MULTIFOCAL LANGERHANS CELL HISTIOCYTOSIS WITH RARE SUBMANDIBULAR GLAND INVOLVEMENT

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INTRODUCTION

- Langerhans cell histocytosis (LCH) is a lymphoproliferative disorder characterized by proliferation of langerhans type of dendritic cells.Histopathologically, a lesion is formed by collections of pathologic Langerhans cells with eosinophils, macrophages and lymphocytes.
- Common sites involved are skin, lymph node, bone and lung and liver.
- LCH can present as unifocal, multifocal or disseminated disease based on extent of lesions and chronicity of disease. Unifocal LCH presents with solitary eosinophilic granuloma. Multifocal LCH is usually chronic with multiple lesions. Disseminated LCH shows diffuse involvement of multiple organs and has poor prognosis.
- Investigations used are ultrasonography, PET-CT scan,MRI etc.
- Diagnosis is done by biopsy and histopathological study of the lesion.
- Treatment includes surgical excision followed by , chemotherapy, corticosteroids, low dose irradiation. High-dose chemotherapy and radiation followed by bone marrow transplantation is reported to be successful in advanced cases.

CASE REPORT

- A 20 year old male patient presents to opd with complaints of swelling in left side of neck since past 2 months, which was insidious in onset and gradually progressive in size. No h/o pain, fever, URTI, difficulty in swallowing, hypersalivation. On examination, a single swelling was noted, 1x2 cm size in left cervical region (level Ib). It had smooth surface, firm consistency, mobile.
- Two years back he came with similar complaints of right sided neck swelling duration 2 months, insidious in onset and progressive. Associated with pain radiating to right ear. No other associated complaints. Excisional biopsy of right submandibular gland revealed features indicative of langerhans cell histiocytosis.

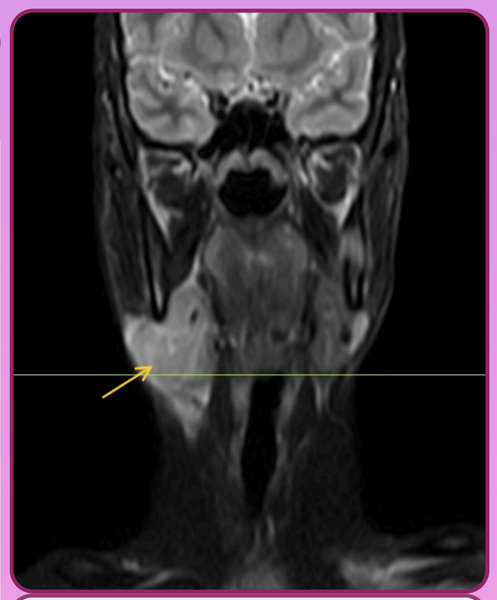

Fig 1: Pre-operative MRI of neck shows enlarged right submandibular gland (45x34mm) with heterogenous enhancement

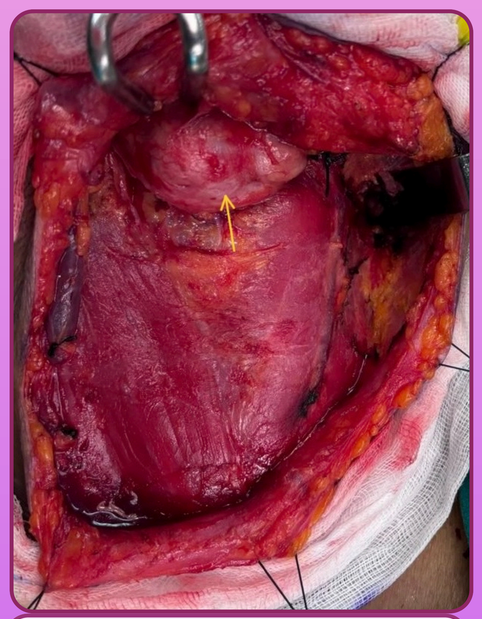

Fig 2: Intra-operative image from right submandibular gland excision surgery

MANAGEMENT

- Upon first admission, the affected right submandibular gland was excised. PET-CT scan done after surgery showed lytic lesions in mandible and maxilla and D12 vertebra. Diagnosis- MULTIFOCAL LANGERHANS CELLS HISTIOCYTOSIS. The patient was started on chemotherapy containing Vinblastine (9mg once weekly) along with oral steroids. The patient was lost to follow up after 9 cycles of chemotherapy.
- Two years later, when patient presented with left submandibular swelling, ultrasonography of neck showed hypoechoic lesion in left submandibular gland and bilateral level IB and II lymph nodes. Core biopsy of left submandibular gland confirmed lesion of Langerhans cell histiocytosis. The left submandibular gland was excised. PET-CT scan done after surgery showed no significant changes in existing osteolytic lesions in maxilla, mandible and vertebra. Chemotherapy was initiated again- Vinblastine 6mg weekly for 6 weeks, Etoposide 50mg for 21 days with Prednisolone 60mg daily (tapered dose). Patient was counselled about need of compliance to treatment and possibility of relapse if non- compliant.

DISCUSSION

Langerhans cell histiocytosis is a rare disease most often diagnosed between the ages of 1 and 3. It most commonly involves bone, skin, lungs, and pituitary gland.
In this case, patient was 18 yr old when multifocal LCH was diagnosed, involving submandibular gland alongwith osteolytic lesions in mandible and vertebra.
Submandibular gland involvement is an extremely rare presentation of the disease.
Treatment modalities used in this case are surgical excision followed by chemotherapy. It also shows relapse of disease due to non-compliance by the patient (discontinuation to chemotherapy).

CONCLUSION

Involvement of submandibular salivary gland in LCH is extremely rare with only a handful of reported cases. As it is a rare disease, there is limited medical literature available. This case presents an opportunity to study the disease and how it responds to available treatment modalities.
The case also highlights the importance of compliance to treatment in patients suffering from chronic diseases requiring lifelong medication.