Chronic Myeloproliferative Disorders: A Rarest Case with Oral Manifestations and Dental Management

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ABSTRACT

Chronic myeloproliferative disorders (CMPD) are rarest hematological disorders (malignant myeloid neoplasms). The three most common chronic myeloproliferative disorders are polycythemia vera, essential thrombocythemia and chronic idiopathic myelofibrosis. Clinical manifestations (including oral manifestations) of these disorders are overlapping with each other and with other hematologic disorders, which makes the diagnosis of CMPD a challenging task. In this article we report a rare to rarest case of CMPD at dental outpatient department, its oral manifestations and its management in dental clinics.

Keywords: Chronic myeloproliferative disorder, Essential thrombocytosis, Idiopathic myelofibrosis, Polycythemia vera.


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INTRODUCTION

Chronic myeloproliferative disorders (CMPD) are rarest hematological disorders (malignant myeloid neoplasms). Traditionally polycythemia vera (PV), essential thrombocythemia (ET) and chronic idiopathic myelofibrosis (IMF) have been classified under the term ‘the chronic myeloproliferative disorders’ (CMPD) because they share the following features: Involvement of a multipotent hematopoietic progenitor cell; dominance of the transformed clone over nontransformed hematopoietic progenitor cells (clonal dominance); overproduction of one or more of the formed elements of the blood in the absence of a definable stimulus; marrow hypercellularity; thrombotic and hemorrhagic diathesis; exuberant extramedullary hematopoiesis; and spontaneous transformation to acute leukemia or the development of marrow fibrosis but at a low rate compared with the rate in chronic myelogenous leukemia (CML).1-4

The recent World Health Organization classification of the chronic myeloproliferative diseases includes in addition to PV, IMF and ET, the following disorders: CML, chronic neutrophilic leukemia, chronic eosinophilic leukemia, the hypereosinophilic syndrome and chronic myeloproliferative disease-unclassifiable.5

Clinical manifestations (including oral manifestations) of these disorders are overlapping with each other and with other hematologic disorders, which makes the diagnosis of CMPD a challenging task. In this article, we report a rare to rarest case of CMPD at dental outpatient department (OPD), its oral manifestations and its management in dental clinics.

CASE REPORT

A 53-year-old female Hindu patient came to us in June 2010, with the chief complaint of bleeding gums, mobility of teeth in lower front teeth region and difficulty in mastication since 1 year. Upon extracting detail history about her general medical health it was revealed that, she was diagnosed with chronic myeloproliferative disorder in September 2004 when she had consulted a gynecologist for excessive menstruation who had advised her routine blood investigations before surgery, which revealed abnormally high hemoglobin percentage, high erythrocyte count, increased hematocrit and high platelet count. She was then referred to the hematono- oncologist by the gynecologist for expert opinion. Upon seeing laboratory reports and confirming the diagnosis, cyto reductive therapy (hydroxyurea bid for 3 days/week) was started by the hematono- oncologist. The doctor had also advised her that due to the risk of thrombotic or hemorrhagic event she must consult him before taking any medications (e.g. non- steroidal anti-inflammatory drugs), injections or before going to any kind of surgical procedure. Her family history was not significant. Intraoral examination (Figs 1 and 2) revealed poor periodontal status with missing 17, 31, 36, 41 and 46 which were exfoliated on their own. Mobility was noted in all the present teeth. Gingiva was red and edematous. Generalized gingival enlargement with loss of stippling and recession was noted. It was soft and friable which was bleeding profusely on touch or provocation. Patient was advised orthopantomogram, which also showed generalized bone loss (Fig. 3). On the basis of thorough examination total extraction followed by complete denture construction was advised to the patient. Medical clearance was taken from the hematono- oncologist before treatment and all required blood investigations were done to keep check on blood picture. Tablet traxamic 500 mg qid (tranexamic acid-antifibrinolytic drug) was started 1 day before treatment to prevent hemorrhagic event. Extraction of 32, 33, 42 teeth with supported bridge was done on the next planned sitting
under complete antibiotic and analgesic coverage. No uneventful complication was noted immediately after extraction and on follow-up appointments after 1 week.

**DISCUSSION AND CONCLUSION**

CMPDs are rare to rarest; least understood and most understudied hematological disorders which require special attention due to the risk of malignant transformation. More commonly they occur in 4th to 6th decade of life and male to female ratio is 1.4:1 but in the reported case the patient is a 53 years old female. Most often these patients are diagnosed accidentally due to hemorrhagic or thrombotic event which is true in our case also as excessive menstrual bleeding was the feature for which patient consulted a gynecologist and her disease was diagnosed on the basis of abnormal blood picture.

Oral manifestations (such as poor periodontal health, gingival enlargement and easy bruising) of a patient of CMPDs disorders are also overlapping with each other and with other hematological disorders. In the reported case poor periodontal health (gingival enlargement with bleeding gums and mobility of teeth) was the chief complaint for which she had consulted us. Thus, it is the duty of an oral stomatologist or diagnostician to think of CMPDs in differential diagnosis, if an old patient with significant past medical history/drug history consult the dental surgeon for poor periodontal health especially with gingival enlargement or easy bruising.

Dental management of such patients includes consultation with a hemato-oncologist and repeated complete blood picture to assign the patient to a risk category (high-risk, intermediate-risk, low-risk) from which treatment recommendations follow and to check the side effects (such as macrocytic or leukemogenic changes) of cytoreductive drugs. High-risk patients can be defined as those 60 years of age or older or those who have had a thrombosis or hemorrhagic event at any age. These patients should be treated mainly with cytoreductive drugs (e.g. hydroxyurea). Intermediate-risk patients are those less than 60 years who have not had thromboses, but who have platelet counts >1,500 × 10^9/l or who have significant cardiovascular risk factors. These patients should have their risk factors treated and may be given low-dose aspirin (40-325 mg). Low-risk patients are those less than 60 years old who have not had thrombosis, who have no cardiovascular risk factors, and whose platelet counts are <1,500 × 10^9/l. These patients can be observed or placed on low-dose aspirin (along with phlebotomy in PV). In the reported case as the patient had a history of hemorrhagic event (excessive menstruation), she was assigned a high-risk category by the hemato-oncologist and was on a cytoreductive drug (hydroxyurea) since 2004. Her latest blood picture did not show any abnormality (in relation to the disease or to the side effects of cytoreductive drug), on the basis of which medical clearance with specific precautions/instructions [such as premedication with an antifibrinolytic drug, an entally acting nonsteroidal anti-inflammatory drugs (NSAIDs) or opioid analgesics and

**Fig. 1:** Generalized gingival enlargement

**Fig. 2:** Gingiva is reddish pink, soft and friable which bleeds on slight provocation

**Fig. 3:** OPG shows generalized and severe bone loss
antibiotics and avoidance of NSAIDs] was obtained from the hemato-oncologist. All the instructions were strictly followed by us in the reported case and patient was managed without any complication due to a good team work.6-8

REFERENCES


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