

## Buccal space hematoma- A rare presentation in chronic myeloid leukemia

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### Abstract

Chronic Myeloid Leukemia (CML) typically seen in older patients, increases with age. Patients often are asymptomatic at the time of diagnosis and the disease is found incidentally on examination of blood. Oral ulceration, petechiae, mucosal bleeding, gingival enlargement, acute periodontitis, noma, trismus and oral infections are the most common oral manifestations. It occurs either due to direct infiltration of leukemic cells (primary) or secondary to underlying thrombocytopenia, neutropenia or impaired granulocyte function. We present a rare case of buccal space hematoma secondary to CML.

**Keywords:** Chronic myeloid leukemia, Buccal space hematoma, Facial swelling, Oral manifestation.

### Introduction

Chronic myeloid leukaemia (CML) is a myeloproliferative neoplasm characterized by an excess of granulocytes (neutrophils, basophils, eosinophils). It is a clonal disorder that originates from a bone marrow haemopoietic stem cell.<sup>1</sup> It can occur at any age, but half of the cases are diagnosed in people >65 years of age. There is a slight male predominance and a higher incidence among those with radiation exposure.<sup>1</sup> It is rarely seen in children and, if found, carries a poor prognosis.<sup>2</sup> Approximately 30% of patients with CML are asymptomatic, the diagnosis being made following an incidental finding of leucocytosis on a full blood count. Symptoms at presentation are often nonspecific and include fatigue, night sweats, weight loss, abdominal discomfort, spontaneous bleeding or bruising. Patients presenting with very high white cell counts can have symptoms associated with leucostasis, including headaches, visual disturbance, dyspnoea and priapism.<sup>3</sup> Oral ulceration, petechiae, mucosal bleeding, gingival enlargement, acute periodontitis, noma, trismus and oral infections are the most common oral manifestations. It occurs either due to direct infiltration of leukemic cells (primary) or secondary to underlying thrombocytopenia, neutropenia or impaired granulocyte function.<sup>2</sup> We discuss a case of buccal space hematoma as a rare presenting feature of CML.

### Case Report

A 28 years old female reported to the Oral & Maxillofacial Surgery outdoor department with sudden appearance of painful swelling over right side cheek for

last three days. Patient was apprehensive, irritable with low grade fever (99.8F). On clinical examination, a soft, fluctuant, tender swelling (4 x 2 cm) over right mid-face region mimicking buccal space infection was noted. Intraoral examination revealed obliteration of buccal vestibule with deep bluish discoloration of overlying mucosa. No odontogenic cause was associated. No history of systemic illness was obtained. Generalized systemic examination revealed hepatosplenomegaly. Hematological examination showed raised total leucocyte count (384000/cu.mm. with blast cells 1%, metamyelocytes 20%). Total red blood cell count (2.51 million/cu.mm) and platelet counts (80000/cu.mm) were decreased. She was anaemic with haemoglobin value of 9.31g/dl. Viral markers (HIV, HBsAG, HCV) were negative. On aspiration of swelling, clotted blood was recovered. Surgical decompression was performed to relieve the pressure. Genetic analysis confirmed the fusion of bcr-abl (90%). The diagnosis of chronic myeloid leukemia was confirmed. Patient was referred to the Department of Medicine for further management. She was advised to take tab Imatinib mesylate (Gleevec) 400 mg once daily. Packed cell RBC, Fresh frozen plasma and platelets were infused to maintain the normal parameters. The swelling reduced in size after one month of course. The routine hematological reports were also within normal limits. Patient was kept under regular follow up.



**Fig.1:** Frontal view showing swelling over the right-side mid face region.



**Fig. 2:** On aspiration blood was recovered.



**Fig. 3:** After one month chemotherapy, resolved swelling over the face.

### Discussion

Leukemia is a disease of mesenchymal origin in which there is an abnormal proliferation or an increased lifespan of myeloid or lymphoid cells. Pathologic changes may occur within the oral cavity early in the course of the disease or may be induced in oral tissues secondary to treatment or as delayed sequelae of the disorder.<sup>4</sup>

CML has a worldwide annual incidence of 1-1.5 cases per 100,000 populations. CML is diagnosed by the identification of reciprocal translocation of chromosomes 9 and 22, also termed as Philadelphia chromosome. With disease progression, additional cytogenetic abnormalities can develop, as well as mutations of the BCR-ABL

kinase domain, which are associated with treatment resistance.<sup>1</sup>

Generally there are three phases of the disease. The initial or chronic stage typically lasts 5 to 6 years. In the chronic phase, the peripheral blood reveals elevated counts of platelets and elevated white blood cell counts. Mature granulocytes among metamyelocytes and myelocytes often are seen. Bone marrow shows near 100% cellularity compared with the usual 50%. Extramedullary hematopoiesis occurs, resulting most commonly in splenomegaly and possible hepatomegaly and lymphadenopathy. The second phase, also known as accelerated stage, presents with anemia, basophilia, and thrombocytopenia. The third stage, blast crisis is marked by a transition from a chronic leukemia to an acute leukemia. Blast crisis leads to rapid deterioration in health and eventual death of the patient.<sup>2,5</sup>

Common oral manifestations of CML are gingival enlargement and mucous membrane bleeding in the form of bruise, purpura, petechiae, ecchymosis, epistaxis because of abnormal platelet function. Patient may present with infections due to dysfunctional neutrophils and because of leukocytosis there may be features of hyper viscosity syndrome e.g. veno-occlusive disease, cerebrovascular accident, myocardial infarction, priapism, visual disturbances and pulmonary infarction.<sup>2,6</sup> In the patients with CML who have bleeding complications, qualitative platelet abnormalities are frequently observed. Thrombocytosis is found in one third of CML patients and one sixth patients have platelet dysfunction.<sup>6</sup>

Soft tissue hematomas are rarely found in blood malignancies. Cases of mediastinal haematoma, haemothorax and large haematoma in the anterior as well as posterior compartment of left thigh have been reported in patients diagnosed with CML, which were treated successfully with hydroxyurea and imatinib.<sup>7,8</sup> Retroperitoneal hematoma also has been diagnosed in a case of CML responding to imatinib and later went for stem cell transplant. There is report of spontaneous spinal epidural hematoma (SSEH) detected through MRI, as the initial presentation of leukemia which was managed by surgical decompression within 48 hours.<sup>9,10</sup> There are incidences of three cases of sub-dural hematoma (SDH) during the remission phase of CML which required surgical intervention. In present case, patient reported us with a facial swelling mimicking as buccal space infection. Hematological investigations confirmed the

diagnosis of CML. Till date no case of CML has been reported presenting initially as buccal space hematoma.

Management of CML has been revolutionized by the introduction of tyrosine kinase inhibitors (TKIs; e.g. imatinib). Imatinib mesylate (Gleevec, 400 mg once daily) acts by inhibiting the TK in its inactive form by binding to the ATP binding site. In adults, doses of 600 to 800 mg can be used during the blast crisis. The response is poor for those who begin their treatment in the blast phase. Monitoring of disease plays an important role.<sup>11-13</sup> CML has a median survival of 5-7 years. The 5-year overall survival in the chronic phase is 92 to 95%. Prognosis in blast-phase CML remains very poor, with a median survival of 7-11 months despite multiple therapies.<sup>14</sup> Surgery is reserved for mucosal disorders requiring biopsy or supportive therapy, gingiva-periodontal disease, soft tissue or bony trauma, mucosal pathology, tumors, or developmental disorders. Surgical decompression was carried out to relieve the pressure and pain from the swelling.

### Conclusion

In a patient presenting with soft tissue haematoma, after exclusion of infection and traumatic cause, hematological disorder should be kept in mind. If haemogram and peripheral blood film show abnormal number and atypical morphological blood cells, then hematological malignancies may be suspected.

**Conflict of Interest:** None.

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