Case Report

Diffused mixed B-cell non-Hodgkin lymphoma of mandible

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Abstract Lymphomas are the third-most common cancer of the oral cavity after squamous cell carcinomas and salivary gland tumors. It is characterized by proliferation of lymphoid cells and their precursor. Diffuse B-cell non-Hodgkin lymphoma is the most common histological type of lymphoma in the head-and-neck region and most commonly affects older men in their seventh decade of life.

Keywords: B-cell, CD 20, non-Hodgkin lymphoma

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INTRODUCTION

Lymphomas are the malignant neoplasms of lymphocyte cell lineage.[1] They are mainly classified as either Hodgkin's or non-Hodgkin's lymphoma.[1] Extranodal presentation of Hodgkin's disease is rare; non-Hodgkin's lymphoma arises primarily within the lymph nodes, but approximately 24% affect extranodal locations such as the stomach, skin, lung, salivary glands and rarely occur in the mouth.[1,2] In the head-and-neck region, Waldeyer's ring is the most common site of extranodal non-Hodgkin lymphoma.[3] The most common clinical appearance of non-Hodgkin's lymphoma in the mouth is a nonhealing, nontender, painless ulceration commonly affecting the vestibule, gingiva or posterior hard palate and resembling a dental abscess of endodontic or periodontal origin.[4] Non-Hodgkin lymphoma is characterized by an abnormal proliferation of T- and B-cells or both, but most such lymphomas have been shown to be predominantly of B-cell origin, more than 90% of patients expressing CD20 antigen.[2,5] When involving the bone, the tumor produces extensive destruction, and lesions on radiological examination usually show ill-defined osteolytic changes.[6]

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CASE REPORT

A 50-year-old male reported to the department of oral medicine and radiology with the chief complaint of swelling in the lower jaw on the left side for 25 days. The patient gave a history of painful decayed lower left posterior teeth 30 days back, which was extracted in the private clinic. Following extraction, swelling did not resolve, gradually increased over a period of 20 days to attain the present size. The patient gave a history of difficulty in chewing. He gave no history of paresthesia. Past medical, surgical and drug histories were unremarkable. He is chronic bidi smoker for 30 years and consumes one bundle of bidi per day.

On general physical examination, the patient built was moderate. Left side submandibular and deep cervical lymph nodes were enlarged, palpable and tender measuring 1.1 cm × 1.4 cm and firm in consistency. The left submandibular lymph node was fixed to the underlying structures. Lymph nodes of other regions were not palpable.

On extraoral examination, a solitary swelling on the left side of the mandible extending from the left lower lip

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to the lower body of mandible, superoinferiorly and mediolaterally, the swelling extending from the angle of the mouth to the angle of mandible on the left side [Figure 1]. The surface of the swelling was smooth. On palpation, swelling was nontender, firm in consistency, nonfluctuant, noncompressible and nonpulsatile.

Intraoral soft-tissue examination revealed an ulcerated mass, extending from 34 tooth region till the ascending ramus of the mandible [Figure 2]. It also obliterated both the buccal and lingual vestibular area on the left side. On palpation, the mass was moderately tender, firm in consistency, nonfluctuant and noncompressible.

Both patient history and clinical examination were suggestive of a malignant neoplasm of the mandible. A differential diagnosis of squamous cell carcinoma, non-Hodgkin lymphoma, Ewing's sarcoma, osteosarcoma, chondrosarcoma, neurosarcoma and fibrosarcoma were given.

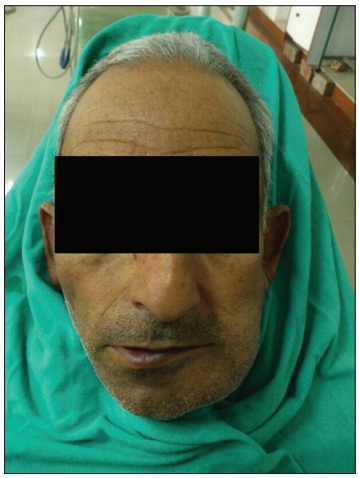

Figure 1: Extraoral photograph showing a left mandibular body swelling, expanding both laterally and inferiorly

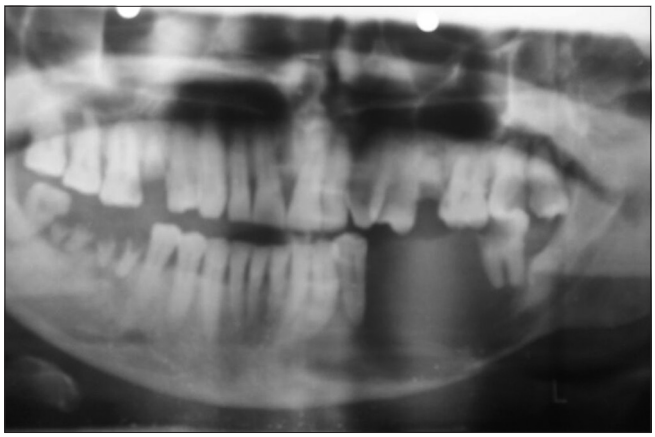

Figure 3: Orthopantomogram revealing pathological fracture of the left lower jaw and a diffuse, large radiolucency extending from 34 tooth region till the ascending ramus of mandible anteroposteriorly

Hematological investigations were performed which showed increased white blood cell count (12,550/mm^3) and raised erythrocyte sedimentation rate (18 mm) was observed. HIV test was also performed but found to be negative.

Radiographically, orthopantomogram was suggestive of pathological fracture of the left lower jaw, and a diffuse, large radiolucency was observed from 34 tooth region till the ascending ramus of the mandible anteroposteriorly leaving a thin border of the mandible [Figure 3].

On incisional biopsy, a soft-tissue specimen was removed in an abundant depth till the bone, measuring of size 2 cm × 1.2 cm × 0.7 cm, creamish white to light brown in color with rough irregular surface and soft to firm in consistency [Figure 4] under local anesthesia with respect to the left side of the mandible and were taken for histopathological examination.

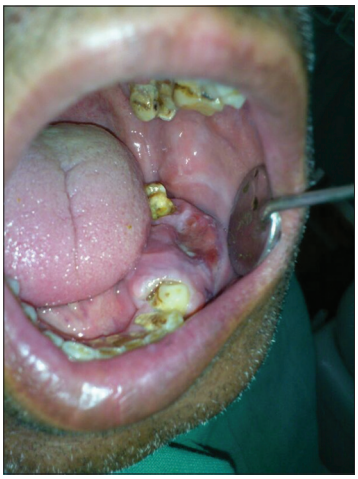

Figure 2: Intraorally an ulcerated mass, extending from 34 tooth region till the ascending ramus of mandible and invading both the buccal and lingual vestibular area of the left side

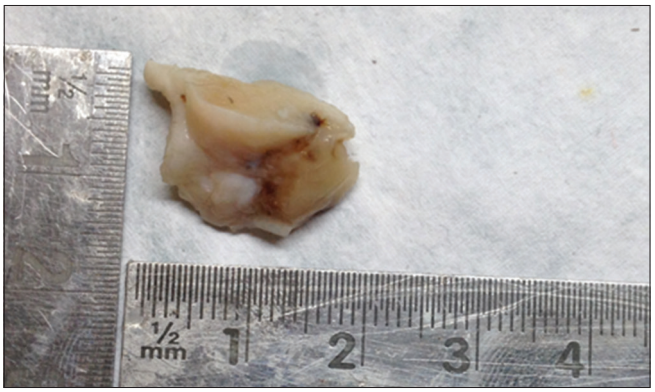

Figure 4: Incisional biopsy photograph showing soft-tissue specimen of size 2 cm × 1.2 cm × 0.7 cm, creamish white to light brown in color with rough irregular surface and soft to firm in consistency

Microscopic examination of the specimen revealed the presence of parakeratinized stratified squamous epithelium with underlying cellular connective tissue stroma. Connective tissue showed the presence of malignant cells resembling small and large lymphocytes arranged in the form of sheets [Figure 5]. The large tumor cells exhibited large, pleomorphic nucleus with hyperchromatic and coarse chromatin. Few cells had prominent, multiple eosinophilic nucleoli. Some large round cells with centrally placed large vesicular nucleus were also seen. These cells were surrounded by a clear halo and exhibited a very lightly stained cytoplasm [Figure 6]. Immature large cells with cleaved nucleus were present. Numerous abnormal mitotic figures that are 1–4 per high-power field, and in some areas, large binucleated cells with pale cytoplasm and prominent nucleoli were seen [Figure 7]. Numerous smaller cells with dark hyperchromatic nuclei and minimal cytoplasm resembling small lymphocytes were also seen. These cells are at least double the size of small lymphocytes. Numerous endothelial lined blood vessels are seen along with hyalinized collagen fibers.

By seeing the round cells in microscopic examination of the present case, various histopathological differential diagnoses were given such as Ewing's sarcoma, peripheral neuroectodermal tumor, rhabdomyosarcoma, synovial sarcoma, non-Hodgkin's lymphoma and desmoplastic small round cell tumor.

Ewing's sarcoma mainly affects the lower extremity and femur region and predominates in the second decade of life.[7] Histopathologically, this tumor is composed of small round cells with regular round nuclei containing finely dispersed chromatin and inconspicuous nucleoli and a narrow rim of clear or pale cytoplasm.[7]

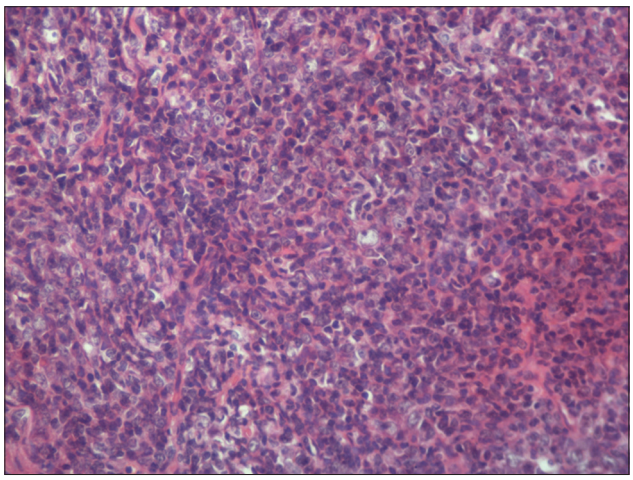

Figure 5: Connective tissue showed the presence of malignant cells resembling small and large lymphocytes arranged in the form of sheets (×4)

Rhabdomyosarcoma is the most commonly found soft-tissue sarcoma in children, wherein the cancer cells are thought to arise from skeletal muscle progenitors.[8] Microscopically, it exhibits cross striation characteristics of skeletal muscle or rhabdomyoblasts.[8]

Synovial sarcoma is rare and most commonly seen in young adults between the ages of 15 and 30 years.[9] The monophasic subtype of synovial sarcoma shows spindle cell sarcoma with hemangiopericytoma-like foci and the biphasic type contained epithelial-like glands and nests, and the presence of mast cells is also an important differential diagnostic feature of synovial sarcoma.[9]

Non-Hodgkin lymphoma shows atypical lymphoid cells arranged in diffuse sheets in a delicate connective tissue stroma with hemorrhagic areas, and tumor cells showed indistinct margins having vesicular, hyperchromatic and pleomorphic nuclei with a prominent nucleolus and abundant eosinophilic cytoplasm.[1]

Desmoplastic small round cell tumors are composed of sharply demarcated nests of varying size with small round or oval cells embedded in the hypervascular desmoplastic stroma, and large tumor cell nests have central necrosis.[10]

B seeing the histopathology of present case which was consistent with histological feature of non-Hodgkin lymphoma as discussed above in differential diagnoses.[1] Hence, the final diagnosis was given non-Hodgkin lymphoma which was further confirmed by the primary immunohistochemical examination. Immunohistochemistry was performed which showed that lymphoid cells were

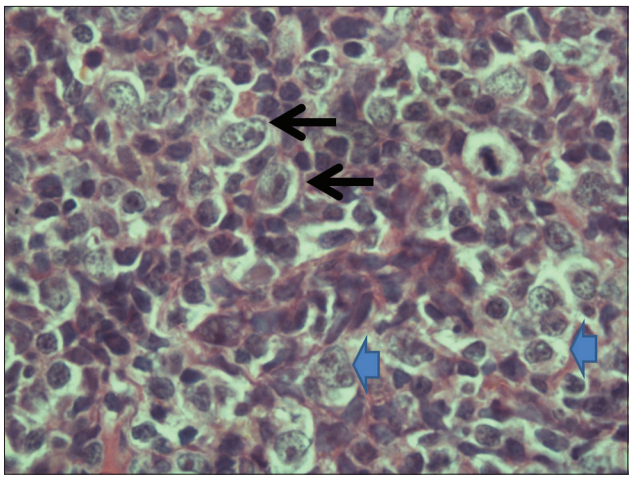

Figure 6: Large binucleated cells with pale cytoplasm and prominent, multiple eosinophilic nucleoli are seen (arrow head), some large round cells with centrally placed large vesicular nucleus surrounded by a clear halo and exhibited a very lightly stained cytoplasm (black arrow) (H&E, ×40)

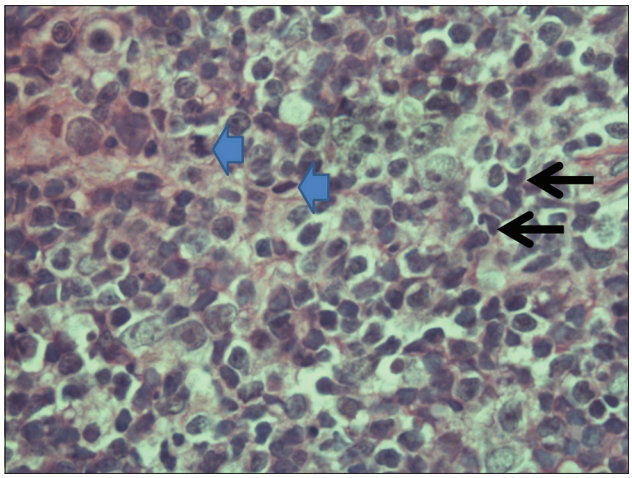

Figure 7: The large tumor cells exhibited large, pleomorphic nucleus with hyperchromatic and coarse chromatin, numerous abnormal mitotic figures that are 1–4 per high-power field (arrow head) and cleaved nucleus (black arrow) are seen

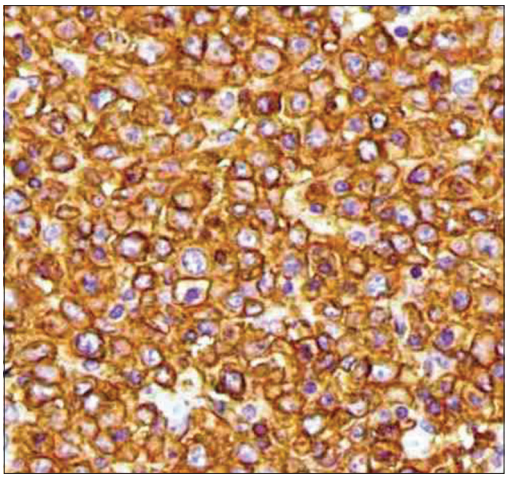

Figure 8: Immunohistochemistry stained picture showing CD20 positive cells

immunopositive for CD20/PAX5 [Figure 8] and were immunonegative for CD30/Bcl6. The Ki-67 proliferative index is approximately 30%. The histopathologic features together with immunohistochemical profile, led to the diagnosis of diffused B-cell lymphoma, mixed small and large cell type.

On asking the patient's relative, no abnormality was found in chest radiograph, abdominal ultrasound, bone scan and bone marrow biopsy. This confirmed this to be a focal lesion. The patient was kept on sequential courses of chemotherapy and treated. Later, he did not report for follow-up. Ethical clearance was not sought in this case as all guidelines prescribed by Helsinki declaration have been followed, and no patient-related personal data were revealed.

DISCUSSION

Malignant lymphomas of the oral cavity represent 5% of all lymphomas, with the diffuse large B-cell lymphoma being the most frequent type.[11] Histologically, diffuse large B-cell lymphoma contains large lymphoid cells with abundant cytoplasm and nuclei, larger than reactive histiocytes.[12] For confirmation of the diagnosis at least, 1 pan-B cell marker, including PAX5, CD79 and CD20, should be expressed distinctively in diffused large B-cell lymphoma.[13] In the present case, lymphoid cells were immunopositive for CD20/PAX5 and immunonegative for CD30/Bcl6. CD20 is classified under pan-B cell marker, and its presence on benign and neoplastic lymphocytes is generally considered specific for B-cell lineage.[14] PAX 5 is a member of the paired box transcription factors involved in the development and is expressed in hematopoietic malignancies of B-cell lineage, and it may be used as an adjunct marker in the diagnosis of classical Hodgkin lymphoma and non-Hodgkin lymphoma.[15]

CONCLUSION

In the present case, the patient presented all the necessary diagnostic criteria. The present case is unique as a "diffused B-cell lymphoma, mixed small and large cell type" of mandible origin which invaded both the buccal and lingual cortical plates. This case presents as a swelling affecting the jaws, which might be encountered in a dental setting. It might be a challenging task for clinicians to diagnose or determine the correct course of management. An understanding of the same can help in timely diagnosis and treatment that can help in reducing the morbidity and mortality in such cases by a collaborative effort by clinicians and histopathologists.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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