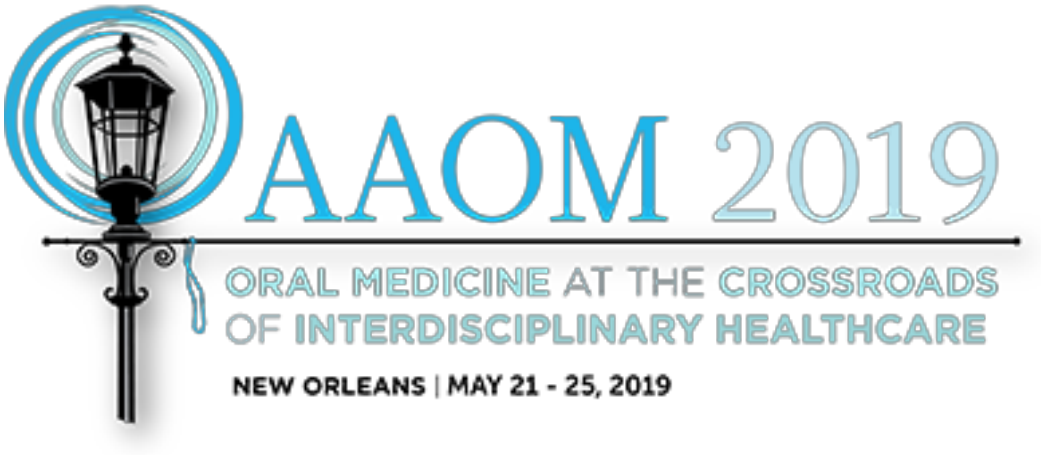


# Case Report Presentations
# Friday, 05/24/2018, 10:40am-11:40pm

# = Presentation Number, *Presenter/Awardee

To conserve space, we list only the institution and the country submitted as 1st organization.

Abstracts Committee:
Chair: Kentaro Ikeda, DDS, MPH
Co-Chair: Bhavik Desai, DMD, PhD

**10:40AM**
**#1:**

**A Rare Case of Non-Langerhans Cell Histiocytosis/ Erdheim-Chester Disease (ECD) Presenting As Desquamative Gingivitis and Oral Mucosal Erythema**

*Muhammad Ali Shazib, Hervé Yves Sroussi
Brigham and Women's Hospital and Dana-Farber Cancer Institute, USA

**Background:**

Erdheim-Chester Disease (ECD) is a rare non-langerhans cell histiocytosis characterized by BRAFV600E mutations in greater than 50% of patients, multi-systemic involvement such as skin (xanthelasma), heart (pericardial effusion), CNS (brain-stem lesions), and bone lesions. ECD has a distinct histopathology and immunochemistry profile which is negative for CD1a, S100, and Langerin. Craniofacial involvement has been reported in less than 10 cases and described as multiple solitary, well-defined radiopaque/ lucent lesions of the jaws. We report a case of ECD which initially included desquamative gingivitis, mucosal erythema, and edema.

**Case Summary:**

A pleasant 62-year-old Caucasian lady was referred from her local dentist to the Division of Oral Medicine and Dentistry, Brigham and Women's Hospital in December 2017 with a chief complain of mouth sensitivity. Her medical history was notable for a 7-year history of intermittent bone pain, non-pruritic erythematous and papular rashes of the neck, extremities and trunk, and gingival erythema. She was diagnosed by a hematologist-oncologist with ECD in December 2010 based in-part on a tibial biopsy and the presence of BRAFV600E mutation. Her medications included prednisone 40 mg QD by mouth, Bactrim QD by mouth, subcutaneous MTX 27.5 mg weekly injection, and dabrafenib 150 mg Q12H. She reported having a 10-year history of intermittent sensitivity, redness, and swelling of the gingiva without any known triggers or patterns. Upon oral examination, there was diffuse desquamation, erythema, and edema of the buccal and palatal gingiva as well as erythema of the mid hard palatal mucosa. Differential diagnosis were Erdheim-Chester Disease, vesiculobullous disorders, and vasculitis. With patient's consent, an incisional biopsy was obtained which revealed the presence of chronic inflammation and a scant histiocytic infiltrate. The patient was managed with topical clobetasol 0.05% via custom-trays to the maxilla and mandible and reported a greater than 75% improvement in sensitivity score after one month of use. Five months later, she presented with two erythematous nodules (#12 and 19) which were consistent with pyogenic granulomas on excisional biopsy.

**Conclusions:**

This is the first report of potential oral mucosal involvement of Erdheim-Chester Disease with the noted desquamative gingivitis and erythema as its oral presentation.

**#2:**

**Clinical Manifestations Leading to a Diagnosis of HIV Infection: A Case Report**

*Richard J. Vargo, Prashanth K. Haribabu
A.T. Still University - Missouri School of Dentistry & Oral Health, USA

**Background:**

Of the approximately 1.1 million people aged 13 years and older living with human immunodeficiency virus (HIV) infection in the United States, an estimated 162,500 are undiagnosed. Numerous clinical manifestations of HIV have been reported, but these have declined in prevalence since the advent of highly active antiretroviral therapy (HAART). Herein, we present a case of a previously undiagnosed patient presenting with multiple supportive clinical manifestations that led to a confirmed diagnosis of HIV.

**Case Summary:**

A 22-year-old male was referred to A.T. Still University - Missouri School of Dentistry & Oral Health with a chief complaint of a six-month history of painful gingivae. Intraoral examination revealed ulcerated, hemorrhagic, and necrotic gingivae with blunted papillae, associated bone loss, and fetid odor consistent with necrotizing ulcerative periodontitis (NUP). Extraoral examination revealed fingernail hyperpigmentation, cervical lymphadenopathy, and multiple smooth-surfaced, sessile, umbilicated papules of the mid-face area suggestive of molluscum contagiosum. A biopsy of the skin lesions confirmed the clinical impression. Despite the lack of reported high-risk behaviors, undiagnosed HIV infection was suspected given the combination of extra- and intraoral manifestations. The patient was prescribed chlorhexidine, metronidazole, and penicillin for NUP and advised to present to his physician that day for HIV testing. At follow-up, intraoral examination revealed pseudomembranous candidiasis but resolving NUP lesions. When presented with the suggestive clinical findings and upon further questioning regarding his sexual history, the patient reported unprotected sexual intercourse with an HIV-positive partner two years prior. Consequently, the patient was referred for rapid HIV testing at a local Infectious Diseases Clinic, and the results came back positive.

**Conclusions:**

This case demonstrates some of the various intraoral and extraoral presentations of HIV, including NUP, fingernail hyperpigmentation, cervical lymphadenopathy, molluscum contagiosum, and pseudomembranous candidiasis. Although the patient's chief complaint was painful gingivae, the presence of his other lesions led to a suspected and later confirmed diagnosis of HIV. Therefore, it is important that clinicians know the various manifestations of HIV infection, especially in patients who are undiagnosed. If present, the possibility of undiagnosed HIV infection should be suspected, and a referral for appropriate testing should be made.

**#3:**

**Primary Diffuse Large B-cell Lymphoma Of Mandible Masquerading As A Toothache**

*Ghaidaa Badabaan, Mahnaz Fatahzadeh
Rutgers School of Dental Medicine, USA

## Background:

Diffuse Large B-cell Lymphoma (DLBL) is an aggressive, fast growing form of non-Hodgkin's disease with rare manifestation in mandible as a primary site. Absence of pathogonomic features in this localization often leads to misdiagnosis as an odontogenic process or it's delayed diagnosis. We report a patient in whom non-odontogenic jaw pain mimicked a toothache prompting multiple dental interventions before persistence of pain and atypical findings led to consideration of a malignant etiology.

## Case Summary

A 54-year-old male presented to oral medicine service for dental clearance prior to radiation for mandibular lymphoma. He had presented to his dentist for evaluation of right-sided jaw pain a few months earlier. On presentation, he had no lymphadenopathy or paresthesia and his oral evaluation was WNL. Radiographic exam showed a deep restoration with pulpal proximity in tooth #31, widened PDL, loss of lamina dura and diffuse periapical rarefaction. Tooth # 31 was diagnosed with pulpitis and underwent RCT. After a brief period of quiescence, jaw pain returned and fractured cusp of tooth #30 bonded to address his symptoms. Despite these measures, jaw pain persisted and tooth #31 gradually developed extreme compressive mobility. Imaging revealed extensive alveolar bone loss, furcation involvement and floating molars. While extracting #31, a large osseous defect was visualized. This raised suspicion of a non-odontogenic etiology for patient's pain and prompted referral for bone biopsy. Microscopic examination of jaw specimen showed a tumor composed of neoplastic lymphocytes with large nuclei, prominent nucleoli and scattered mitotic figures. Immunohistochemical stains were positive for CD45, CD20, CD79a & BCL-6. PET-CT showed a solitary Lytic lesion measuring 3.5 cm in length with intense metabolic activity in right mandibular body. Oncology work up confirmed primary extra-nodal DLBL and patient was started on chemotherapy. Follow up imaging showed marked reduction in metabolic activity (3.8 compared to previous 44.4). He subsequently received low dose radiotherapy for residual disease in mandible and achieved remission.

## Conclusion

Although uncommon, dental providers should include malignancy including lymphoma in differential diagnosis of jaw pain, particularly when thorough evaluation fails to disclose a dental etiology, routine dental interventions fail to control symptoms or atypical clinical or radiographic findings develop.

**#4**

**Oral Cavity Malignant Lymphomas Are Exceptionally Rare, Highly Aggressive And Are Most Commonly A Sign Of Relapse – A Ten Year Retrospective Analysis In A Single Institution**

*Tal Berg, Sharon Akrish, Boaz Frenkel, Adi Rachmiel
Rambam Healthcare Campus, Israel

**Background:**

Malignant non – Hodgkin's lymphoma (NHL) represents approximately 5% of all malignancies of the head and neck and is the third most common after SCC and salivary gland tumors. NHL originates from B- or T-cell of lymphatic tissue. Only 24% of NHL affect extra-nodal locations. The head and neck is the second most common frequent site of extra-nodal manifestation. Only 0.1% to 5% present in the oral cavity of which the majority are Diffuse Large B Cell Lymphomas (DLBCL). Intra-oral cases usually appear as part of a progressive or recurrent disease when treatment failed. Primary NHL in the oral cavity are exceptionally rare. In our study, we conducted a retrospective clinical-pathologic analysis of all intra-oral soft tissue malignant lymphoma cases.

**Case Summary:**

Between 2008 – 2018, 11 cases of intra-oral soft tissue NHL were found and included extremely rare cases such as NK/T cell palatal lymphoma, Mantle cell lymphoma, Burkitt's lymphoma that transformed into DLBC and Mycosis Fungoides (Cutaneous T cell lymphoma) of the tongue. All patients were HIV negative. Ages ranging from 14 to 77 years old. Presentation of lesions was diverse and ranged from swelling mimicking dental abscess to deep ulcers mimicking SCC and even deep fungal infections. Thus, a fast and accurate differential diagnosis was of high importance. In some of the patients, oral cavity was the primary site of the disease and in part was a sign of relapse or part of disseminated illness. The most common location involved the hard palate. Over 90% of all intra-oral lymphomas were the first sign of relapse. 55% of these patients died within 6 months of oral presentation, therefore, early detection of intra-oral relapses is critical and highly important.

**Conclusions:**

Oral mucosal expression of NHL is rare and, in most cases, the first sign of relapse. Many types of NHL can be presented in oral soft tissues, most of them are fatal, so clinicians should take NHL into consideration when making their differential diagnosis: NHL lesions can mimic periodontal disease, acute abscess or even other malignancy.

**11:10AM**
**#5:**

**Nonspecific Oral Mucositis in Patient with Crohn Disease**

*Kristen Robbins, Dana Schwartz, Robert Greer, Kentaro Ikeda
University of Colorado, USA

**Background:**

A wide variety of oral lesions have been reported in Crohn's Disease (CD) with a reported prevalence of 20-50%. We report a case of lip swelling and severe mucositis affecting the lips and the oral mucosa in a patient with CD.

**Case Summary:**

A 48-year-old female presented with 2-year history of painful gingiva and lips. She had seen 26 doctors for this condition and had undergone multiple biopsies with nonspecific results, including a most recent perioral biopsy showing spongiotic dermatitis with neutrophils and eosinophils and negative direct immunofluorescence (DIF). Attempted treatments included chlorhexidine rinse, dexamethasone rinse, azathioprine, an increase infliximab dosage that she had been taking for CD, and prednisone. Among these treatments, only prednisone helped to reduce the severity of the symptoms. Past medical history included CD, celiac disease, and meningioma. Medications included infliximab every 8 weeks for 7 years, azathioprine, amitriptyline, sertraline, and pantoprazole. Her CD had been well controlled and multiple endoscopies/colonoscopies after the onset of the oral lesions confirmed no current CD lesions. Examination revealed swelling and generalized erythema with multiple ulcerations and blisters of the lips, generalized erythema on all facial gingiva and vestibules, and a slight hyperplastic appearance of anterior facial gingiva simulating orofacial granulomatosis. Prior biopsies were done on perioral cutaneous tissues. We performed a biopsy of oral mucosa. The result again indicated nonspecific inflammation with a negative DIF. The oral lesions have responded relatively well to systemic steroids.

**Conclusions:**

This case may demonstrate multiple conditions presenting simultaneously. Oral biopsy revealed a solitary focus of granulomatous inflammation which may be indicative of orofacial granulomatosis. However, the lesion biopsied was also unusual in that inflammation extended into the epithelium, a relatively unusual feature for granulomatous inflammatory disease and a pattern more commonly seen in vesiculobullous eruptions. Since this patient's CD has been well controlled, it is difficult to believe that these oral lesions are associated with CD activity. One reasonable assumption is that the treatment of the patient's CD for years with infliximab might have hindered what we could have seen histologically/immunologically in order to establish more definitive diagnosis.

**#6:**

**Multiple Myeloma masquerading as Medication Related Osteonecrosis of the Jaw (MRONJ): A Case Study**

*Yoon ah Danskin, Dalal Alhajji, SaeHee Yom, Cherry L. Estilo
Memorial Sloan Kettering Cancer Center, USA

**Background:**

Bisphosphonate such as zoledronic acid is commonly used to treat bone metastasis from conditions such as multiple myeloma or from solid tumors such as breast or prostate cancer. Medication-related osteonecrosis of the jaw (MRONJ) is one of the complications of bisphosphonate treatment. The reported incidence of MRONJ among cancer patients with bone metastasis treated with zoledronic acid is 1.3%. MRONJ has been reported to have an incidence rate of up to 18.5%. The clinical and radiographic presentation of MRONJ is very similar to the presentation of bone metastasis, making early diagnosis and timely treatment challenging. This is a case report of a patient with multiple myeloma with zoledronic acid history, whose initial presentation of an intraoral lesion made it challenging to diagnose between bone metastasis and MRONJ.

**Case Summary:**

This is a case report of a 67-year-old female with multiple myeloma with zoledronic acid history of 28 doses from 2013 to 2016, in remission. The patient initially presented with a one-week history of firm gingival swelling buccal and lingual to a fixed partial denture (FPD) in the left mandible. Panoramic radiograph showed a well-defined radiolucent lesion. Considering her antiresorptive history, MRONJ was considered as one of the diagnoses. However, due to the unusual hyperkeratotic nature of the buccal gingiva, relapsed disease was included in the differential diagnosis. Due to the unconventional clinical manifestation and concern for relapsed disease, CT guided bony biopsy was completed, which revealed plasma cell neoplasm and necrotic bone. The patient was treated with palliative radiation therapy and antibiotic, which decreased her symptoms significantly. Eventually, a portion of the FPD was removed, which revealed an exposed bony site with a mobile bony sequestrum. Once the sequestrum was removed, it led to complete gingival healing of the exposed site.

**Conclusions:**

Maxillofacial manifestation of bone metastasis is common but is often overlooked. Therefore, it should be considered in the differential diagnosis when a patient with antiresorptive history presents with gingival mass and/or exophytic bone. A good clinical judgement and a well-timed bony biopsy and diagnostic imaging can help to lead to correct diagnosis and optimal treatment.

**#7:**

**Localized CO2 Laser Treatment of a Recalcitrant Oral Ulceration in Pemphigus Vulgaris**

*Vinitha Gopal-Murthy, Nita Chainani-Wu
Oral Medicine Clinic, USA

**Background:**

Pemphigus vulgaris (PV) is a potentially fatal, autoimmune, vesiculobullous, mucocutaneous disease. Recalcitrant oral lesions of pemphigus vulgaris can result in significant oral discomfort and interfere with eating and oral hygiene. Increasing the dosage of systemic medications to control such lesions results in increased risk of side-effects.Currently there is 1 published report describing 2 patients with healing of oral recalcitrant lesions after carbon dioxide (CO2) laser vaporization.

**Case Summary:**

We describe a case of a 51-year old male patient diagnosed with PV by biopsy and direct immunofluorescence in 2013. At baseline, he had a history of premature ventricular contractions, allergy to penicillin, occasional alcohol intake, and no tobacco use. He was on analgesics for mouth pain. After further baseline laboratory testing, he was started on Prednisone and mycophenolate, with good response. These medications were slowly tapered over the next 2 years using clinical signs to guide the taper. At that time the mycophenolate was changed to IVIG monthly infusions due to adverse effects. During the 4.5 year follow-up period after diagnosis, while his systemic medications were being tapered, his oral and skin lesions were well-controlled other than minor transient flares. However, a painful, ulcerated lesion on the facial gingiva between #11 and 12 was non-responsive, even with use of topical clobetasol in trays. This recalcitrant lesion was vaporized with a CO2 laser (3W to 5.5 W, continuous mode) under local anesthesia. This procedure resulted in complete healing of the ulceration with no recurrence at the most recent 9-month postoperative examination.

**Conclusions:**

Adjunctive procedures that can facilitate a decrease in cumulative dosage of corticosteroids and immunosuppressants have great value in management of PV. CO2 laser vaporization is a safe procedure with minimal morbidity, and no long-term side-effects, and should be considered as an adjunctive treatment option for management of recalcitrant lesions in patients with oral PV.

**#8:**

**Saliva a Mirror to Oral Health in Neurofibromatosis type-1 Patients**

*Jayasurya Kondapaneni, Eshwar Thota, Nallan C.S.K Chaitanya, John Jims Veeravalli, Sourab Abhyankar, Sai Krishna Manchala, Humaira Samreen, Kavya Alluri

Panineeya Mahavidyalaya Institute of Dental Sciences and Research Center, India

**Background:**

Neurofibromatosis consists of complex group of syndromes which occurs due to inactivation of the various tumor suppressor genes precipitating in multiple cutaneous neuromas and schwannomas in the body. Neurofibromatosis type 1(NF1)/ Von-Recklinghausen's disease/ Watsons disease effects 1 in 30,000 individuals and occurs due to mutation in the tumor suppressor gene NF1 locus 17q11.2 and is one of the largest genes in the human genome. It is inherited in autosomal dominant pattern.

**Case Summary:**

Cross sectional study was conducted in a family suffering from neurofibromatosis over three generations, consisting of 20 members and the people meeting the diagnostic criteria for NF1 according to National Institutes of Health (NIH) (Stumpf et al.., 1988) were selected. Pedigree analysis, Clinical examination, Ramfjord periodontal index & Sialometry were performed for following parameters. Salivary flow rate- Whole unstimulated saliva was considered for salivary flow rate and is measured by asking the patient to spit out saliva in a sterile container and the amount of saliva is quantitatively measured. Salivary pH- Salivary pH of whole unstimulated saliva was measured by using PH-meter. Salivary alpha amylase activity - salivary alpha amylase activity measured by starch-saliva-iodine experiment.

**Conclusions:**

On clinical examination all the people in the family show a distinctive osseous lesion at the base of the tongue. Salivary analysis revealed decreased salivary flowrate and increased pH making the individuals prone to periodontitis due to decreased clearance and enzymatic activity. Alterations in the salivary glands (acini and ducts) is caused by mutations in the NF1 gene. Neurofibromin is known to play important role in salivary glands cell-division and differentiation via Rho-rock-LIM kinase-2 pathway, hence defect in Neurofibromin causes defective salivary gland cells differentiation causing non-functional cells making the individual prone to hyposalivation.