Idiopathic Thrombocytopenic Purpura With Black Oral Mucosal Lesions

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Idiopathic thrombocytopenic purpura (ITP) is an acquired autoimmune disorder in which autoantibodies are made against platelets, causing accelerated platelet destruction. History and physical examination are most often normal except for petechiae, commonly seen in the lower extremities. Hemorrhagic bullae of mucous membranes can indicate the presence of severe thrombocytopenia. We report a case of ITP in a 33-year-old man who presented with insidious onset of black oral mucosal lesions.


Case Report

A 33-year-old man presented with a 2-week history of “black spots” in his mouth. The first lesion appeared on the left buccal mucosa, with 4 more black papules appearing on the right buccal mucosa in the next several days. He felt congested and noticed blood-tinged mucus when blowing his nose. He was seen at an urgent care center and was given a 10-day course of amoxicillin for what had been diagnosed as sinusitis. The patient then noticed a bruise on his right arm and another on his left leg. After noting tiny red spots on his legs and feet, he notified a family physician, who gave him methylprednisolone. When this treatment did not alleviate his eruption, he was referred to a dermatologist.

Physical examination revealed a well-nourished, well-developed, cooperative man in no acute distress. His vital signs were stable, and he had a normal temperature. An irregular 8-mm black papule was seen on the left buccal mucosa, and 4 smooth shiny black papules were observed on the right buccal mucosa (Figures 1 and 2). There were 2 ecchymoses, approximately 3 to 4 cm in diameter—one on the right arm and one on the left leg. Multiple red hemorrhagic nonblanching macules 1 to 2 mm in diameter were present on the anteromedial lower legs and dorsa of his feet and were barely visible. The patient’s spleen was palpable just below the left central margin with inspiration. No hepatomegaly or lymphadenopathy was detected.

Differential diagnosis included metastatic melanoma because of the shiny black appearance of the papules. Acute leukemia and idiopathic thrombocytopenic purpura (ITP) were considered in light of the petechial and ecchymotic lesions, suggesting a hemorrhagic diathesis. Actinomycosis, histoplasmosis, aspergillosis, and cryptococcosis were included in the differential diagnosis because oral granulomatous papules also occur in these diseases. However, considering the patient’s general appearance and lack of fever or malaise, these diagnoses were considered less likely causes of the black papules.

Laboratory results revealed that hemoglobin, hematocrit, 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 3000/μL (reference range, 150–450 × 10³/μL). Peripheral smear showed normal size platelets. The patient was diagnosed with ITP and sent to the emergency room where his internist and a hematologist examined him. In view of the acute onset of the lesions and the patient’s age, a bone marrow biopsy was considered unnecessary and treatment was initiated. He received 1 U of platelets, intravenous immunoglobulin G, and prednisone starting at 80 mg/d. After 2 weeks, his platelet count was 130,000/μL.

Comment

ITP, also known as immune thrombocytopenic purpura, is an acquired disease in which autoantibodies are made against platelets, causing accelerated platelet destruction. ITP is classified as primary or...
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secondary to an underlying disease and may be acute or chronic. ITP occurs most often between the ages of 18 and 40 years and is twice as common in women than in men. It occurs in approximately 38 per million adults per year.

Approximately 30% to 40% of adults with ITP have no symptoms. This form of the disease has an insidious onset and rarely results in spontaneous resolution. Patients with ITP usually present with platelet counts greater than 50,000/µL. Individuals with platelet counts between 30,000 and 50,000/µL might experience excessive bruising from minimal trauma. When platelet counts are between 10,000 and 30,000/µL, petechiae or ecchymoses suddenly develop. Patients with platelets below 10,000/µL are at risk of internal hemorrhage.

The majority of adults present with a long-standing history of petechiae and purpura, and many adults are diagnosed incidentally as a result of routine platelet counting. Although the remainder of the history and physical examination are normal, the symptoms and signs of bleeding are not. Occurring most commonly in dependent regions, petechiae are not palpable and they are numerous in the mucous membranes where hemorrhagic bullae can occur when severe thrombocytopenia is present. Signs and symptoms are predictable from the commonly recognized pattern of bleeding associated with congenital platelet function disorders. Purpura, menorrhagia, epistaxis, and gingival bleeding are common; gastrointestinal bleeding and hematuria are less common. Hemorrhagic bullae of mucous membranes can occur with severe thrombocytopenia.

Laboratory findings usually are suitable indicators of ITP. Isolated thrombocytopenia is the primary abnormality. Hemoglobin concentration usually is normal unless substantial hemorrhage associated with thrombocytopenia has resulted in anemia. The white blood cell count typically is within reference range.

Other conditions such as infection with human immunodeficiency virus or hepatitis C, chronic liver disease with hypersplenism, myelodysplastic syndromes, systemic lupus erythematosus, and chronic diffuse intravascular coagulation can mimic ITP. Excluding other causes of thrombocytopenia is the most efficient way of diagnosing ITP. The absence of systemic symptoms, as well as the duration of the bleeding signs, helps to rule out secondary forms and other diagnoses. The diagnosis of ITP is mainly based on history, physical examination, and complete blood cell count, with examination of the peripheral smear. Bone marrow aspiration is recommended to establish a diagnosis in patients older than 60 years and to rule out myelodysplastic syndromes.

Treatment of ITP should be individualized. Glucocorticoid therapy (ie, prednisone 1–2 mg/kg per day) is the appropriate initial therapy for moderate to severe thrombocytopenia and symptomatic purpura. Intravenous immunoglobulin G therapy (2 g/kg given over 2–5 days) is indicated for patients with platelet counts less than 30,000/µL and patients with severe life-threatening bleeding. Platelet transfusions and hospitalization usually are reserved for patients with platelet counts less than 20,000/µL. Splenectomy is considered when platelet counts remain below 30,000/µL after 4 to 6 weeks of medical treatment. Anti-D immunoglobulin therapy currently is used only for Rh-positive patients with severe thrombocytopenia unresponsive to oral agents but is being studied for initial therapy. It is given as a single dose (75 μg/kg) and may be repeated based on platelet response. Most patients with mild to moderate asymptomatic thrombocytopenia (those who can...
maintain platelet counts >30,000/µL) can be safely followed with no treatment.2,9

REFERENCES