Case Report

Langerhans Cell Histiocytosis (LCH) and Diabetes Insipidus with Mandibular lesion

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Abstract: Langerhans cell histicytosis (LCH) is a rare disorder that primarily affects children. Its occurrence in adult is very rare. We report a case of 42 year old female patient who presented polyuria and polydipsia, loosing teeth and diplopia added to symptom .The diagnostic workup revealed endocrine involvement with diabetes insipidus. The x ray Orthopantomogram (OPG) showed destructive bone lesion of mandible. Biopsy of lesion revealed histiocytosis X. We herein describe the case report of Langerhans cell histiocytosis on mandible and involvement endocrine system.

Bibliographic Information of this article:
[Sayeh Alizad jahani, Somaye Khosravi, Mohammad Reza Mohajeri Tehrani, Bgher Larijani. **Langerhans Cell Histiocytosis (LCH) and Diabetes Insipidus with Mandibular lesion**. Electronic Physician, 2012;4(2):516-519. Available at: http://www.ephysician.ir/2012/516-519.pdf]. (ISSN: 2008-5842). http://www.ephysician.ir

Keywords: Langerhans cell histicytosis (LCH); Diabetes insipidus (DI); Mandibular lesion

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1. Introduction

Langerhans cell histiocytosis (LCH) is a rare disorder characterized by the excessive proliferation of Langerhans cells in various tissues. LCH places in the mononuclear phagocyte system and can infiltrate almost any place of the body. LCH be presented as a localized lesion or extensive systemic disease (1–3). The clinical manifestation of LCH is extremely, Patient may suffer single system or multisystem involvement. Presenting symptoms in order of frequency are skin rash, dyspnea, polydipsia and polyuria (diabetes insipidus/DI), bone pain, lymphadenopathy, gingival ulcer, and memory disorders. The incidence of LCH appears to be 3 to 5 per million children and in adult it is estimated 1 to 2 cases of LCH occur per million populations (4). The etiology of LCH is unknown; some studies suggested that the etiology may be related to immunological abnormalities resulting from a suppressor cell deficiency (5-6) and other efforts to define a viral etiology have not been successful (6-7). Dibetes insipidus (DI) is the most common endocrine abnormality in LCH; 4 % of LCH may present with an apparent idiopathic presentation of DI previous to other lesions are recognized (8). Of 47 adult patients in one study, the primary sites of bone involvement were Jaw (30 %), skull (21 %), vertebral (13 %) (9). Pain and bony swelling are the most commonly presenting complaint. Intraoral findings include gingival ulcer (50%), loosening teeth (17%) and premature exfoliation of the teeth, precocious eruption of permanent dentition, ectopic eruption of permanent molars and halitosis (5, 10, and 11). Histological confirmation is necessary for diagnosis. Because endocrinology manifestation in LCH relatively is rare, we report a case of an Iranian adult patient suffering from LCH on jaw and DI.

2. Case Presentation

The patient was a 42 years old housewife presented with polyoria, polydipsia and nocturia since 14 years ago. Central DI (Diabetes insipidus) was treated with nasal spray desmopressin acetate (DDAVP) 2puf /daily and symptom temporarily resolved .Two year later treatment patient developed amenorrhea and consequently presented with loos mandibular teeth which subsequently extracted.

The X-Ray (OPG) examination (Figure1) and CTscan (Figure2), showed destructive bone lesion of middle mandible .family history were unremarkable.last pregnancy of LCH patient was terminated by cesarean section 14

years ago she did not experience unusual bleeding. Three months after delivery she was able to breast feed. Seven years after first presentation the patient was suffer from Diabetes Mellitus that was treated with hypoglacemic oral agents. Diplopia gradually added to the symptoms.

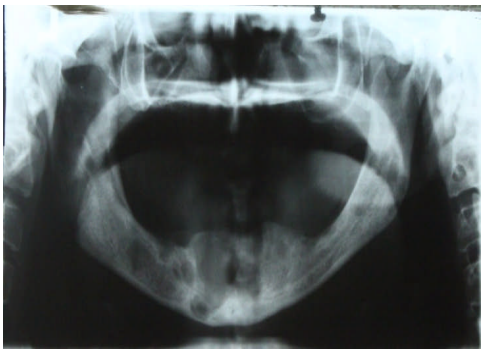

Figure 1. The X-Ray (OPG): (showed destructive bone lesion of middle mandible

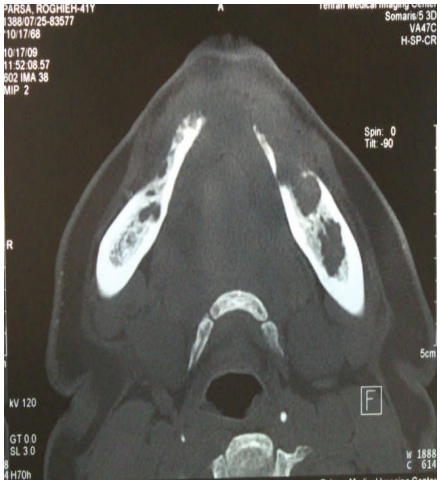

Figure 2. CTscan Axial view. Lytic lesion in left mandible and lytic lesion in medial ramus of mandible

In Physical examination, the superficial lymph nodes were not palpable and no hepatosplenomegaly was noted. There were no abnormalities of cardiac or pulmonary function, Neurological examination at that time, revealed diplopia and bitemporal visual field defects. Hematological data including Complete Blood Count, Electrolytes ,Blood Urea, Nitrogen, Creatinin Erythrocyte Sedemination Rate were whitin normal limits , Fasting blood sugar:115mg/dl, HbA_1C:5.5 (4/4_6/7), Endocrinological test revealed the following values: T4 [Tetroiodothyronine: 7.1µ gr/dl (4/6-12/4)], TSH [thyroid stimulating hormone 0.3mIU/ml (0/1-5)], T3 [Triiodothyronine: 113ng/100ml (78-182)], Prolactin 9.4ng/ml (2-13), MorningCortisolin: 5µgr/dl (9/4-26), Luteiniing hormone 0.5 mIU/ml (13/5-96), Follicle stimulating hormone1.1 mIU/ml (27_129), Stradiol: 27Pmol/ml.

As mentioned in OPG showed a destructive bone lesion on middle mandible and 2 lesions on left ramus. After biopsy the lesion was resected totally by a maxillofacial surgeon. Biopsy of lesions demonstrated presence of proliferation histiocytic cell and positive for S100 proteined CD_{19} indicated Langerhans cell histiocytosis (histiocytosis X). Bone scan showed increase activity in mandible due to infiltration lesion, focal lesion in distal part of right femur, suspicious lesion in the right parietal bone. Brain MRI with contrast showed: enhancement lesion in suprasellar 10×5mm which is attached a chiasma that can be associated with histiocytosis (Figure 3).

Prednisolone (PSL) gradually was started and tapered and then12 cycles of chemotherapy with vinblastine were given every 3 weeks for two days. During chemotherapy proteinuria occurred based on consultation with the

nephrology service and according to Renal Ultra Sonography which was normal and membranoglomerulopathy was reported in biopsy, previous treatment to continue were recommended. After completing chemotherapy, she underwent radiotherapy for 12 sessions. Follow-up 6 months after radiothrapy showed no recurrence or evidence of systemic involvement.

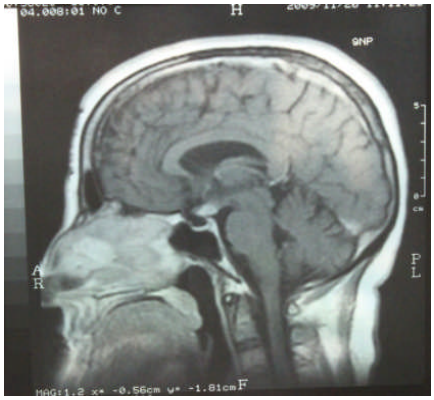

Figure 3. Brain MRI inhacment lesion in suprasellar 10×5mm which is attached a chiasma that can be associated with histiocytosis

3. Discussions

LCH is very rare and multifaceted that includes Letter-Siwe disease, Hand-Schuller-Christian disease and eosinophilic granuloma (EG). LCH may be systemic or localized the pathogenesis and etiology of LCH remains as uncertain. Finding positive markers in lesional cells, e.g S-100 Makes definitive diagnosis of the disease. LCH endocrine manifestation are involvement hypothalamus, Pituitary, thyroid gland, pancreas. It shows exacting tendency for involvement of the hypothalamo–pituitary axis (HPA), leading to diabetes insipidus (DI) and/or anterior pituitary dysfunction. DI is most common endocrine involvement; incidence is approximately 50% (15). DI can predict the presence of LCH and is usually constant and does not react to some obtainable treatment (16). Most frequent anterior pituitary hormone deficiency is Growth hormone (GH) (17). The second most ordinary anterior pituitary hormone deficiency in adults with LCH is Gonadotropin deficiency. Disorder in levels Adrenocorticotropin (ACTH) is reported in 1–2% of LCH patients, Prolactin (PRL) levels differ, and fairly Elevated (18). Hypothalamus involvement in LCH show to neurological and neuropsychological disorders such as disorder of appetite, sleeping pattern, behavioral, skills, body temperature regulation and memory impairment Thyroid and pancrass involvement is uncommon. Bone lesion may be silent in some sites but in mouth are specially trouble because of tooth loose mandible lesion tend to devastate alveolar bone. Prognosis of LCH depends on age of patient extent of disease and presence of vital organ failure.

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