CR19

A rare presentation of Langerhans cell sarcoma in the parotid gland

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Keywords: Langerhans cell sarcoma, radiotherapy, rare tumor, salivary glands

INTRODUCTION/OBJECTIVES: Langerhans cell sarcoma (LCS) is a very rare, high-grade malignancy of the Langerhans cells and may vary from isolated lesion to multifocal disease. Parotid gland involvement is an exceptionally rare clinical presentation of LCS even in patients with multifocal disease.

CASE PRESENTATION: We report a case of LCS in the left parotid gland that occurred in a 63-year-old male. The patient presented with a palpable mass on the left side of the face. Ultrasonography showed three hypoechoic nodules in the left parotid gland. The patient underwent parotidectomy and selective neck dissection levels II-III. Immunohistochemical studies of the tumor tissue showed marked positivity to S100 and CD68, and focally positive CD1A staining. The presence of Birbeck granules on electron microscopy couldn't be reliably established. With 20 mitoses per 10 HPFs, tumor tissue exhibited high proliferative activity. Tumor was confined to the parotid gland with no involvement of adjacent lymph nodes. On the initial assessment, there were no signs of systemic dissemination. Four months post parotidectomy, PET-CT revealed focally increased metabolism in the soft tissue nodule located along the posterior edge of the gonial angle on the left side protruding medially, which raised suspicion of disease recurrence. Ultrasonically guided FNA did not show the presence of malignant cells. Surgery was followed by adjuvant radiotherapy 66 Gy in 33 fractions. There has been no evidence of recurrence or disease progression over the past 14 months since diagnosis.

CONCLUSION: LCS should be kept in mind in the differential diagnosis in patients with parotid enlargement.

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ACUTE LIVER FAILURE IN SECONDARY HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS AFTER COVID – 19 VACCINATION

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Keywords: adult multisystem inflammatory syndrome, acute liver failure, covid-19 vaccination, hemophagocytic lymphohistiocytosis

INTRODUCTION/OBJECTIVES: Hemophagocytic lymphohistiocytosis (HLH) is hyperinflammatory hyperferritinemic syndrome. Secondary HLH is known in adults and can be triggered by infections, malignancies or autoimmune response. If not recognized, HLH leads to multiorgan failure and death. CASE PRESENTATION: A 61-year-old male came to emergency room complaining of continuous high fever which started 10 days after COVID-19 vaccination. Patient was prescribed with co-amoxiclav. 14 days later, fever still not diminishing, patient was admitted to hospital. He tested negative for SARS-CoV-2 twice. Despite empirical antibiotic treatment, fever persisted. MSCT showed hepatosplenomegaly. Based on high fever, hepatosplenomegaly and hyperferritinemia, hemophagocytic syndrome was suspected. After administrating dexamethasone, fever decreased. 7 days post-hospitalization, coagulation deficit(PT 2.71), high transaminases(AST 3658 IU/L, ALT 4494 IU/L), jaundice(bilirubin 269µmol/L) and stage III portal encephalopathy indicated acute liver failure. Patient was transferred to a larger medical center for high urgency liver transplantation as trans-jugular liver biopsy confirmed sub-massive liver necrosis. Criteria for high urgency transplantation were not met because diagnostic tests confirmed secondary hemophagocytic lymphohistiocytosis probably as a part of adult multisystem inflammatory syndrome following vaccination. Patient complied to 6 out of 8 HLH-2004 diagnostic criteria - high fever, hepatosplenomegaly, he-

CONCLUSION: HLH may result in secondary acute liver failure, but emergency transplantation is not a treatment option.