CASE REPORT

Intraoral ecchymosis in myelodysplastic syndrome with myelofibrosis: A case report

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Abstract

Myelodysplastic syndrome (MDS) with myelofibrosis having intraoral manifestations as ecchymosis, unprompted gingival bleeding, and generalized gingival overgrowth with associated periodontal disease is one of the rare cases. MDS is a clonal disorder of pluripotent hematopoietic stem cells. Myelofibrosis is a disorder where normal bone marrow is replaced with a fibrous scar tissue. It is a complication of MDS. Here, we are discussing a rare case report of MDS with myelofibrosis manifesting as intraoral ecchymosis. MDS with myelofibrosis is aggressive disorder of elderly whose pathophysiology remains unknown with a poor prognosis. When myelofibrosis is observed in a bone marrow specimen, further investigation of the patient with respect to splenomegaly, leukocyte count, peripheral blood smear and bone marrow findings, and karyotype will serve as a guide to correct diagnosis.

Keywords: Intraoral ecchymosis, myelofibrosis, myelodysplastic syndrome, pancytopenia

Introduction

Myelodysplastic syndrome (MDS) is a clonal hematological disorder. Fibrosis of bone marrow can be associated with MDS, which is a very uncommon and constitutes only 5% of all cases. The characteristics of the disease are pancytopenia, minimal organomegaly, hypercellular bone marrow with marked fibrosis, dysplasia of erythrocytes, leukocytes and thrombocytes, and proliferation of megakaryocytes.[1-3] Oral manifestations of MDS are nonspecific and are usually consequences of cytopenia. In the review of literature, there are no case reports of oral manifestations of MDS with myelofibrosis. Here, we are presenting an intraoral manifestation of MDS with myelofibrosis.

Case Report

A male patient aged 70 years reported to the Department of Oral Medicine and Radiology with the chief complaint of missing upper teeth since 3 years and desired to get it replaced. The history of presenting illness was mobility followed by exfoliation of upper teeth. He gave a positive medical history of angioplasty done 15 years back and hypercholesterolemia. The patient was under antiplatelet (tablet ecospirin 75 mg) and hypolipidimic drugs (tablet lipitor) once daily. On general physical examination, a pallor was present in the nail beds and lower palpebral conjunctiva. On intraoral examination, oral mucosa was pale and multiple focal purplish erythematous areas were noted on the left and right buccal mucosa extending from retrocommisural area to lower buccal vestibule approximately measuring about 3 cm × 4 cm in size situated in relation to teeth 33, 34, 35 and 43, 44, 45 and similar lesions were also seen on ventral aspect of tongue and floor of mouth. On palpation, all the inspectory findings were confirmed. Lesions were non-tender and non-compressible suggestive of submucosal ecchymosis [Figures 1 and 2]. Marginal gingival erythema was present with bleeding on probing with mild inflammatory gingival enlargement. On hard tissue examination completely edentulous upper arch and partially edentulous lower arch was seen. A provisional diagnosis of anemic stomatitis was made, and the patient was advised for routine blood investigations such as complete blood count, hemoglobin (Hb) %, and peripheral blood smear. Complete blood examination revealed red blood cell (RBC) count 3.1 million/cumm, WBC count was 5400 cells/cumm, Hb were 7 g/dl, packed cell volume was decreased. Platelet count was 1.86 lakhs. Peripheral blood smear showed macrocytic anemia. The patient was referred...
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Figure 1: Submucosal ecchymosis in the left retrocomissural area and ventral surface of tongue

Figure 2: Submucosal ecchymosis in the right retrocomissural area and floor of mouth

to general physician for complete check up and consent for dental treatment. Patient reported back after 3 months being diagnosed as MDS with myelofibrosis. Patient had undergone bone marrow biopsy. Patient was non-responsive to lenalidomide, erythropoietin and danazol, therefore, was advised for regular blood transfusion. Patient was advised for complete hemogram once in every 2 weeks. Endoscopy examination revealed multiple erosions in the gastric mucosa. Patient’s recent blood picture was as follows RBC count 3.4 million/cumm, WBC count was 10,510 cells/cumm, lymphocytes counts were decreased, and monocytes were increased in number. Hb was 10.8 g/dl, packed cell volume was decreased. Platelet count was 2.41 lakhs. Patient was given consent for the dental procedure by a consultant in clinical hematology and hemato-oncology.

Discussion

The MDS, collectively characterized by clonal hematopoiesis, peripheral blood cytopenias, dysplastic cell morphology and risk of leukemic progression, continue to pose difficult challenges for patients and physicians. The etiology is commonly unknown. Less commonly, it is associated with antineoplastic chemotherapy and smoking, radiotherapy, environmental exposure to benzene petroleum products are the risk factors. MDS most commonly affects elderly individuals >70 years of age, it is uncommon in childhood. Clonal cytogenetic abnormalities in marrow cells are seen in about 50-60% of individuals in MDS. It can be associated with chromosomal loss or chromosomal gain. In this case, there was trisomy 8 unlike acute myelogenous leukemia (AML). Increased apoptosis of myeloid cells would provide an explanation of ineffective hematopoiesis leading to blood cytopenia like symptoms of anemia, infection due to neutropenia, bleeding due to thrombocytopenia, or due to thrombocytopenia, or due to thrombocytopenia. Several mechanisms underlie thrombocytopenia in MDS including ineffective platelet production secondary to disordered maturation and proliferation of megakaryocytes or their precursors increased megakaryocyte programmed cell death, increased peripheral destruction of platelets and autoantibody-mediated destruction of platelets and megakaryocytes. The clinical effects of thrombocytopenia in MDS can be exacerbated by functional platelet defects, including decreased surface expression of glycoproteins and abnormal alpha and dense granules. Bleeding events in MDS range from minor petechial or gingival bleeding to life-threatening gastrointestinal or intracranial hemorrhage. Bleeding rates vary widely across reported MDS series; with spontaneous mild bleeding occurring in 18-23% regardless of platelet count. As in our case bleeding spots were seen in oral cavity as well as gastric mucosa. Thrombocytopenia in MDS is not only associated with bleeding but is also an independent predictor of progression to AML and reduced survival.

The bone marrow aspirate is usually normocellular or hypocellular and sometimes hypercellular owing to marrow fibrosis. Myelofibrosis is a complication of MDS. It is an indication of rapid progression to leukemia or poor prognosis for survival. MDS with MF is closely associated with the neoplastic proliferation of megakaryoblasts. The cause for myelofibrosis in MDS is attributed due to an increase in the levels of plasma transforming growth factor-beta 1.

MDS is a diagnosis of exclusion; one should exclude several causes of myelodysplastic features of marrow precursors such as megaloblastic anemia, sideroblastic anemia, aplastic anemia, infections with viral particularly HIV, chronic liver disease, and alcohol abuse. The drugs available to treat the condition are erythropoietin, darbepoetin, lenigrastim (GCSF), 5-azacytidine, decitabine, anti-thymocyte globulin, cyclosporine, lenalidomide, deferasirox, and deferroxamine. The Treatment of anemia in MDS is usually done by blood transfusion. The mainstay of the treatment has been entirely supportive with whole blood
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Intraoral ecchymosis in myelodysplastic syndrome with myelofibrosis.\(^{[1]}\) The final treatment option is autologous stem cell transplantation.\(^{[2]}\)

**Conclusion**

MDS with myelofibrosis is an aggressive disorder of elderly whose pathophysiology remains unknown with poor prognosis. We described a unique patient with MDS and myelofibrosis having intraoral ecchymosis which was a key for further investigations and helps in identifying the condition.

**Clinical significance**

The majority of systemic diseases will be having oral manifestations in one or more various stages of the disease. The recognition of oral lesions, correlating it to the underlying systemic disease and prompt referral to the concerned medical practitioner should be prime commitment of the dental practitioner which plays a crucial role in timely diagnosis and prognosis of the systemic disease.

**References**