



**ADVANCES IN ORAL
MEDICINE** from Bench
to Bedside to Bench
AAOM 2018

Case Report Award Presentations Friday, 04/13/2018, 11:00am-12:00pm

= 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

#1: 11:00

Multifocal Granular Cell Tumors Occurring with Geographic Tongue

***Saeed Arem**, Richard Jordan, Caroline Shiboski, School of Dentistry
University of California San Francisco, USA

Background:

Granular cell tumors occur in the oral cavity and other sites including the skin and are characterized by the accumulation of plump cells with granular cytoplasm. Often appearing yellow clinically, these tumors are benign, but may recur. Here we report a case of multiple GCTs occurring on the dorsum of the tongue of a 59-year-old woman who also had geographic tongue.

Case summary:

A 59-year-female, referred for evaluation of three masses in the middle of the tongue. The lesions were unchanged since initial presentation two weeks earlier and no treatment had been rendered. The patient reported a burning sensation in the anterior half of tongue that preceded the lesions. Past medical, family, social, and dental history were unremarkable. Intraoral examination revealed three round, well-defined nodules with normal overlying mucosa located right of midline of the anterior portion of the tongue, firm to palpation and measuring each 1 x 1cm, 4 x 4mm, and 2 x 3mm, respectively. The patient's tongue also showed depapillated areas surrounded by white rims. Differential diagnoses for the nodules included GCT and lipoma. Diagnoses of geographic tongue and fungal overgrowth were also made. An antifungal was prescribed, and one week later excisional biopsies were performed of the masses. The tumor cells were S-100 and NKI-C3 positive but negative for MiTF confirming the diagnosis of GCT.

Conclusions:

GCTs are typically solitary asymptomatic yellowish nodules. This case is unusual because of the multifocal presentation, mucosal color, and burning pain, which was explained by concurrent geographic tongue.

#2: 11:12

Severe Oral Mucosal Hypersensitivity Reaction to Apixaban

*Meghan Gahm, Ayathi Apostolopoulos, Malik Altoos, Kentaro Ikeda

University of Colorado, USA

Background:

Allergic reactions to direct factor Xa inhibitors (DFXaI) are very rare and not well documented. Through PubMed search, we could not identify any reported case of oral hypersensitivity reaction to DFXaI. We present a case of oral mucosal sloughing associated with the use of Apixaban.

Case Summary:

A 72-year-old male presented with 8-months history of severe sloughing of the oral mucosa. This caused enough irritation to disturb his eating, resulting in a weight loss of 60 lbs in the eight months. He could not identify any aggravating or alleviating factors. Past management included fluconazole on two separate occasions without improvement. He was treated for an intestinal parasite up until one month prior to the onset of sloughing. After the failure of fluconazole trials, the oral lesions were thought to be related to possible recurrent parasitic infection and anti-parasite medication was recently reinstated, but it was ineffective. His past medical history included atrial fibrillation and intestinal parasite infection. His current medications included dofetilide, apixaban, florinal with codeine and ivermectin. There had been no reported change in the medications or oral products in the past 12 months. Examination revealed diffuse significant mucosal sloughing with underlying white lesions on all surfaces of buccal mucosa, labial mucosa, and gingiva. Biopsy was performed with results suggesting non-diagnostic to any specific disease and re-biopsy was suggested. He returned 5 weeks later with no sign of mucosal sloughing. Upon further questions, he reported that he tapered down apixaban over 2 weeks and subsequently discontinued 2 weeks prior. His mouth started to feel better soon after tapering down and completely healed 2 weeks after the discontinuation. He also admitted that he failed to report that the timing of starting apixaban was shortly before the onset of the mucosal sloughing. At this point, diagnosis of mucosal allergic reaction to apixaban was made with recommendation to use alternative future anticoagulation therapy.

Conclusions:

Due to the current trend in increasing use of DFXaI, health care providers will likely face more patients taking these medications. Awareness of these potential side-effects is crucial to diagnose and manage this type of case.

#3: 11:24

IgG4-related Disease Mimicking Extranodal Lymphoma of the Hard Palatal Mucosa:

A case report and review of the literature

***Alaa Bukhari**, Britta Magnuson, Mark Lerman

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Background:

Sicca syndrome is characterized by dryness of exocrine glands, in particular the lacrimal and salivary glands, and is a clinical hallmark of Sjögren syndrome (SS). Non-Hodgkin lymphoma (NHL) represents a complication of SS in nearly 5% of patients and the risk of lymphoma development in SS is 44-fold higher compared to the general population (Royer B. et al, 1997) However, the incidence of lymphoma in patients with Sicca syndrome in the absence of a definitive diagnosis of SS is not as well characterized.

Case Summary:

A 66-year-old female presented to Tufts University School of Dental Medicine with bilateral swelling of the parotid and submandibular glands. The patient had been followed by rheumatology with symptoms of SS, though this diagnosis had never been rendered definitively. She had a medical history significant for hypertension, atrial fibrillation, osteopenia, thyroid nodules, gastroesophageal reflux disease, dry eyes, and dry mouth. Her medications included lisinopril, metoprolol, dofetilide, rivaroxaban, cevimeline, calcium and multivitamins. CT scans in 2014 and 2017 revealed marked bilateral symmetric enlargement of the parotid and submandibular glands. On general examination, the patient was afebrile with no symptoms of night sweats or cough but with 8-pounds weight loss in past six months. Extraoral examination revealed firm swelling of parotid and submandibular glands. Intraoral examination revealed diffuse bilateral swelling of the hard palatal mucosa. The tissues exhibited red-purple coloration and a boggy consistency. There was no sign of bleeding or discharge. An incisional biopsy of the palatal swelling was performed. Histopathology revealed hyperkeratotic stratified squamous epithelium with overlying an atypical proliferation of lymphoid cells consistent with a B-cell marginal zone lymphoma. The patient was referred to hematology/oncology for further evaluation and management.

Conclusions:

Diagnostic criteria for SS have undergone many revisions over the years and this remains a challenging diagnosis to render, often involving providers in multiple specialties. This case underscores the importance of careful follow-up of patients with Sicca syndrome for development of lymphoma, even in the absence of a definitive diagnosis of SS.

#4 11:36

Chronic Mucocutaneous Candidiasis due to Gain-of-Function Mutation in STAT1

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Background:

Chronic mucocutaneous candidiasis (CMC) is a heterogeneous group of primary immunodeficiency diseases characterized by susceptibility to chronic or recurrent superficial *Candida* infection of skin, nails and mucous membranes. Gain-of-function mutations in the STAT1 gene (STAT1-GOF) are the most common reported cause of CMC. We describe the clinical and genetic findings for three patients with CMC due to novel gain-of-function mutations in the STAT1 gene (STAT1-GOF).

Case Summary:

Three cases are: A 56 year old Ugandan male with a heterozygous mutation c.850G>A (Glu284Lys) in exon 10. A 34 year old English female with a heterozygous mutation c.209G>C (Arg70Pro) in exon 4. A 66 year old Italian male with a heterozygous mutation c.2159C>T (Thr720Ole) in exon 24. All three cases presented to Oral Medicine with recurrent oral candidiasis and were subsequently referred to immunology for systematic investigation. The mainstays of treatment included suppression of *Candida* infection, using a combination of intermittent systemic therapy with long-term topical suppression and regular surveillance of the oral cavity to detect early dysplastic changes. The majority of mutations previously reported are confined to the STAT1 coiled-coil and DNA binding domains. In this series, two mutations identified lie outside of the typical regions, but altered function has been predicted in-silico and confirmed in-vitro.

Conclusions:

STAT1-GOF mutations are the most common genetic aetiology for CMC and mutation analysis should be considered in any patient with recurrent or chronic candidiasis. These mutations lead to defective responses in type 1 and type 17 helper T cells (Th1 and Th17) and are associated with a variety of phenotypes, which, depending on the mutation, predispose to candidiasis, fungal infections other than candidiasis, infection with *Staphylococci*, *Mycobacteria* & *Herpesviridae*, autoimmune disorders, as well as carcinomas and aneurysms. Prompt referral for immunological investigation allows early initiation of treatment and screening to identify specific genetic defects and reduces the significant morbidity and mortality associated with this condition.

#5: 11:48

**Oncogenic Osteomalacia Caused by a Phosphaturic Mesenchymal Tumor of the Mandible:
A Case Report**

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Background:

Tumor-induced osteomalacia (TIO) is a rare paraneoplastic syndrome in which patients present with hypophosphatemia and osteomalacia with clinical features that include bone pain, fractures and muscle weakness. The etiology is high levels of phosphate and vitamin D-regulating hormone, fibroblast growth factor 23 (FGF23). In TIO, FGF23 can be secreted by a variety of tumors, most commonly mesenchymal tumors that are typically small and difficult to locate. FGF23 acts primarily at the renal tubules and impairs phosphate resorption, leading to hypophosphatemia that inhibits the 1-alpha hydroxylase. To date, less than 310 cases of TIO have been reported and involvement of the mandible is exceptionally rare with only 8 reported cases.

Case Summary:

A 42-year-old male presented to our Institution with a 1-year history of pain in his ribs, hips, lower back, and feet. Radiologic examination revealed a decrease in bone density (spine T-score of - 4) and multiple pathologic fractures of the femur, ilium, and trochanter. Laboratory evaluation showed hypophosphatemia (1.6, normal 2.5 – 4.5 mg/dl), a tubular reabsorption of phosphate of 61% (normal 85-94%), and an elevated FGF23 (322, normal <180). Imaging and physical examination identified 3 potential sites: mandibular right first molar region; a cystic area in the left posterior cervical region; and a small nodule of right epididymis. A PET/CT (F-18 fluorodeoxyglucose) scan showed increased uptake in the mandibular right first molar region, where the patient had had a recent molar extraction. Intra-oral examination revealed no striking evidence of infection or mass. A CT scan showed an enhancing lytic expansile lesion. A deep bone biopsy was positive for mixed connective tissue tumor with spindle cell proliferation and scattered giant cells, with no identifiable malignant features. A surgical resection included a right segmental mandibulectomy, right neck dissection and tracheostomy followed by microvascular reconstruction. The final histopathology revealed no malignant features and completeness of the resection was confirmed by normalization of serum phosphate and an increase in serum 1,25 dihydroxy vitamin D3 from 27 (prior to surgery) to 366 pg/ml (normal 18-64 pg/ml) postoperatively.

Conclusions:

In conclusion, successful management of this debilitating condition required complete resection of the tumor followed by reconstructive surgery.