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ELEVENTH ANNUAL MEETING

Parotid Gland Extramedullary Hairy Cell Leukemia: A Case Report With Review of Literature

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INTRODUCTION

Hairy cell leukemia (HCL) is rare hematologic malignancy with an extremely rare manifestation of extramedullary involvement, which can occur with or without bone marrow infiltration. We report a 42-year-old man with extramedullary HCL in the right parotid gland without cytopenia, bone marrow infiltration, or splenic involvement.

CASE REPORT

We report a 42-year-old man with extramedullary HCL in the right parotid gland without cytopenia, bone marrow infiltration, or splenic involvement.

Core biopsy showed infiltration by small to medium lymphoid cells with oval nuclei containing homogenous chromatin and inconspicuous nuclei with distinct borders; the surrounding cytoplasm was clear and abundant.

Immunohistochemistry was positive for leukocyte common antigen (LCA), cluster of differentiation (CD)-10, CD20, CD25, B-cell lymphoma (BCL)-2, BCL6, and cyclin D1; negative for CD3 and CD5; with a Ki-67 score of 10% to 15%. Cyclin D1 positivity is also seen in mantle cell lymphoma. In our patient, molecular analysis showed ***BRAF* V600E** mutation with a frequency of 34.2%, a more sensitive and specific marker for HCL.

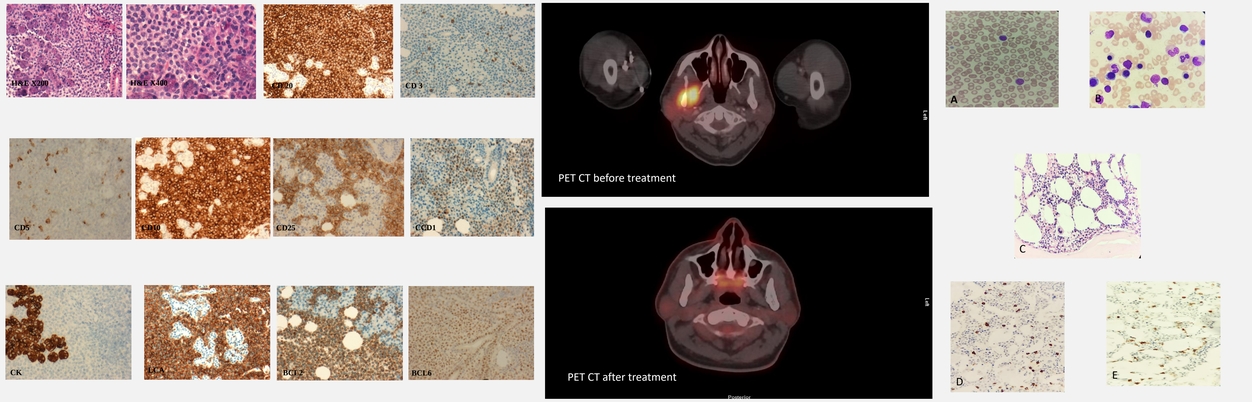
Positron emission computed tomography (PET CT) showed a soft tissue mass (4x2.7 cm) in the superficial lobe of the right parotid gland (maximum standardized uptake value [SUVmax], 7); a metabolically active, ill-defined soft tissue mass (4.5x4 cm) in the right suprazygomatic masticator space, infiltrating the lateral pterygoid and temporalis with erosions in the mandibular condyle (SUVmax, 9.5); and metabolically active foci in both femurs, the lower sacrum, the L1 vertebra, and the left third rib (SUVmax, 8)

TREATMENT

He received 5 days of cladribine and 8 doses of weekly rituximab. Extramedullary involvement does not warrant special treatment, except local radiation in select cases for symptomatic and larger lesions; our patient did not require local radiation. Post therapy, he had very good clinical and radiologic improvement with complete resolution of the parotid gland lesions.

Review of Literature

	Age/Gender	Extramedullary site	BM involvement	Treatment	outcome
1	39/M	Parotid gland	No	Surgery and Radiation	CR
2	49/F	Breast	No	Watch and wait	-
3	40/F	Right submandibular gland	No	Cladribine	CR
4	48/M	Duodenum	Yes	Cladribine	CR
Current case	42/M	Parotid gland	No	Cladribine and Rituximab	CR



Immunohistochemistry was positive for CD10, CD20, CD25, B-cell lymphoma (BCL)-2, BCL6, and cyclin D1; negative for CD3 and CD5; with a Ki-67 score of 10% to 15%. PET CT scan showing FDG uptake in parotid gland, before and after treatment.

Peripheral blood smear(A) showing no abnormal lymphocytes or hairy cells , Bone marrow aspiration smear(B), showing myeloid and erythroid precursors in various stages of maturation, small mature lymphocytes, BM biopsy(C),Immunohistochemistry of bone marrow trephine biopsy of same case, show few scattered small lymphocytes mixed CD20+(D) and CD3+(E)

CONCLUSIONS

To our knowledge, this is one of the few reported cases of salivary gland involvement by HCL. Patient well responded to Cladribine +Rituximab therapy.. Extramedullary involvement most often doesn't need special treatment, except in few cases local radiation may be required.

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