Richter's Transformation in an Unusual Site: Non-Hodgkin Lymphoma in the Parotid Region

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To the editor,

A 73-year-old female patient, a known diabetic, presented to our tertiary care center with swelling on the right side of the face for the past 1 month, insidious in onset and gradually increasing in size. The patient had a past history of chronic lymphocytic leukemia (CLL) that was diagnosed at an outside hospital 3 years ago, for which she was taking treatment comprising of tablet chlorambucil. Further details of initial diagnosis were not known. Her Eastern Cooperative Oncology Group performance status indicated a score of 0 (able to perform all daily activities). Upon hematological investigations, she had a hemoglobin of 13.6 g%, total count of 56,800 cells/cu.mm, platelet counts was 3.29 lakhs, and serum lactate dehydrogenase level was 163 U/L (normal: 208–370 U/L). Uric acid and aspartate aminotransferase levels were low. Bone marrow biopsy was not performed. Computed tomography and positron emission tomography showed a fluorodeoxyglucose (FDG)-avid mass lesion with mandible erosion in the right parotid with a maximum standardized uptake value of 11.8 (►**Fig. 1A, B**). The scan also showed

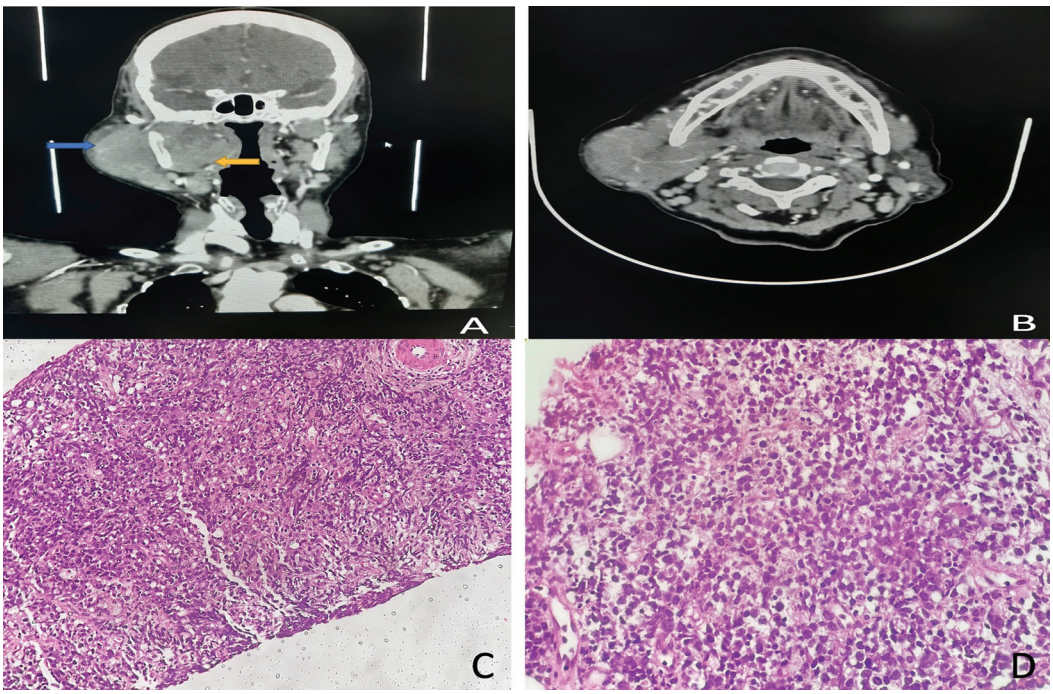

Fig. 1 (**A** and **B**) Computed tomography of the neck with contrast showing an ill-defined homogeneously enhancing soft tissue density involving the superficial lobe (blue arrow) and deep lobe (yellow arrow) of the right parotid gland with extensions. (**C** and **D**) Hematoxylin and eosin-stained sections in 20× (**C**) and 40× (**D**) magnifications, respectively, showing infiltration by sheets of atypical large lymphoid cells, with the presence of brisk mitosis and apoptosis.

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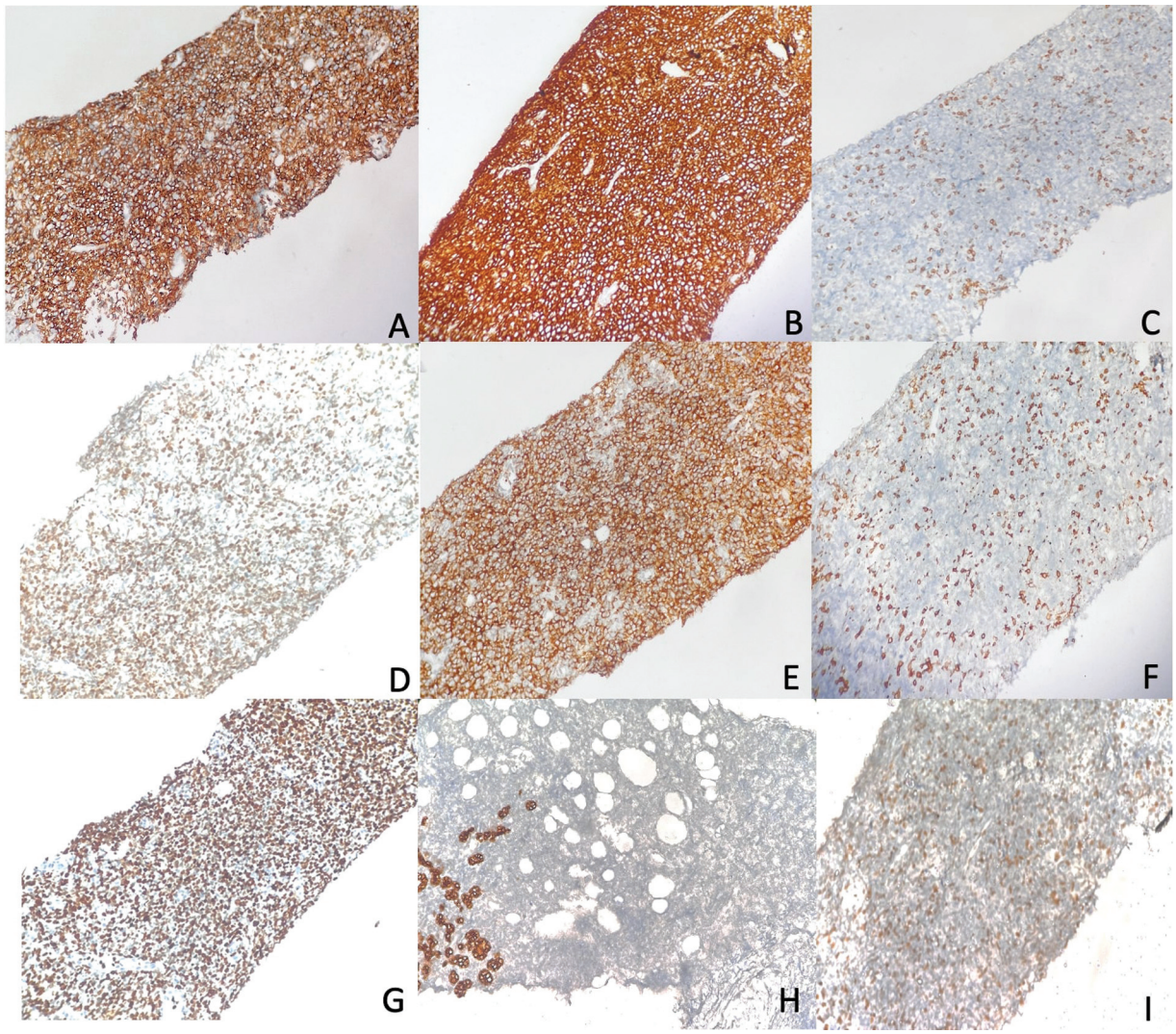

Fig. 2 Immunohistochemical findings in the tumor cells in 20 × : (**A**) CD45 positive in the tumor, (**B**) CD20 diffusely positive in the tumor cells, (**C**) CD3 scattered positivity noted, (**D**) Bcl6 positive, (**E**) Bcl2 diffusely positive, (**F**) CD5 positive in scattered cells, (**G**) Ki67 80% in the tumor cells, (**H**) cytokeratin positive in entrapped glands and negative in tumor cells, and (**I**) C-myc focally positive.

multiple mildly prominent and mildly enlarged lymph nodes in the neck and bilateral axilla, with a maximum size of 15 mm on the right and 17 mm on the left and pelvis with no minimal FDG uptake. An image-guided accurate biopsy from the right parotid gland was performed.

Grossly, we received six linear cores of soft tissue, the largest measuring 0.5 cm and the smallest measuring 0.2 cm. Upon microscopic examination, sections showed diffuse infiltrating lesion composed of sheets of large cells with scant cytoplasm, increased nucleocytoplasmic ratio, nuclear pleomorphism, and coarse chromatin. Brisk mitotic figures and apoptotic bodies were also seen (►**Fig. 1C, D**).

Immunohistochemistry (IHC) performed showed diffuse positivity for CD45, CD20, BCL6, and BCL2 and focally for CD5 in the large cells, while they were negative for CD3, CD10, CK7, cyclin D, and MUM1. Ki67 labeling index was 80% (►**Fig. 2A–I**). The IHC findings confirmed the diagnosis of a diffuse large B cell lymphoma (DLBCL) germinal center type, involving the parotid gland, establishing a Richter's transformation from CLL. Following this, she was again started on rituximab-containing chemotherapy that also included injection doxorubicin, vincristine, and cyclophosphamide. However, she finally succumbed to the disease 3 months later.

Richter's transformation refers to a phenomenon in which CLL or small lymphocytic lymphoma transforms into a more aggressive form of lymphoma, usually DLBCL, with incidence rates of 2 to 10%.[1] The exact cause has yet to be understood, but mutations such as *TP53*, *c-Myc*, and *NOTCH1* have been documented.[2,3] Richter transformation occurs in salivary glands such as the submandibular gland; however, it is extremely rare to occur in the parotid gland.[4] The prognosis for such cases is generally less favorable, and treatment is challenging. Hence, a high index of suspicion is crucial for a timely diagnosis and to plan appropriate treatment. Recent therapies such as chimeric antigen receptor T-cell therapy and treatment with monoclonal antibodies have demonstrated encouraging results and are in the clinical trial stage.[5] This case was presented

due to its rare location and distinct clinicopathological implication.

Authors' Contributions

J.J. was involved in literature search, data acquisition, data analysis, manuscript preparation. A.B. was involved with the concept, design, definition of intellectual content, clinical study, manuscript editing, review and guarantor. L.D. and N.R. were involved with definition of intellectual content, clinical study, manuscript review and editing.

Patient Consent

Patient consent is not required due to the retrospective nature of the study.

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Conflict of Interest

None declared.

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