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ORAL MANIFESTATION OF BLOOD DISEASES (LITERATURE DATA)

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Abstract

The aim of this paper is to review orofacial manifestation of hematologic diseases (anemia, leukemia and some hemorrhagic syndromes). It is very important that the dental profession recognize these early signs, for many of these conditions may be cured if recognized in their early stage and operative procedures should not be carried out in cases in which a disease of serious prognosis is present. More importance, hematologic disease for dentists are: iron deficiency anemia; megaloblastic anemia; Plummer-Vinson syndrome; agranulocytosis; leukemia; coagulation disorders of homeostasis. Oral signs and symptoms may be the first clinical evidence of underlying blood disorders, and the dental health practitioner must pay careful attention to possible indicators of occult disease. Diseases of blood frequently affect soft and the hard tissues of mouth with different characteristics.

Key words: hemostatic disorders; anemia; agranulocytosis; leucosis;

Introduction

Red and white blood cells are the cornerstones of systemic health, and disorders of these cells may have profound effects on all organ system In the periodontium and oral mucosa, erythrocytes carry oxygen and nutritients to the tissues. Platelets are critical for hemostasis and for production of critical immunoinflammatory mediators. White blood cells protect periodontium and oral mucosa from bacterial, viral and fungal pathogens. These cells exist in a exquisite homeostatic balance. Disorders that disturb this balance may manifest within the periodontium or on other mucosal surface.

The various blood diseases present polymorphic clinical expressions, one of which is the relatively constant involvement of oral structures. The dentist is often consulted by the patients suffering from one of the hematologic disorders who, unaware of this condition, only seek relief of his harassing physical discomfort, Lichuman (1), Meyer (2), Provin (3).

Oral manifestation of many diseases of the blood is clinically similar to those lesions, which occur in the oral cavity, as a result of some local phenomenon, usually irritation or infection. For this reason a specific diagnosis of blood dyscrasia is difficult, is not impossible, to establish on the basis of the oral findings alone.

Diseases red blood cells (anemia) with oral manifestation

Anemia is defined as an abnormal reduction in the number of circulating red blood cells, the quantity of hemoglobin and the volume of packed red cells in a given unit of blood.

A number of different types of anemia may exhibit oral manifestations, but more frequently with oral manifestation are: iron deficiency anemia; pernicious anemia, Plumer-Vinson syndrome and so on.

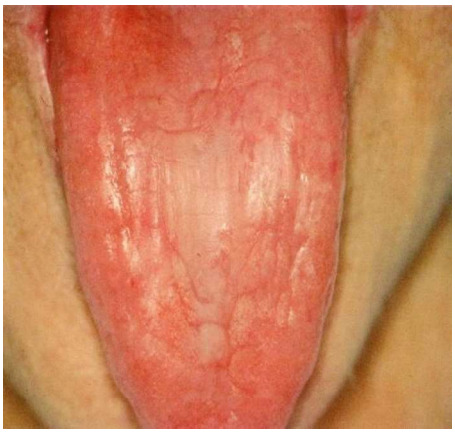
1.1. Hypochromic anemia (iron deficiency anemia)

Iron deficiency anemia is the most common hematological disorders, Mc Farlane (4). It can be suspected from a low MCV and the presence of hypochromic, microlytic red cells. Lesion on the oral cavity can be noticed soon after signs of anemia are noticed and the disease diagnosed, at the same time when disease is diagnosed. However, oral

lesion could be the primary clinical manifestation of the disorder. Accordingly, it can be said that the role of dentists might be very important concerning diagnostic, prognostic and therapeutic issues.

Iron deficiency anemia may manifest in the orofacial region as an atrophic glossitis (fig 1), mucosal pallor and angular cheilitis (fig 2). Atrophic glossitis "flattening of the tongue papillae", resulting in a smooth and erythematous tongue may mimic migratory glossitis.

Migratory glossitis, also known as geographic tongue, is a condition of unknown etiology that affects 12% of population, Long (5). Angular stomatitis, however, is often associated with fungal infection (Candida albicans), lip sucking and dehydration, Zegarelli (6).



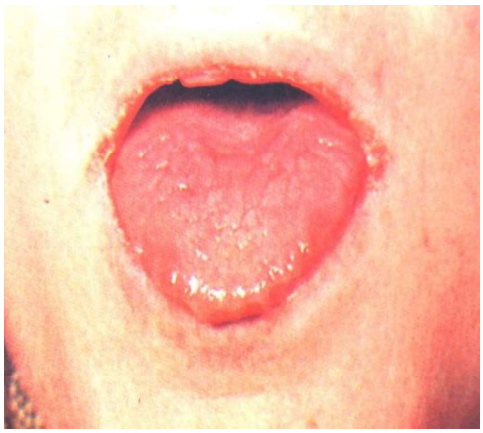


Fig.1 fig 2

1.2. Pernicious anemia (megaloblastic anemia)

Pernicious anemia is one of the most important of all forms of anemia. It is chronic in nature and represents a very typical pattern of clinical manifestation. It is disease of adults, seldom occurring before the age of 30.

This may be caused by a vitamin B12 deficiency (commonly from pernicious anemia, surgical resection of the ileum or small intestinal diverticulitis) or by a folic acid deficiency.

A typical triad of symptoms often has been described in pernicious anemia: generalized weakness; sore, painful, or burning tongue and numbness or tingling of the extremities. There are numerous other clinical manifestations of the disease in addition to the triad listed above. These include fatigue ability, difficulty in walking and coordination, loss of appetite and loss of weight, nausea and vomiting, diarrhea, abdominal pain and generalized characteristics like lemon-yellow pallor of the skin.

The oral manifestations of pernicious anemia may be extremely severe and quite discomforting to the patient so that dental consultation frequently may be sought before medical advice. The most common complaint is a painful, burning sensation of the tongue which itself, upon clinical examination appears beefy red in color, sometimes with small focal areas of ulceration. This glossitis with the accompanying glossodynia and glossopyrosis, is most prevalent in the early stage of the disease. The lingual signs and symptoms occasionally undergo spontaneous remission but almost inevitably recur. While the pain and burning sensation usually are confined to the tongue, they may extend to include much of the mucosa of the entire oral cavity. Ultimately, the papillae of the tongue undergo atrophy, with the result that the tongue appears very smooth and shiny or "bald" fig 3 and 4.The term Hunters glossitis or Moeller's glossitis, has been used to describe the tongue in this stage of the disease as well as the term "bald tongue". In any stage of alteration of the normal lingual mucosa, the patients may complain of an altered taste sensation. Faccini (7), Field (8)

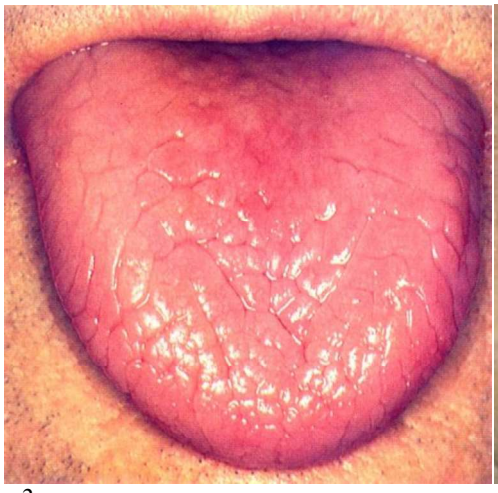


Fig 3

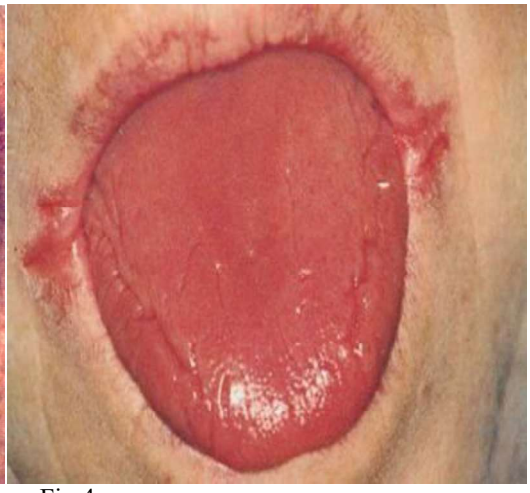


Fig 4

1.3. Plumer-Vinson syndrome (Patterson-Brown-Kelly syndrome)

The Plumer-Vinson syndrome is present with symptom complex caused by iron deficiency, Hoffman (9). However, the Plumer-Vinson syndrome has been found to occur mainly in women, particularly in the third to the fifth decade. These patients have a typical lemon-yellow pallor of the skin and complain of weakness and easy fatigability. Gastrointestinal disturbance such as nausea, vomiting and diarrhea are relatively common. Achlorhydria is almost invariably found. Koilonychias or spoon shaped fingernails, often with longitudinal ridges, is also commonly seen in these patients.

The oral manifestation is prominent in patients with Plumer-Vinson syndrome. There is frequently cracking or fissuring at the corners of the mouth suggestive of the angular cheilosis or ariboflavinosis. In addition, the tongue is generally red, smooth and painful. The atrophic glossitis is similar to that seen in pernicious anemia. Ultimately all the oral mucosa membranes become pale and atrophic. Difficulty in swallowing or dysphagia, is also a common complaint, Hoffman (9)

Disease involving white blood cells with oral manifestation

2.1. Agranulocytosis

Agranulocytosis is a serious disease involving the white blood cells. It is often with reference to etiology as primary agranulocytosis being that form of disease in which the etiology is unknown and secondary agranulocytosis being that form in which the cause is recognized.

Agranulocytosis occurs at any age, but is somewhat more common in adults, particularly women.

The disease commences with high fever, accompanied by chills and sore throat. The patient suffers from malaise, weakness, and prostration. The skin appears pale and anemic, or in some cases, jaundiced. The most characteristic feature of the disease is the presence of infection, particularly of oral cavity, but also throughout the gastrointestinal tract, respiratory tract and skin. Regional lymphadenitis accompanies the infection in any this location.

The oral lesions constitute an important phase of the clinical aspects of agranulocytosis. These appear as necrotizing ulcerations in the oral mucosa, gingiva, tonsils and pharynx, fig 5, fig 6.

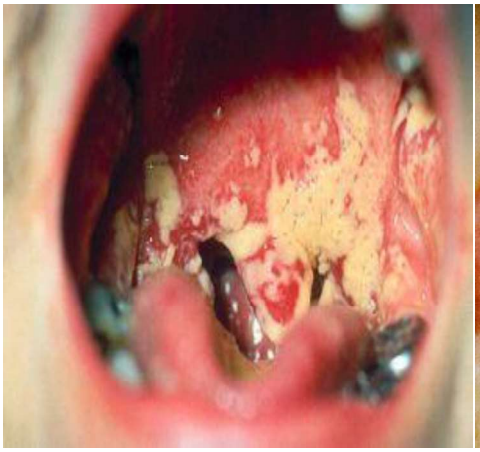


Fig 5

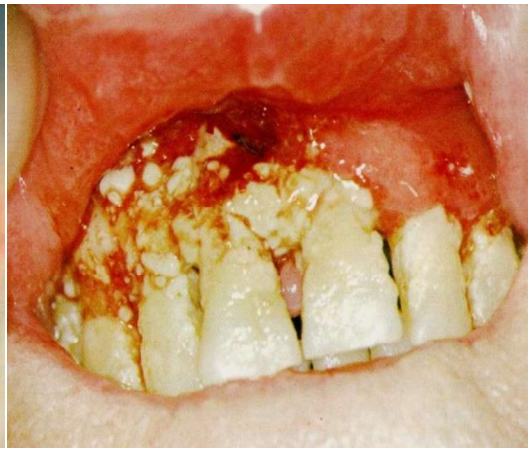


Fig 6

Particularly involved are three gingivas and palates .The lesions appear as raged necrotic ulcers covered by a gray or even black membrane, severe gingivitis rapidly progressive bone loss, and teeth loss may appear at an early age.

2.2. Leukemia

Leukemia is a disease characterized by the progressive overproduction of white blood cells which usually appear in the circulating blood in an immature form. This proliferation of white blood cells or their precursors occurs in such an uncoordinated and independent fashion that leukemia is generally considered a true malignant neoplasm, particularly since the disease is very often fatal, Bruch (10), Bricker (11)

The age of the patients affected by leukemia varies remarkably, but generally may be correlated with the course of the disease. Thus acute leukemia is more common in children and young adults, while chronic leukemia are most frequently seen in adults of middle age or older, Provn (3),Mc Kenna (12).

Acute leukemia is characterized by a rapid onset of weakness, malaise, fever, and generalized swelling of lymph nodes. Petechiae and ecchymoses of both skin and mucous membrane are common and with the lymphadenopathy may be the first manifestation of the disease. Bone and joint pains often occur referable to bone marrow involvement. Hepatomegaly and splenomegaly are sometimes seen, but usually in the acute form of the disease. Oral manifestations often are the first signs of the disease.

Chronic leukemia is often very insidious in its development, and clinical signs and symptoms may not appear until the disease in quite advanced. Patients may manifest a progressive weakness and malaise with loose of weight. A mild anemic pallor may be present as well as hepatomegaly and lymphadenopathy.

Oral manifestation of leukemia occurs in both the acute and chronic forms of the disease. Burket has reported oral lesions in 87% of patients with monocytic leukemia and 23% of patients with lymphocytic leukemia.

The most common oral manifestation of leukemia is gingival hyperplasia sometimes so severe that almost completely covers the teeth. In such cases, the gingiva is soft, boggy, deep red in color and tends to bleed spontaneously or following mild trauma. In nearly all cases, the hyperplasia is generalized except for edentulous areas.

The periodontal membrane may also exhibit pathological changes in the form of hemorrhage and necrosis, with subsequent necrosis of alveolar bonne.

Rapid loosing of the teeth due to necrosis of periodontal ligament has been reported, and destruction of the alveolar bonne also occurs in some cases. The use of panoramic radiographs, in a study of 214 children with acute leukemia has been reported by Curtis (14), to be useful in demonstrating previously overlooked changes in the jaws.

Oral mucosal lesions are greatly significant in the patients with acute leukemia since they are frequent and represent the points of intrusion of infectious agents in the development of serious systemic infections.

Infections are mainly bacterial, as numerous bacteria exist in the oral cavity, and, beside pathogenic microorganisms, their causative agents can be conditionally pathogenic microorganisms, as well, being that immunocompromised patients are involved. Fungal infections are also frequent and they are mainly of the Candida species in our climate, while most frequent viruses are Herpes simplex and Zoste, fig. 7 and 8.

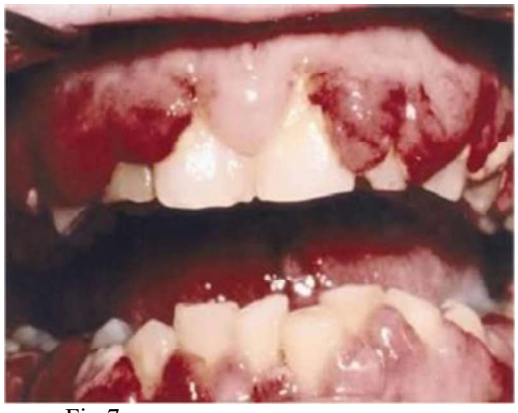


Fig 7

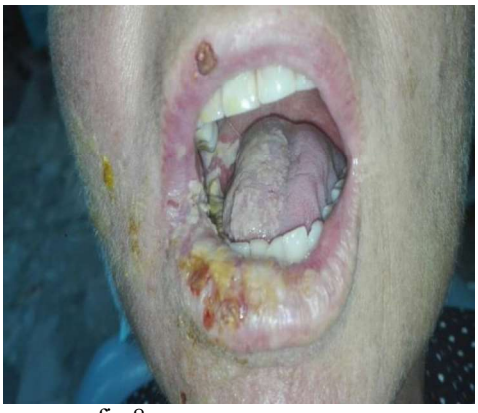


fig 8

Diseases of blood platelets

3.1. Thrombocytopenia

Abnormal bleeding associated with thrombocytopenia (low platelet count) or abnormal platelet function is characterized by spontaneous skin purpura, mucosal hemorrhages and prolonged bleeding after trauma, Girolamo (15).

Primary thrombocytopenia, although of obscure origin, has been suspected of being an autoimmune disorder in which a person becomes immunized against his own platelets.

Secondary thrombocytopenia frequently occurs following administration or assimilation of a variety of drugs and chemical including the barbiturates, sulfonamides, benzol, X ray radiation, sodium salicylate and so on.

Facial petechiae, conjuctivaes hemorrhage and hemorrhagic bullae in the oral mucous membrane occur in primary deficiency of platelets. These features are also seen in secondary thrombocytopenia due to the myeloblastic syndrome, autoimmune disorders, aplastic anemia and bacterial infections, collagen vascular disease, disseminated intravascular coagulopathy and drugs, Girolamo (15).

The oral manifestation consists of spontaneous gingival bleeding and excessive prolonged bleeding from minor oral surgical procedures such as dental extractions. Mucosal ecchymosis may be seen occasionally.

3.2. Hemophilia

Hemophilia is an X-linked hereditary disorder. Hemophilia A is a deficiency of factor VIII, while hemophilia B (Cristmas disease) is a deficiency factor IX. Factor VIII factor IX are important in the intrinsic phase of blood coagulation and their deficiency is considered severe when plasma activity of the deficiency in considered severe when plasma activity of the deficient factor is<1IU/dl normal range, 50-199, moderate if it ranges between 2 and 5IU/dl and mild if it is between 6 and 40 IU/dl, Kasper (16).

The disease is transmitted by affected males through infected daughters to grandsons. Therefore the disease is manifested in males but carried by females who themselves an unaffected. Generally, the sons of a man with hemophilia are normal and do not carry the defect.

Hemophilic patients clinically manifest prolonged and excessive bleeding following even the most minor traumatic injury.

Severe, prolonged gingival bleeding even following the most significant gingival injury is extremely common. Severe hemorrhage also occurs even with normal tooth eruption and exfoliation.

3.2. Platelet disorders in homeostasis

Abnormal bleeding associated with thrombocytopenia (low platelet count) or abnormal platelet function is characterized by spontaneous skin purpura, mucosal hemorrhages and prolonged bleeding after trauma, Chandra, (17).

Oral manifestation of thrombocytopenic purpura is the severe and often profuse gingival hemorrhage which occurs in the majority of cases. This hemorrhage may be spontaneous and often arises in the absence of skin lesion.

Petechiae also occur on the oral mucosa, commonly on the palate and appear as numerous tiny, grouped clusters of reddish spots only a millimeter or less in diameter. Actual ecchymoses do occur here occasionally.

The tendency for excessive bleeding contraindicates any oral surgical procedure, particularly tooth extraction, until the deficiency has been compensated.

Conclusion

Oral signs and symptoms may be the first clinical evidence of underlying blood disorders, and the dental health practitioner must pay careful attention to possible indicators of disease. Diseases of blood frequently affect soft and hard tissues of mouth with different characteristics. Oral manifestation of blood disease affect color of the mucosa hypertrophy of gingival, mucosal destruction in the form of ulceration, bleeding and hemorrhage, color of the tooth with red discoloration, lymph node and affect the bone with decreased density and enlarged marrow space. Every dentist should be familiarized the wide variety of oral manifestations of the blood diseases to differentiate from other diseases.

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