Hand Schuller Christian Disease: A Case Report with Oral Manifestation

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Abstract: Langerhan’s cell histiocytosis is a group of diseases that result from langerhan’s cells or their precursors abnormal proliferation. The clinical manifestations are the result of infiltration and accumulation of the langerhan’s cells in tissues. We report a case of a three years’ boy with osteolytic lesions involving occipital, maxillary and mandibular bone. Additional periodontal tissues destruction was seen involving all four quadrants. Diagnosis was confirmed radiographically and histologically.

Keywords: Histiocytosis, Hand-schuller-christian disease, periodontal disease

1. Introduction

Langerhan cell histiocytosis (LCH) is manifested by proliferation mononuclear cells and prominent eosinophil infiltrate resulting in destruction of hard and soft tissues (1). Chronic disseminated form (Hand-Schuller-Christian disease) frequently seen with multiple osseous lesions and with extra skeletal lesions. Classical triad of Hand-Schuller-Christian disease is lytic bone lesions, exophthalmos, and diabetes insipidus.

Chronic localized form (Eosinophilic granuloma) characterized by only unifocal or multifocal bone lesions mainly seen in adult (3).

Here, we report a case of chronic disseminated Langerhan cell disease (Hand-Schüller-Christian disease) presenting in a three years old patient.

2. Case Report

A three years old boy accompanied by his father attended the National Ribat University, Pediatric dentistry department, in September 2014, his father reported that his son has a history of gradual increase in teeth mobility started two months ago.

On extra oral examination, the patient had brachycephalic skull, slight hypertelorism, exophthalmic left eye and submandibular lymphadenopathy.

The pathogenesis of LCH is unknown, however, various theories have suggested contribution of immunologic dysregulations, environment, infections, genetics, and neoplastic process (2).

LCH may affect any organ, although the lymph nodes, liver, and spleen are involved in most cases (1).

Langerhan cell disease manifests in three forms the acute disseminated form, chronic disseminated form and the chronic localized form.

The acute disseminated form (Letterer-Siwe disease) has a multiple rapidly progressive system involvement and often occurring mainly in infants.

The lower right mandibular molars had grade III mobility.

A full blood count was performed along with blood glucose levels to eliminate underlying systemic disease. The patient has a microcytic hypochromic blood picture and thrombocytosis.

CT showed osteolytic lesions involving occipital, maxillary and mandibular bones.

Chest x-ray revealed no present pathology and the abdomen U/S showed a 1 CM left mid pole renal stone and multiple enlarged P/A lymph nodes.

The gingivae were painful, erythematous and enlarged.
The meninges were almost exposed to environment. Scintigraphy with Tc-99m was normal. The immunohistochemistry report of the teeth and attached soft tissue revealed sheets of histiocytes with abundant pale eosinophilic cytoplasm and vesicular nuclei with an indented “coffee bean” appearance accompanied by an admixture of eosinophils and neutrophils.

Bone marrow aspirate and trephine biopsy revealed that the bone marrow is involved by histiocytic disorder.

The DPT revealed generalized irregular bone destruction.

The Diagnosis of Hand Schuller Christian Disease was set.

The patient was put on .. times a day for ... pediatric onchologist chemo

3. Discussion

Hand Schuller Christian is seen in children and infants and rarely in adults. LCH peak incidence between 5 and 10 years of age (4).

The classical triad of Hand Schuller Christian disease – Lytic lesions, exophthalmos and diabetes insipidus is seen only in one-third of patients (5).

50% of cases reporting with diabetes insipidus and with the most common site involved is the skull (6).

Tenderness, swelling and facial asymmetry are usually seen when the facial bones are involved.

Other frequently involved bones are ribs, femur, vertebrae, and pelvis. Sometimes, popular lesions appear in the skin.

Usually, oral manifestations are usually the earliest signs, which include unpleasant taste, sore mouth, gingivitis, halitosis and loose teeth. Bone loss may mimic advanced periodontal disease. Scooped out bone loss starting typically at the mid root region, commonly affecting the posterior mandibular alveolus may present (4).

Alveolar involvement can be extensive and causes the teeth to appear as if they are “floating in air.”(7)

Multidisciplinary team approaches is utilized in the treatment of the disease.

Localized oral lesions is treated by surgical curettage or excision. Intra-lesional corticosteroids injection or low dose systemic oral corticosteroids. Multi-systemic disease needs systemic chemotherapy. Dental practitioner has a role in the management the oral manifestations of the disease. Surgical intervention has to be done once the growth of jaw is complete. Prosthetic management can be given to

Other therapies include supportive therapy, control of diabetic insipidus and surgery in specific indications.

4. Conclusion

Management of patients with Patients with Hand Schuller Christian disease generally requires a multidisciplinary approach. Early diagnosis and treatment of the disease will improve the life expectancy.

References