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Oral Pathology

Letterer Siwe Disease

Letterer Siwe disease now known as Langerhans Histiocytosis is describe as a rare reactive and proliferative disease of histiocytes with unknown etiology, it is characterized by excessive proliferation of histiocytes called Langerhans cells. LCH can be divided into 2 categories, non malignant disorder and malignant disorder. The malignant disorders include Letterer Siwe disease and variants of histiocytic lymphoma. It occurs mainly in the pediatric population from 1 to 15 years of age with the peak incidence from 2 to 4 years of age but a few cases in adults has also been reported. Then incidence is more common in males compared to females. This condition can target almost any aspect of the body. In 60 % of the cases it affects the head and neck. In the head and neck region it appears as a punched out lesions in the skull, maxilla, mandible, sternum and other flat bones and causes rapid resorption of the alveolar bone, leading to floating teeth appearance in the radiographs. In most cases this disease manifests initially in the oral cavity, with careful clinical, radiological examination and biopsy this disease can be diagnosed.

Base on the journal “Langerhans Histiocytosis in a Child – Diagnosed by Oral Manifestations” A 4-year-old female child reported to the Department of Pedodontics and Preventive Dentistry with a chief complaint of pain, swelling and burning sensation in the lower left back region of the mouth. An extra oral and intra oral examination was performed. The

patient had a erythematous macule located on the upper lip, face was asymmetry, and swelling in the left mandibular region was observe, area was diffuse and soft to palpate. Intra oral examination showed gingival recession and a nodular swelling in the lower left region. based on clinical finding, patient was then sent for radiographic investigation. Orthopantomograph revealed multiple radiolucent lesions in the mandible on the left side extending from the canine region to the ramus of the mandible. There was also marked alveolar bone loss showing floating teeth. Based on the clinical examination, radiographic findings and histopathological report a confirmatory diagnosis of Langerhan's Histiocytosis was made. Patient was referred to Kidwai Memorial Institute of Oncology, Bengaluru for chemotherapy and radiotherapy. After treatment Patient had a complete clinical remission.

Another case was reported involving a 3 year old boy. "Langerhans Cell Histiocytosis: A Case Report" according to this journal, A 3-years-old boy, who was admitted to Maharani Laxmi Bai Medical College, Jhansi present with 8 months fever, insidious, mild to moderate grade, and intermittent. Patient medical history revealed that he was diagnosed with pilmonary Koch's and prescribed anti-tubercular treatment 5 months prior to admission. After one month of taking the medication, patient started developing red color maculopapular rash on his skin. He also had an history of purulent ear discharge and gum swelling. During examination, patient had an oral ulcer located on the hard palate, hypopigmented maculopapular rashes over his back, chest and abdomen, there were also multiple depressions present on palpation. Skiagram and computed tomography revealed and confirmed multiple punched out lesions in frontal, and parietal bone. Skull X-ray shows multiple irregular osteolytic lesions, followed by a CT scan for confirmation. a chest X-ray was also perform from the posterior-anterior view revealing multiple cavitary

lesions. CT scan revealed honeycomb of lung parenchyma. After performing multiple of test with the help of radiographs and assessment finding, the patient was diagnosed with LCH with multiorgan involvement and organ failure as multisystem LCH with involvement of risk organs according to histiocytosis society. Treating a child with multiorgan involvement, . LCH III treatment protocol is currently the most common therapeutic strategy. The journal also stated that, As a result of recent therapeutic trials, it has been shown that the single best prognostic indicator is a patient's response to chemotherapy during the 6 weeks induction phase. Patients who respond to chemotherapy during the 1-6th week of therapy have a 88-91% survival rate, but survival rates drop to 17-34% in patients who fail to respond.

LCH is a rare and unpredictable disease, it's natural history vary from a rapidly, fatal progressive disease to spontanous resolution. As a dental hygienist its very important to become familiar with these diseases. Being able to identify a suspicious lesion during our assessment phase of care, follow by a biopsy could save a person life. Base on the journal "Langerhans Cell Histiocytosis: A Case Report" Letterer-Siwe disease has a high mortality rate. The prognosis in these patients depends on the patient's age, the extent of disease, and the degree of organ dysfunction. The mortality rate is 50% or higher. With Early diagnosis, patient has a better chance to survive.