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OROFACIAL MANIFESTATIONS OF HEMATOLOGICAL DISORDERS

Dental Science

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ABSTRACT

Various anemic disorders which shows different orofacial manifestations are iron deficiency anemia,plummer Vinson syndrome,megaloblastic anemia,sickle cell anemia,thalassemia and aplastic anemia.These orofacial manifestations are facial pallor,atrophic glossitis,angular stomatitis,magenta tongue,midface growth etc.

Spontaneous & post traumatic hemorrhage,prolong bleeding after tooth extraction are sign of different hemostatic disorder such as Von-Willebrand disease or Hemophilia.

Oral manifestations of most of the hematological diseases are nonspecific.

In this article I have reviewed various literatures to identify orofacial manifestations of various hematological disorders.

KEYWORDS

Anemia, Atrophic glossitis,Hemophilia,Neutropenia

INTRODUCTION:

Oral cavity can be described as mirror of general health; as it reflects the presence of any systemic disease. Though, changes of oral cavity are not specific for any systemic disease, yet they can be described as distinctive & characteristic. This is true for hematopoietic system too.[1] Many of the cases of blood dyscrasias begins with subjective complains related to oral cavity, hence patients primarily visits to dentists to seek medical guidance.

The knowledge regarding orofacial manifestations of hematological diseases are important, as these signs and symptoms could be the first clinical presentation, which may help the dentist to diagnose & refer the cases to a hematologist.[1,2]

On the other hand, patients with known hematopoietic disorder or patients with general symptoms of hematopoietic disorder such as weakness,dyspnea etc or patients, whose relative is having bleeding & clotting disorders should be checked properly for any oral lesions and it should be taken care of .[1,2,3,4]

Along with oral mucosa, changes occur in adjacent bone in different hematological disease.[1,2,3]

Hematological diseases can be classified under four groups; disorders of red blood cells, disorders of white blood cells, histiocytic disorders and bleeding disorders.

In this article, different orofacial manifestations of hematological disorders are discussed elaborately.

Oral manifestations:

Petechiae and Ecchymoses:

When tiny red dots occur into the skin due to bleeding from broken blood vessels,it is known as petechia on the other hand when a large bruised area is formed due to collection of blood in larger flat areas under the skin is called ecchymosis.

These lesions are often found on the dorsum of tongue,over the mucosa of the floor of mouth,on palate,lips in connection with different hemorrhagic diatheses.[5,6,7]

These signs suggest the diagnostic consideration of Purpura;which may be thrombocytopenic or idiopathic or a secondary variety in conjunction with different hematopoietic or other diseases,such as Leukemia,Aplastic anemic, Scurvy etc.or toxic or infectious conditions.[5,6,7]

In thrombocytopenia either there is low platelet count or there are abnormal platelet functions.

Though similar signs are observed, yet in thrombocytopenic purpura,except bleeding tendency the overlying epithelium looks normal. On the other hand,in leukemia or aplastic anemia along with this special feature of pinpoint bleeding spot,they may show infiltrations and ulcers.[5,6,7] In scurvy, along with petechial hemorrhage,gingival margins are infected, swollen and have a purplish hue.

In some infectious process such as in sub acute bacterial endocarditis,similar phenomena resembling petechiae is observed.

Ecchymotic lesions and petechiae on oral mucosa,tongue,lips can also be seen in hereditary hemorrhagic telangiectasia and Ehlers-Danlos syndrome.[5,6,7]

Bleeding from mouth & Gum / Prolong bleeding after trauma:

Prolong and abnormal bleeding from mouth and gum after trauma is characteristic features of platelet disorder (qualitative or quantitative).[2,8]

On the other hand, along with continuous bleeding from oral cavity,brown discoloration of tooth due to deposit of fosters of hemosiderin and other blood degradation products on the surfaces of teeth is observed in von Willebrand's disease.[2,3,8,9]

Bleeding from multiple sites of oral cavity is common in Hemophilia. Deficiency of clotting factor –VIII is known as hemophilia-A, and deficiency of clotting factor-IX is known as hemophilia-B or Christmas disease. These factors help in blood clotting in intrinsic pathway.

Normal ranges of these factors are 50-100 IU/dl. When the ranges come down to 6-40 IU/dl,it is considered mild hemophilia; when it comes down to 2-5 IU/dl it is known as moderate hemophilia and when it comes down below1 IU/dl it is considered as severe hemophilia.[2,3,8,9,10,11,12]

Iatrogenic factors and poor oral hygiene practice may induce the bleeding in hemophilia. Even vigorous brushing or use of tooth picks can leads to bleeding.[11,12]

Kaneda et al mentioned that frequency of hemorrhage from different intraoral sites in hemophilia are gingiva (64%), dentalpulp (13%), tongue (7.5%),lip(7%),palate(2%) & buccal mucosa (1%).[13]

Analysis of patients' history plays an important role in diagnosis of hemophilia. Male gender, previous history of prolong bleeding, similar history of bleeding episode of male members of maternal side will help clinician to diagnose a patient with hemophilia.[2,3,9,10,11,12]

In hemophilia,minimal provocation will lead to severe bleeding. Due to exfoliation of deciduous teeth and eruption of permanent teeth, repeated prolong bleeding episode is common. Tooth extraction can be dangerous.[10,11,12]

In hemophilia, Intraorally no single point of origin of the bleeding can be located as well as gingiva doesnot look inflamed.

Hemophilia is a clotting disorder; hence blood remains unclotted for

long time. If it occurs below oral mucosa,it spreads widely and may give pressure on vital structures.

Similar bleeding episodes can be observed from oral mucosa and gum in leukemia (acute or chronic),hodgkin's disease and aplastic anemia.Bleeding episode for these disease is mainly due to deficiency of platelet. Leukemia patients will not give any history of previous episode of bleeding or any history of bleeding episode of any male maternal relatives as hemophiliacs.[2,3,9]

In all the bleeding disorders, infection of gingival is very common; as accumulated blood deposits act as a proper culture media for various microrganisms.

Tongue Changes:
Careful examination of tongue reveals various signs of hematological diseases. Large number of papillae is present in dorsum of the tongue, which gives a roughened appearance.

In pernicious anemia, tongue become red, swollen and feel sore in early days; later atrophy of mucosa and tongue papillae is observed, which gives the tongue a smooth, pale, shiny and glistering appearance.[14,15,16,17]

In this disorder (vitamin B12 deficiency), painful tongue and burning tongue is very common. [14,15,16,17]

Hypochromic microcytic anemia is mainly due to iron deficiency. In this anemia,due to atrophy of tongue papillae,tongue become 'beefy' red. In this disorder sometimes tongue becomes 'shrunken'.[1,2,9]

'Beefy' red tongue is also seen in avitaminosis,pellagra and sprue.
In most of the cases,fungal infection is associated with depapillation of tongue.

Angular Stomatitis and Cheilolosis:
Painful fissures at the corner of mouth is called as angular stomatitis;on the other hand, dry scaling of lips and corner of mouth is known as cheilosis. Both the lesions are common in iron deficiency anemia.[1,2,9]

Ulcers:
Recurrent apthous ulcer:
Minor apthous ulcer is common in iron deficiency anemia. These ulcers starts as vesicular elevation.Due to accumulation of serous fluid,the epithelium peels off and leaves a denuded surface,which is very painful due to uncovered nerve ending.

Various studies showed positive correlation between serum ferritin level and recurrent apthous stomatitis.[1,2,9]

Necrotic ulcers:
In agranulocytosis or in severe neutropenia necrotic type of ulceration is observed. This ulcer is different from apthous ulcer,which is regular in outline and clean in appearance. The necrotic ulcers are dirty looking and covered with grayish white membrane.[1,2,9]

In apthous ulcer, a zone of redness is found surrounding the lesions due to inflammation. On the other hand, no inflammation is present in necrotic ulcers. In these lesions,bacterial growth increases,tissue-cells undergo necrosis and foul odor is present.[1,2,9]

Similar type of necrotic ulcers are also seen in aplastic anemia, aleukemic leukemia,infectious mononeucleosis;as well as damage to bone marrow from radiation,arsenic or benzoyl poisoning where depression of mature leukocyte production occurs.[1,2,9]

Gingival growth:
In few varieties of leukemia,specially in monocytic type of leukemia;distortion of gingival margin and gingival mass is observed. The growth is due to extensive infiltration of masses of leukemoid cells into oral mucosa as well as submucosa at the area of gingiva.[1,2,9,18,19]

This gingival growth resembles drug induced gingival hyperplasia, specially Dilantin induced gingival hyperplasia.[1,2,9,18,19]

Localised treatment by dental surgeon will not improve the condition; instead tooth extraction may provoke a sudden increase in severity of underlying disease.[1,2,9,18,19]

Tonsillar enlargement:
Tonsill may get enlarged in lymphatic leukemia. The enlargement is rapid and severe. Even, it may occur before lymphadenopathy. As the lesion differs from ordinary tonsillitis,tonsillectomy should not be done; as it may lead to acute exacerbation of the underlying disease and severe hemorrhage.[1,2,9,20]

Lymphatic tissue and node enlargement:
In different hematological diseases,hyperplasia and hypertrophy of lymphatic tissue is a very common;specially enlargement of submaxillary and cervical lymph nodes.

In Hodgkin's disese, lymphadenopathy of face and jaw could be the first sign to diagnose the disease. In this disorder,nodes are unilaterally enlarged and discrete. It may appear that,only single chain of lymph nodes are affected. On palpation lymphnodes have rubbery or resilient consistency.[1,2,9,20]

On the other hand,in Lymphosarcoma,lymphnodes are firmer, indurated, adherent and less likely to be discrete. In this disorder also, cervical lymphadenopathy may be the first sign which helps in diagnosis.[1,2,9,20]

Lymph node enlargement may be seen in chronic lymphocytic leukemia.These lymph nodes only can be altered by radiation therapy.[1,2,9,20]

Changes in Bone:
Changes of jaw bone and facial bones are very common in different hematological diseases. Multiple myeloma,plasmacytoma etc are very prominent among them.[1,2,9]

Punched out radiolucency:
Punched out radiolucency is commonly seen in skull bone, ribs and spine of patients with multiple myeloma. These areas are painful and form due to replacement of bone structure by tumor like masses formed by myeloma cells.[1,2,9]

Moth eaten appearance:
Moth eaten appearance of rarefaction in mandible and skullbone is observed in few hematological diseases. These diseases are xanthomatosis or Hand-Schuller Christian diseases. These bony defects are filled with lipoid foam cells.[1,2,9]

Mottled appearance:
Mottled appearance of jaw bone is observed in radioactive poisoning. When radiation is used to treat oral tumor;it affects osseous component of mandible also. Due to this, radiation induced osteitis develops. Due to impared healing of bone; infection, osteomyelitis, sequestration develops.[1]

Mid-face overgrowth:
Midface overgrowth is commonly seen among patients' with Thalassaemia. These patients show bossing of the skull, enlarged maxilla, and prominent molar eminences. Due to overdevelopment of maxilla;spacing between teeth,increased overjet and other malocclusion develops.[1,2,9]

To compensate the severe anemic situation, there is intense compensatory hyperplasia of marrow and expansion of marrow cavity as a result; 'Chipmunk' face is observed.[1,2,9]

Mandibular Osteomyelitis:
Osteomyelitis is commonly observed in sickle cell anemia. Compared to maxilla, blood supply of mandible is insufficient. As a result, ischemic infract and osteonecrosis happens;which allows streptococcus or salmonella to proliferate.[21,22,23]

Changes in teeth:
Radioactive poisoning can be observed in teeth also. Caries like lesions are most prominent due to limited decalcification. They are termed “focal necrosis”.[1]

On the other hand asymptomatic pulpal necrosis is seen in sickle cell anemia.[24]

Changes in Periodontal Ligament:
Loss of attachment and periodontal destruction is common finding of Neutropenia. In this disorder,premature exfoliation of deciduous teeth is also a common finding.

Involvement of periodontal ligament space is also a frequent findings of non-hodgkins lymphoma.[1,2,9,20]

Pallor of buccal mucosa:

A study on Venizuelan population showed that,the most common intraoral soft tissue changes in sickle cell anemia was pallor of buccal mucosa.[25]

Paresthesia of Nerve:

Neuropathy of inferior alveolar nerve and paresthesia of lower lip is common finding of sickle cell anemia.[26,27]

Paresthesia is also a common finding of non-Hodgkin lymphoma.[28]

CONCLUSION:

Oral manifestations of hematological disorders are common and often seen in theearly course of the disease.Hence any unusual changes in oral cavity;such as bleeding tendencies,alteration of colour of mucosa,ulceration of oral cavity,lymph node enlargement may be manifestations of hematological disorder.

Hence, it should be correlated with history and clinical findings.
Dental surgeons should be aware about oral manifestations of hematological disorders.

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