Lymphoblastic lymphoma (LBL) is a rare aggressive neoplasm of T-cell or B-cell precursors resembling acute lymphoblastic leukemia, with no or limited bone marrow involvement, that develops more frequently in children and young adults. Lymphoblastic lymphoma of the B-cell type is uncommon, and extranodal presentation is even rarer. We report what is, to the best of our knowledge, the first reported case of B-lymphoblastic lymphoma (B-LBL) of the hard palate.

CASE REPORT

A 49-year-old man presented with pain and swelling in the hard palate for 3 months. Examination of the oral cavity showed diffuse, soft, nontender swelling in the hard palate (Figure 1a). There was no pallor, lymphadenopathy, or organomegaly. Computed tomography scan of the head revealed irregular rarefaction of the anterior aspect of the hard palate. An incisional biopsy of the lesion disclosed subepithelium diffusely infiltrated with a monotonous population of medium to large cells with vesicular nuclei, prominent nucleoli, and scanty cytoplasm (Figure 2). On immunohistochemistry, the cells were positive for CD20, CD34, Bcl2, and Tdt with an MIB labeling index of about 90%. The picture was diagnostic of B-LBL. His hemoglobin was 15 g/dL; platelets, 3.5 lakhs/mm³; and total leucocyte count, 7300/mm³. Lactate dehydrogenase was 540 U/L. His cerebrospinal fluid and bone marrow studies were normal. Computed tomography scans of the neck, thorax, abdomen, and pelvis were normal, and he was staged as stage 1. The patient was started on rituximab, cyclophosphamide, vincristine, doxorubicin, and dexamethasone (R-Hyper CVAD protocol). After completion of one cycle of chemotherapy, the lesion regressed (Figure 1b). He is on maintenance chemotherapy.
of the first cycle of chemotherapy, his symptoms subsided and the lesions showed significant clinical and radiological regression (Figure 1b). At present, he is on maintenance chemotherapy.

**DISCUSSION**

LBL is a highly aggressive neoplasm of lymphoblasts that may be of either T-cell origin (T-LBL) or B-cell origin. Lymphoblastic lymphoma accounts for approximately 2% of all non-Hodgkin lymphomas, out of which T-LBL constitutes around 90% of cases (1). LBLs are grouped together with acute lymphoblastic leukemia in the 2008 World Health Organization classification of hematopoietic malignancies (2). These two entities are biologically very close but not identical; in LBL, the bone marrow is not involved or is only partially involved, with less than 20% infiltrating blast cells. LBL occurs commonly in children, mostly males. T-LBL usually presents with a mediastinal mass, central nervous system involvement, and pleural and pericardial effusion, whereas B-LBL presentation is more limited than that of T-LBL and the localized disease usually involves single nodal or extranodal sites such as skin, bone, and soft tissue (3, 4).

Lymphoid lesions of the palate can be either lymphomatous or benign lymphoid hyperplasia. Oral lymphomas are relatively rare and constitute about 4% of all oral malignancies (5). The oral cavity is the primary site of approximately 2% of all extranodal lymphomas (6). Lymphomas can affect both bony and soft tissue of the oral cavity, with the most frequent localization being the tonsil. The most common type is diffuse large B-cell lymphoma, but mantle cell lymphoma, marginal zone B-cell lymphoma, Burkitt’s lymphoma, lymphomablastic lymphoma, peripheral T-cell lymphoma, and anaplastic large cell lymphoma have also been reported in the oral cavity (7). However, B-LBL arising from the hard palate has not been reported previously.

Clinical manifestations depend on the location of the lesion. The most common clinical appearance of non-Hodgkin lymphoma in the mouth is a nonhealing, painless ulceration (8). Patients may complain of localized or diffuse soft tissue swelling, pain, mucosal discoloration, paresthesias, anesthesia, and loosening of teeth (9).

Lymphoblastic lymphomas are treated similar to acute lymphoblastic leukemia with combination chemotherapy protocols consisting of intensive remission-induction chemotherapy, central nervous system prophylaxis, a consolidation chemotherapy, and subsequent maintenance therapy. Treatment of LBL with protocols derived from acute lymphoblastic leukemia therapy are effective, with an 82% event-free survival and an 85% overall survival at 5 years (10).