

Letterer Siwe

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Letterer Siwe Disease affects about 1 in 200,000 people each year.(6) This paper will answer the following questions what is Letterer Siwe Disease? Who does it affect? How is this disease treated? What is the prognosis? And how does this relate to a dental hygiene setting. Letter Siwe Disease is a unique disease which affects the generation of the histiocytes and eosinophilic leukocytes.(2) This disease mainly affects infants.(5) This paper will focus on describing the background of the disease, the symptoms, treatment and how it affects the oral cavity. With the use of articles and textbook information will be used to explain all the information about Letterer Siwe.

Histiocytosis is the broad name for the group of syndromes that have abnormal number of production of histiocytes, the function of histiocytes are to fight infections.(6) The 3 major groups of histiocytoses are Langerhans Cell Histiocytosis (Histiocytosis X), Malignant Histiocytosis Syndrome (T-cell Lymphoma), and Non-Langerhans Cell Histiocytosis (Hemophagocytic Syndrome).(6) Letterer Siwe falls into the group of Langerhans Cell Histiocytosis (Histiocytosis X), this disease is the most common and serious of its group.(4) Histiocytoses is the multiplying of histiocytes, these cells are produced in the bone marrow.(4) Letterer Siwe is also known as Acute disseminated Hystiocytosis which it affects the skin, internal organs and bone marrow.(5) This disease has been known as a cancer-like condition, but researchers have begun to say that its an autoimmune disease, which means that the cells are attacking the body.(6)

Letterer Siwe disease is an aggressive disease which if not treated immediately it is fatal in a short time.(3) This disease affects children around age under 3 years old, some of the symptoms that they suffer are abdominal pain, bone pain, fever, jaundice, rash, seborrheic dermatitis of the scalp, swollen lymph glands, and weight loss.(3/5) Since this disease is aggressive it affects multiple organs of the body such as liver, lung, lymph nodes, skin, bone marrow, and bone. Certain manifestations are found with this disease like eczema, enlargement of the liver and the spleen, anemia, hemorrhages, enlargement of the lymph nodes, and severe bone loss. (3/4) In order to diagnose this disease biopsy of the skin, bone marrow biopsy is needed to check for the presence of langerhans cells, complete blood count, and skeletal survey.(6)

The treatment of Letterer Siwe depends on how severe the case is. Some of the medications given are Cyclophosphamide, Etoposide, Methotrexate, Vinblastine. Other treatments could be used such as radiation or surgery to treat bone lesions.(6/4) The prognosis of this disease is poor, due to how aggressive the disease is and how it affects multiple organs of the body. Patients that are under 2 years of age and have multiple involvement of organs with this disease, even with an aggressive treatment, still have a mortality greater than 50%.(4)

Patients that have Letterer siwe may have oral lesions as an early indication of this disease, there is a 77% incidence of oral lesion found with this disease.(3) The most common presentation is pain and bony swelling. Some of the intraoral findings are gingival necrosis, mucosal ulceration, loosening and premature exfoliation of the teeth, and halitosis.(2) Due to alveolar bone loss in a x-ray the teeth are surrounded by radiolucency and the term “floating teeth” is used to describe it. The excessive mobility turns out to premature loss of the teeth affected. (2,3)

Letterer Siwe is important to a dental hygienist since most of the patients that do have this disease start to develop oral lesions. The oral lesions are the first possible indications of this disease. That is why it is important to carefully do an intra and extra oral exam to all the patients even though the patients might look healthy. Along with the intra and extra oral exam the patients should also have x-rays done to check if the jaw bones are healthy. It is also important to always follow up with the patients condition and make sure the best treatment is given to each patient.

In conclusion Letterer Siwe is a rare but serious disease that affects children under the age of 2. It is also important for the dental hygienist to be aware of oral lesions that affect these patients that have this disease. In this paper the disease Letterer siwe was described including the background of the disease, the symptoms, treatment, how it affects the oral cavity and the importance in the dental hygiene setting. Articles and textbook information were used to explain all the information about Letterer Siwe.

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