

Oral Manifestations of Immune Thrombocytopenic Purpura: A Diagnosis of Exclusion

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Abstract: Platelets or thrombocytes play an indispensable role in haemostasis to limit excess blood loss due to vascular damage. Platelet dysfunction either qualitative or quantitative, therefore, leads to bleeding disorders. Thrombocytopenia, characterised by decrease in platelet count can occur in different haematological disorders. The distinctive feature of immune thrombocytopenic purpura (ITP) is autoantibody induced platelet destruction leading to low platelet count. In this case clinical and exhaustive investigations of a patient with ITP, in the absence of an obvious systemic disease, are presented. This case spotlights the importance of thoroughly evaluating oral manifestations of any systemic disorder, which may serve to be the first clue towards diagnosis.

Keywords: Platelets; Thrombocytopenia; Immune; Autoantibody.

I. Introduction

Oral cavity is contemplated as mirror to the body because several important diseases are manifested orally. Majority of the oral signs and symptoms are atypical, but should be acknowledged for the likelihood of a contemporaneous underlying systemic disorder. The oral manifestations of haematological diseases may be the initial clinical sign that serves to inform the clinician for a detailed assessment (1). Primary immune thrombocytopenia or idiopathic thrombocytopenic purpura or Immune Thrombocytopenic Purpura (ITP) is an autoimmune disorder, typified by antiplatelet antibodies and immune mediated platelet destruction with a consequent decrease in peripheral platelet count (2). The underlying mechanism of thrombocytopenia in ITP is believed to be immune mediated; however, recent concepts highlight the complex mechanisms with interplay of T cell mediated cytotoxicity and impaired platelet production (2, 3). ITP is classified as acute (<6 months) or chronic (>6 months) based on the duration of its presentation. Acute form is generally seen in children while adults tend to have the chronic form.

ITP is diagnosis of exclusion. Before diagnosing ITP, causes of thrombocytopenia such as thrombotic thrombocytopenic purpura, disseminated intravascular coagulation, bone marrow depression, myelodysplastic syndrome, haemolytic-uremic syndrome, any malignancy etc. need to be ruled out (4). We present a case in which oral manifestations served as initial clue for diagnosing ITP.

II. Case Description

A female aged 67, reported with the main complaint as sudden onset of intraoral bleeding since 2 days and history of breathlessness and fatigue since 2 months. The medical history revealed absence of any systemic disorder. Furthermore, patient reported abstinence from any medication in last 4 months and no trauma history was present. Intra-oral examination revealed multiple large hematoma and ecchymotic patches over labial gingiva, buccal mucosa, dorsum and ventrum of tongue, with areas of clotted blood and active oozing of blood from gingiva (Fig 1). The periodontal health was compromised. The extra-oral physical examination revealed, multiple petechiae and discoloration over the extremities, patient being unaware of it. With a provisional diagnosis of anaemia with thrombocytopenia, the patient was referred to the haematologist for investigations. After complete haematological tests, platelet count was found as 17,000/cmm and haemoglobin as 5gm%. The other hematologic parameters read as RBC: 2.5 million/cmm, WBC: 6600/cmm; bleeding time (Dukes method): 10min; clotting time (Lee & White method): 8min; prothrombin time: 8sec; ESR: 70mm/hr. Bone marrow aspirate was non-contributory.

The peripheral blood smear showed mild hypochromia, mild anisocytosis, and degenerated platelets. Immunologic tests HBsAG, Anti-HBs, Anti-HIV were negative. There was no evidence of lymphadenopathy, hepatomegaly or splenomegaly. Anti-nuclear antibody test (ANA) was advised considering the acute onset at

this age and relation of ANA with the chronicity of the lesion. The optical density (OD) ratio of 2.24 was found with ANA ELISA test which is interpreted as positive for IgG ANA. The patient received platelet concentrate (6 units), whole blood transfusion, prednisone 10mg BD and general care. At 20 days follow up, the petechiae decreased in size and ultimately disappeared after 45 days (**Fig 2**). The patient was asymptomatic with complete resolution of intraoral hematomas, bleeding and petechiae on the skin. Based on the investigations, absence of any other obvious cause of thrombocytopenia and patient's response to the treatment a final diagnosis of ITP was given.

III. Discussion

The first sign of thrombocytopenia, most often is traumatic or spontaneous bleeding from gingiva. Oral mucosa, notably buccal mucosa and soft palate usually demonstrate petechial haemorrhage and ecchymosis. ITP, a haematological disorder characterised primarily by immune mediated platelet destruction with mucosal and skin bleeding has an additional component of decreased platelet production unable to stride with platelet destruction. It presents most commonly in females during first to third decade of life, however, there is no definite age and sex predilection (**5**). Intraorally, ITP manifests as petechiae, purpura, ecchymosis, haematomas that develop over several days and are seen more often in trauma prone areas like buccal mucosa, lateral borders of the tongue, and junction between the soft and hard palate. Spontaneous mucocutaneous and gingival haemorrhage can also occur (**6**). In the present case the lesions were seen on gingiva, buccal mucosa, ventrum and lateral border of tongue.

The diagnostic hallmark of ITP is thrombocytopenia without any alternative cause. The severity of the disease is estimated based on the platelet count. Severe thrombocytopenia ($5000/\mu\text{l}$) may result in intracranial bleeding or haemorrhage. Marked cutaneous or mucosal bleeding, epistaxis, haematuria generally develops at platelet counts under $10000/\mu\text{L}$. Individuals with platelet count above $30,000/\mu\text{l}$ may or may not complain of easy bruising (**7**). On rare occasions, symptoms of bleeding are seen incommensurate with the platelet count, secondary to platelet dysfunction (**8**). Since diagnosis of ITP is by exclusion, a comprehensive history taking is important to exclude other causes of platelet dysfunction. Management depends on the severity of bleeding. Attaining haemostatic platelet count with minimum drug toxicity should be considered. In the present case platelet and blood transfusion along with steroids resulted in complete resolution of the lesions.

IV. Figures

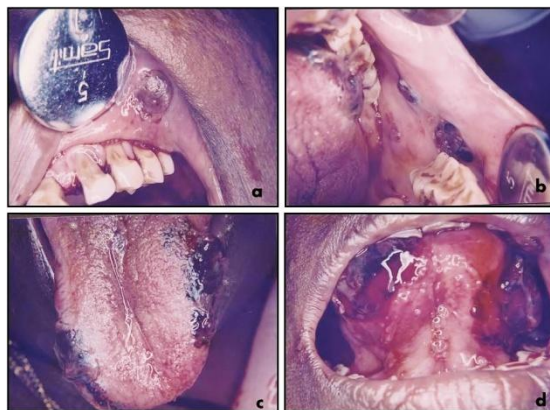


Figure 1: The pre-treatment clinical appearance. Hematoma with areas of bleeding on labial gingiva (1a), buccal mucosa (1b), lateral border of the tongue (1c) and areas of ecchymosis on the ventral surface of the tongue (1d)

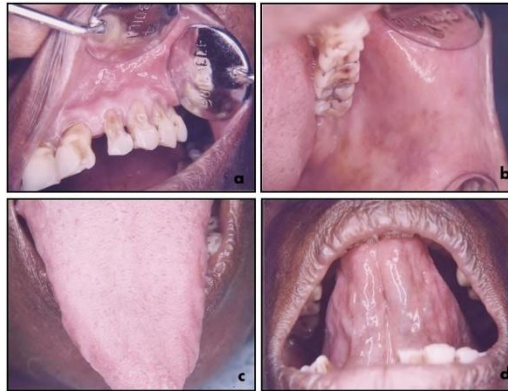


Figure 2:The post-treatment clinical appearance showing complete resolution of the lesions.

V. Conclusion

The symptoms of haematological disorders may be encountered initially in the oral cavity. Therefore the dental surgeon should carefully take note of history and intraoral as well as extraoral manifestations of these disorders. Planning medical or dental treatment is of utmost importance in patients with low platelet count ($<30,000/\mu\text{l}$). Appropriate consultation with the haematologist to determine the severity of the disease is, therefore, essential.

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