Abstract:
Idiopathic thrombocytopenic purpura (ITP) also called as Immune thrombocytopenic purpura is an acquired autoimmune disorder in which auto-antibodies are made against platelets leading to increased platelet destruction. Immune thrombocytopenia, increased splenic sequestration, infections such as dengue fever, decreased bone marrow production and adverse drug reactions are the major causes of ITP. Patients with ITP often present with clinical manifestations of bruising and bleeding. Albeit these manifestations are detected most frequently on the skin; on the lower extremities, the oral cavity also may exhibit signs of petechiae, purpura and bleeding. Haemorrhagic bullae of mucous membranes in the oral cavity can indicate the existence of severe thrombocytopenia. We report two female cases presented with oral features of petechial haemorrhage, pigmented brownish black macules with bleeding, indicative of a bleeding disorders. Laboratory test results revealed the idiopathic thrombocytopenia which necessitates the importance of diagnosis of oral manifestations of thrombocytopenia.

KEYWORDS: Idiopathic thrombocytopenic purpura, Immune thrombocytopenic purpura, petechiae, thrombocytopenia, Bleeding disorders.

Introduction

Platelets (thrombocytes) derived from megakaryocytes have an elemental function in the blood clotting system and contribute in the initial
stage of the haemostatic process through adhering to the collagen and aggregating to form the clot \(^1\) thus, disorders accompanied by decreased platelet number or platelet dysfunction may give rise to pathologic bleeding states.\(^2\) Platelet count below 50,000 cu mm is referred as thrombocytopenia.\(^3\) Normal range is between 1,50000 to 4,50000 cu mm.\(^2,3\) Thrombocytopenia or decrease platelet function can take place in the course of a variety of mechanisms, including autoimmune destruction, infections such as dengue fever, spleen sequestration, tumor cell infiltration of bone marrow, and many adverse drug reactions.\(^4\) Idiopathic thrombocytopenic purpura (ITP) also known as immune thrombocytopenic purpura is an acquired autoimmune disease in which there is accelerated platelet destruction due to autoantibodies (7S IgG) made against platelets.\(^5\) As the disorder is of immunological nature, in which the host's platelets are destroyed by host's immune system so the many authors recommend the term “idiopathic” be replaced by “autoimmune”.\(^6\) It occurs in two clinical types; acute self-limiting form which affects exclusively children with the history of severe viral infection and chronic form seen mostly in adults. Acute ITP is common in both sexes and has a peak incidence of 3-5 years.\(^1,4,5\) Chronic form is commonly seen in 20-40 years adults with female predilection (3:1).\(^7\) Evidences available suggest spontaneous remission is seen in only 5% of adults with chronic ITP.\(^8\) Spontaneous Gingival bleeding or due to minor traumatic event like tooth brushing, hard food biting and flossing, is mostly the first sign of thrombocytopenia.\(^4\) The soft palate and buccal mucosa, may display ecchymoses and petechial haemorrhages. In cases with very low platelet counts deep red to black haemorrhagic bullae may occur.\(^9\)

This paper presents two rare cases of idiopathic thrombocytopenic purpura in females with different intraoral presentations.

**CASE REPORT:**

**Case 1:**

A 40 year old female reported to the Dept. of oral medicine and radiology with a complaint of brownish red pigmentations in her mouth and all over the body. She noticed haemorrhagic petechiae and red brown spots in mouth 2 weeks back. She also noticed reddish pigmented spots on back, arms and both legs and bleeding through mouth while gargling and brushing. Physical examination revealed a well-nourished, well-developed, cooperative woman in no acute distress. Her vital signs were stable.

On extra oral examination multiple small reddish brown haemorrhagic macules of approximately 1-2 cm in diameter seen on both arms, both legs and slightly larger in size on back, [Figure 1, 2, 3]. Bluish discolouration of sclera also noted, [Figure 4]. Intraoral examination revealed single reddish haemorrhagic papule of approximately 1x1.5 cm in size noticed on lower labial mucosa, [Figure 5] and also same presentation which is of smaller in size noticed on left buccal mucosa [Figure 6], soft palate, left lateral border of tongue, [Figure 7] and gingiva, [Figure 8]. Small tiny multiple haemorrhagic spots also seen on ventral surface of tongue and floor of mouth, [Figure 9]. In the differential diagnosis acute leukemia and idiopathic thrombocytopenic purpura (ITP) were considered in light of the petechial and ecchymotic lesions, suggesting a hemorrhagic diathesis.
Laboratory results revealed that haemoglobin, haematocrit, and white blood cell counts, as well as prothrombin and partial thromboplastin times, were all within reference range. The platelet count, however, was drastically low at 30000 cu mm (reference range, 1,50000–4,50000 cu mm). Peripheral smear showed normal size platelets. Hence on the basis of all investigatory findings patient was diagnosed with ITP and sent to hematologist for further examination. In treatment she received 1 U of platelets, intravenous immunoglobulin G, and prednisone starting at 80 mg/d. Patient was followed up after 2 weeks her platelet count was raised to upto 135,000 mm$^3$.
**Case 2:**

Another 65 year old female reported to the Dept. of oral medicine and radiology with a complaint of bleeding from mouth and brownish pigmentation on all over the body. She noticed bleeding from oral mucosa and tongue and also noticed haemorrhagic patches on tongue and floor of the mouth. She also noticed brownish pigmented patches of varying sizes on face, arm and neck.

On extra-oral examination single large brownish black pigmented macule of approximately 2x3 cm in size and also few small sized brownish black macules noted on middle third region of right side of face, [Figure 10]. These types of large sized macules are also noted on clavicular region on neck and forearms, [Figure 11, 12]. On intraoral examination large reddish haemorrhagic papule of approximately 2x2.5 cm in size noticed on right lateral border of tongue and also small sized papules noticed on right and left lateral border of tongue and floor of mouth, [Figure 13]. On palpation these papules were easy to bleed.

Differential diagnosis includes metastatic melanoma because of the shiny black appearance of the papules. Acute leukaemia, Actinomycosis, histoplasmosis, aspergillosis, and cryptococcosis were included in the differential diagnosis because oral granulomatous papules also occur in these diseases.

In laboratory results the platelet count was significantly low at 45000 cu mm (reference range, 150000 – 450000 cu mm). Normal size platelets showed in peripheral smear. Hence final diagnosis of Idiopathic thrombocytopenic purpura was concluded and referred to haematologist.

In treatment Corticosteroids is been prescribed; Prednisone 80mg/day. 1 U of platelets was given and also intravenous immunoglobulin G. Patient was followed up for 1 year within 2 weeks her platelet count was raised to 1, 40000 mm³ and subsequently oral mucosal lesions are also been subsided.
Discussion:

ITP is diagnosed by the exclusion of other diseases associated with low platelet count. Diagnosis is best achieved by proper history, physical examination, complete blood count and peripheral smear examination. About 30% to 40% of adults having ITP have no symptoms. Clinical manifestation of ITP shows great disparity. Frequent muco-cutaneous lesions seen in ITP are Petechiae, purpura, easy bruising and hematoma. Rare manifestations are hematuria, GI bleeding and intracranial haemorrhage although intracranial haemorrhage is most common cause of death. Petechiae and ecchymoses are tends to develop on skin. Purpura likely to be form primarily in areas of increased venous pressure, such as extremities like legs. Unprompted gingival bleeding occurs on decreased platelet count less than 20,000 cu mm. ITP can exist in a subclinical form, and severe haemorrhage due to lacerations or minor medical and dental surgical procedures can be its first clinical manifestation. Laboratory results are generally suitable indicators of ITP. Haemoglobin count is typically normal unless significant haemorrhage associated with thrombocytopenia has resulted in anaemia. The WBC count usually is in normal reference range. Other findings are prolonged bleeding time, poor clot retraction;
normal clotting time and positive tourniquet test are other findings. Peripheral smear shows reduced platelet count and is crucial to exclude thrombotic thrombocytopenic purpura and acute leukemia. Early treatment of choice for the patient having very low platelet counts value like 10,000 to 20,000 cu mm leading to intense bleeding is with Intravenous Immunoglobulin (IV-Ig) alone or in combination with Intravenous methyl prednisolone. In conditions of non-emergency initial treatment can be prednisolone 1-2 mg/kg/day). For patients intolerant to corticosteroids, intravenous IV Ig anti-D can be given. The newer therapeutic modalities with varied mechanisms of action, for example rituximab, anti-D, and thrombopoietin like agents can be administered in treatment of ITP.

References:


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