thrombocytopenia. Adults presenting with a platelet count <30,000 cu mm^{-1} are usually treated with oral glucocorticoids oral prednisone 1 mg kg life threatening beeding requires immediate administration of platelet transfusions (George, 2009). Ideally, treatments for ITP should be effective, safe, tolerable and inexpensive. Initial corticosteroid treatment should be limited in duration to avoid intolerable side effects. The goal of treatment is to achieve a safe platelet count to prevent serious bleeding. A normal platelet count may provide confidence that ITP has resolved, but is not the primary goal. Prednisone (or prednisolone) at a daily dose of 1 mg kg^{-1} is commonly used. Other regimens, such as a high dose of dexamethasone (40 mg) daily for 4 days may be more effective (Cheng *et al*., 2003). Spontaneous remission occurs in >80% of cases in children but is uncommon in adults. In acute ITP (children) the sex distribution is equal while in chronic ITP (adult) the sex ratio is 2:1 in favour of females. Peak prevalence occurs in adults aged 20-50 years, while peak prevalence occurs in children aged 2-4 years. Approximately, 40% of all patients are <10 years (Silvermann, 2006).

CONCLUSION

Despite major advances in the understanding of the molecular basis of many blood disorders, the diagnosis of ITP remains one of exclusion. There are currently no clinical or laboratory parameters that are able to establish the diagnosis of ITP with accuracy. The otolaryngologists may be the first port of call as in the two cases here presented; hence they should keep an open mind in the diagnosis and management of cases of epistaxis. Concerted effort should always be made to establish a possible cause of any case of epistaxis so that appropriate treatment is instituted rather than more control of bleeding.

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