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Oral Case Presentations

OC 1
Oral malignant melanoma

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A 48 year-old Iranian female who complained of a palatal swelling that she first noticed 3 months prior to the dental school. Her past medical history is unremarkable. No history of tobacco smoker and alcohol intake was referred. Clinical examination of the oral cavity revealed an over-sized pigmented firm mass in the hard palate. The lack of any pain sensation was the reason for the delay of seeing a physician. The irregular pigmented mass was multilobular with ill-defined border. She reported that the lesion had rapidly increased in size over the past 3 months. During neck palpation a 2 × 1 cm firm mobile non-tender mass was palpated at the submandibular region. A maxillary occlusal radiography and biopsy was performed. A maxillary occlusal and chest X-ray was normal. Histologic examination of the specimen demonstrated extensive infiltration of the mucosa by neoplastic predominantly epitheloid cells and atypical melanocytes. The cells were round to oval, having eosinophilic nucleoli and actively producing and relasing melanin pigment and atypical mitotic figures. They were negative S-100 protein antibody. The diagnosis was oral melanoma and in this case differential diagnosis of melanoma includes, nevi, Kaposi's sarcoma and many other conditions sharing some clinical and microscopic characteristics. Our patient cxcluded any option of invasive treatment. She treated for palliation by radiotherapy. Medical information was provided to the patient and her family regarding the diagnosis, staging therapeutic options and prognosis. The patient died 6 months later.

OC 2
Melkersson-Rosenthal syndrome (MRS). A report of 14 cases

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Melkersson-Rosenthal syndrome (MRS) is a rare disease which is part of orofacial granulomatosis (sarcoidosis, Crohn disease, foreign body reaction, chronic granulomatous disease). Melkersson-Rosenthal syndrome is known for more than 100 years but over this fact its diagnosis is still a problem. MRS is characterized by triad of symptoms: recurrent macrocheilia of granulomatous origin, lingua plicata and facial paralysis. The complete triad is rare. Macrocheilia is almost always diagnosed, others two symptoms are confirmed less frequently. Authors are presenting results of 14 patients with MRS, seven men and seven females. Diagnosis was confirmed by biopsy. Symptoms present were: macrocheilia 14 cases, lingua plicata 7 cases, facial paralysis 4 cases, intraluminar obturation in lymphatic vessels 13 cases.
Therapy: (i) Macrocheilia intralesional application of depot corticosteroids 14, successful (macrocheilia diminish to the normal size and attacks were less frequent) 5, partly successful (macrocheilia diminished to the acceptable size for patient) 6, insufficient (surgical intervention was indicated) 3 (ii) Facial paralysis – resistent to therapy
Summary: (i) MRS is mostly characterized by an incomplete triad. In our set of patients the macrocheilia was always present. (ii) Diagnosis must be confirmed by biopsy. (iii) Application of depot corticosteroids was successful in 11 cases. The surgical intervention was indicated in three cases.
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OC 3
Periodic syndromes presenting with recurrent oral ulceration

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Objectives: Aphthous-like ulceration (ALU) is well recognized in some immunodeficiency states, chronic viral infections, rheumatological disorders and dermatological diseases. Another rare association is with the autoinflammatory syndromes: a diverse group of disorders characterized by recurrent attacks of inflammation in the absence of high-titre autoantibodies or antigen-specific T-lymphocytes. Many of these are hereditary and are referred to as inherited periodic fevers syndromes (IPFS). ALU is a recognised manifestation of several auto-inflammatory disorders including in periodic fever, aphthous-stomatitis, pharyngitis, adenitis (PFAPA), familial mediterranean fever (FMF), hyper IgD and periodic fever syndrome (HIDS), pyogenic sterile arthritis, pyoderma gangrenosum, acne (PAPA) and tumor necrosis factor receptor associated periodic syndrome (TRAPS). ALU has been alluded to in PFAPA (Pinto *et al* Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2006;102:35–9) but there are no reports of any of these in the dental literature. We detail two cases of ALU in periodic syndromes.
Findings: Case 1 A Caucasian family presented with recurring mouth ulcers – a mother, her sister, son, and paternal uncle. Only the son also described episodic fevers. Findings included neutrophilia and raised erythrocyte sedimentation rate and the affected individuals were found to be heterozygote for the TNFRSF1A mutation/polymorphism R92Q, the commonest cause of mild TRAPS in the UK. Case 2 A Caucasian family in which the mother, a 47-year old female presented with an 8-year history of recurrent oral ulceration, fatigue, fever, dyspnoea, arthralgia, acrocyanosis and abdominal discomfort recurring every 4–5 weeks lasting 2–3 days. Her daughter at age 21 started developing recurrent fevers and skin lesions but no oral ulcers. Investigations including the acute phase response were normal, and gene sequencing demonstrated no mutations in MEFV exon 2 or 10, (responsible for familial mediterranean fever), or TNFRSF1A exons 2 to 7. She is controlled on mycophenolate mofetil and corticosteroids during disease flares. Her syndrome is as yet not defined.
Conclusions: Atypical manifestations of autoinflammatory diseases such as presentation with ALU are increasingly recognized; particularly among the less severe mutations which are often also present as low frequency polymorphisms. One case of TRAPS has already recently been reported with mouth ulceration (Saulsbury *et al*. J Pediatr. 2005;146:283–5). IPFS can only be diagnosed if the diseases are considered and specific genetic sequencing performed. Recognition of these syndromes is important as they carry risks of serious long-term complications particularly AA amyloidosis, and often respond well to specific treatments.

OC 4
Oro-facial manifestations of HIV infection – a case series: continued management challenges in the UK

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Introduction: HAART profoundly decreased oro-facial disease presentation in the UK. However drug resistance, compliance and adverse effects, epidemic evolution, social and behavioural factors continue to impact on presentation and management. We detail three HIV-infected individuals recently presenting diverse oro-facial/systemic manifestations and management challenges.
Case reports: A: 48-year old Caucasian male with high risk sexual behaviour with a 2 month history of oral discomfort and dysphagia failing to respond to fluconazole. Examination revealed ulceration of the tongue, lips and palate consistent with lues maligna. Seventeen days of IM penicillin led to resolution. Contact tracing was impossible. B: 17-year old Afro-Caribbean female with perinatally-acquired, multi-drug resistant HIV infection, with severe necrotising periodontitis progressing to necrotising stomatitis. This responded well to frequent local debridement and systemic antibiotics but she subsequently developed severe HIV-related oral ulceration responding to thalidomide. Non-compliance and lack of insight despite a life time of infection impacted on management. C: 18-year old Afro-Caribbean female with severe HIV-related immune thrombocytopenia, needle-phobia and non-compliance with HAART following the loss of her main carer, with extensive caries and necrotizing ulcerative gingivitis. She became the sole carer for two younger relatives and was unable to manage her own health need. Stabilisation occurred with local therapy and systemic antibiotic and difficulties in managing the thrombocytopenia before multiple extractions were overcome.
Conclusion: HAART failure, a return to high risk sexual behaviour, social and cultural factors impact on an increased frequency and nature of presentation, diagnosis and successful management of HIV-infected individuals in London.

OC 5
Joint patient/health care professional conferences – what do they offer?

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Aim: Evaluate a conference for patients and healthcare professionals (HCP) alongside each other.
Method: The Trigeminal Neuralgia Association (TNA) UK, a patient support group, organised a two-day conference for its members. The talks were delivered by a multidisciplinary group of HCPs. Alongside this was a 1-day event for HCP organised by the chairperson of the Medical Advisory Board of TNA. The HCP conference, delivered by a multidisciplinary team, was accredited by the Royal College of Physicians. Patients and HCPs both evaluated the conferences.
Results: Thirty patients evaluated the conference on a scale of 0–10. They all found the conference fulfilled their expectations, particularly the facility to interact with fellow

sufferers and experts, with mean scores over nine. It improved patients' confidence in managing their trigeminal neuralgia, score 8.6. The 20 HCPs who attended, rating on a 5-point scale, scored a mean of 4.5 or above on overall value, meeting education needs and fulfilling objectives. Both groups greatly appreciated the opportunity to talk with each other during breaks and the HCPs were surprised by the reflective and searching questions put to the panel.
Conclusion: Conferences attended by both patients and HCPs provide an opportunity for HCPs to learn what really matters to patients and, it is hoped, provide more patient-centred care in the future.
Relevance: Patients with long-term conditions need to self manage and for this they need high quality information provided in a variety of formats.

OC 6

Rich in eosinophils atypical CD30+ T-lymphoproliferative process with ulcers of the oral mucosa of the type of traumatic eosinophilic granuloma

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Introduction: Traumatic eosinophilic granuloma (TEG) of the oral mucosa is a chronic, self-limited lesion of unknown pathogenesis, which commonly manifests as an ulcer with elevated borders. Histologically, TEG shows a diffuse, dense and polymorphic cellular infiltrate, eosinophilic rich or not, involving the superficial mucosa and the underlying soft tissues.
Aim: The aim of the present study was to present a case of multifocal oral TEG and to emphasize the pathoanatomical heterogeneity of the diseases with the clinical features of TEG.
Case report: A 13-year-old male presented with pain during teeth brushing for the last 5 weeks. The clinical examination revealed ulcers with elevated, indurated borders at the tongue and the attached gingiva of the right maxilla's 1st and 2nd premolar teeth. Clinical differential diagnosis included ulcerative stomatitis, Langerhans cell histiocytosis and non-Hodgkin's lymphoma. An incisional biopsy was performed and the lesions healed completely. Laboratory evaluation was negative. Histological, immunohistochemical and molecular analysis of the lesions revealed an atypical CD30+ T-lymphoproliferative process with abnormal immunophenotype, epitheliotropism and intense eosinophilic infiltrate along with clonic character IgM(k) plasmacytic infiltrate.
Discussion/Results: (i) TEG clinically mimics squamous cell carcinoma, ulcerative gingivitis, Langerhans cell histiocytosis and malignant lymphoproliferative processes, while the histological differential diagnosis includes Langerhans cell histiocytosis, Hodgkin's and non-Hodgkin's lymphoma. (ii) TEG is a group of benign diseases that shows pathoanatomical and histogenetical heterogeneity and includes the atypical or not CD30+ T-lymphoproliferative eosinophil-rich process of the oral mucosa. It is essential to differentiate TEG from malignant lesions, especially peripheral T-lymphoma from large CD30+ positive cells.

OC 7

Amyloidosis: a clinical case with oral and cardiac involvement

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Signs and symptoms: We present a case of a 70-year old man that presented to the Oral Medicine clinic with a history of persistent mouth ulcers and severe difficulty in eating.
Medical history and social history: The patient was known to suffer from chronic renal failure and recurrent light chain multiple myeloma that had been diagnosed 6 years previously. The multiple myeloma had been treated with autologous stem cell transplantation and five cycles of high dose medical therapy involving thalidomide, dexamethasone and pamidronate.
Oral disease history: The oral disease had started approximately 3 months previously and included persistent mouth ulcers and pain in the tongue. The resultant difficulty eating had caused weight loss and was compromising treatment of the myeloma. Intraoral examination revealed macroglossia, restricted tongue movement, ulcerated lesions on the borders of the tongue and an ulcerated mass in the floor of the mouth.
Diagnosis: A biopsy of the oral lesions revealed the presence of extensive amyloid deposits affecting the tongue and floor of mouth. Further investigations revealed features of systemic amyloidosis, with heavy amyloid deposits in cardiac muscle and tongue, with important clinical consequences.
Treatment: Unfortunately despite referral to a specialist centre for the treatment of amyloidosis no treatments proved effective and the patient died as a consequence of his cardiac involvement. We report this very severe case and detail the latest therapeutic options for managing this condition.

OC 8

Intraoral disease in hereditary epidermolysis bullosa reflects mutation-based alteration of the basement membrane zone

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During early odontogenesis, the basement membrane is thought to be important in epithelio–mesenchymal interactions and the subsequent interdigitation between inner enamel epithelium and the underlying dentine resulting in the highly complex amelodentinal junction. Mutations in genes of the major anchoring proteins within the basement membrane zone (BMZ) might therefore be expected either to seriously compromise ameloblast differentiation and/or to interfere with normal basement membrane formation and degradation and thus the fixation of ameloblasts to their underlying matrix. Indeed, teeth of patients suffering from junctional epidermolysis bullosa (JEB), in which BMZ proteins like laminin five are structurally and functionally compromised, are regularly and often severely affected by abnormal dental development. We present a 42-year old JEB-patient with pronounced intraoral involvement to demonstrate the effects of molecular alterations during odontogenesis with impaired development of dental supporting structures. In addition, the spectrum of intraoral disease in EB in general, and relevant etiologic factors are discussed based on our experience with patients seen in the EB-house Austria, a specialized medical center dedicated to EB-specific therapy and research. Finally, we provide major strategies to oppose common EB-specific problems in dental care, including (i) prevention e.g. by counseling and regular follow-ups by a dentist, (ii) restoration of enamel and dentin defects with fillings and stainless steel crowns to minimize wearing and to guarantee structure and function of teeth, (iii) extraction of most severely affected teeth with osteolytic foci to remove continuous sources of oral infections and, finally, (iv) approaches to restore masticatory function and esthetics by e.g. removable prostheses, implant-tissue-supported overdentures or osseointegrated implants.

OC 9

Squamous cell carcinoma of the lower lip arising in lesions of discoid lupus erythematosus: a case report

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Signs and symptoms: A 40-year old woman was referred for evaluation of a painless, ulcerated mass on the right lower labial vestibule.
Medical history and social history: The patient had a 21-year history of discoid lupus erythematosus (DLE) mainly affecting the skin of the face and scalp, which had been treated with steroids, immunosuppressors and immunomodulators. At presentation, she was taking hydroxychloroquine and methylprednisolone.
Oral disease history: According to the patient, the labial vestibular mass appeared before 10 days, while oral mucosal lesions had been present since the onset of DLE with remissions and exacerbations. Physical examination revealed multiple facial cutaneous lesions. On the right lower labial vestibule, a 1.5×1.5 cm, broad-based, indurated, exophytic mass with a crusted, ulcerated surface was noticed. Intraoral examination revealed ulcers, atrophic areas and whitish plaques on the buccal and labial mucosa, hard palate and tongue. There were no palpable neck masses.
Diagnoses: A clinical diagnosis of squamous cell carcinoma (SCC) of the lower lip developing in the context of DLE, along with oral lesions of DLE, was considered. Biopsy of the labial mass revealed a low to intermediate grade SCC, while the intraoral lesions were histologically consistent with DLE.
Treatment: The tumor was completely excised. Systemic and topical steroids were used for the treatment of the intraoral lesions. Four months later, no signs of tumor recurrence were observed.
Conclusions: Squamous cell carcinoma developing in lesions of DLE is a rare occurrence with only 11 previously reported cases arising in the lips of patients with chronic DLE involvement.

OC 10

Malignant tumors mimicking periapical lesions

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The purpose of this study is to present three cases of malignant tumors appearing as periapical lesions. The records of the patients were reviewed. Demographic data, presenting symptoms, radiographic findings, bone scintigraphy, histopathology and clinical management were analyzed. The patients were 23, 33 and 35 years. Two cases were presented in the mandible, related to the first and second mandibular molar. One

case was related to the maxilla posterior. The clinical presentations were: exophytic soft tissue mass, tooth pain, paresthesia of the lower lip and a periapical lesion (Patient 1); paresthesia and periapical lesion (Patient 2); facial enlargement, pain and image of a residual cyst (Patient 3). The patients were referred to an oncologist, the provided treatment protocols were: chemotherapy, radiotherapy and chemotherapy, surgery (for all three). In view of these cases it can be said that meticulous work-up of jaw lesions suspected of being malignant, may be life saving or extend the patient's survival period.

OC 11

An unusual portrait of secondary syphilis

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We describe a case of a 45-year old man referred to our unit for a 1-month history of an asymptomatic 'leukoplakia-like' plaque and for several painful whitish lesions. The patient had been treated with topical corticosteroids and antimycotic, with no favourable improvement of lesions. In the anamnesis, he referred to be a non-smoker, to suffer of HCV-related chronic hepatitis, to be penicillin-allergic. Furthermore, he referred an asymptomatic ulcerative lesions on the glans penis 7 months previously, disappeared spontaneously after 2 weeks. He noted that the appearance of oral lesions was followed by a diffuse cutaneous rash and by fever, pharyngitis, asthenia and dysphagia. Oral examination revealed a well delimited, raised and with a corrugate and non-homogeneous surface 'leukoplakia-like' plaque of the right buccal mucosa. Furthermore, painful white mucous plaques, some with an eroded/ulcerated area and surrounded by an erythematous border were observed on the right lateral margin of the tongue, on the lower labial mucosa and on the left commissure. Oral lesions were associated with painless lymphadenopathy. Because of these anamnestic and clinical data a secondary syphilis with oral lesions was considered. Histological examination of the 'leukoplakia-like' lesion demonstrated an unspecific inflammatory infiltrate in the lamina propria and acanthosis, papillomatosis and hyperkeratosis of the epithelium. HIV serology was negative. The serologic tests for syphilis confirmed our initial hypotheses. This case emphasizes the importance of considering secondary syphilis in the differential diagnosis of white and/or ulcerative oral lesions.

OC 12

Early surgical laser-assisted management of bisphosphonates – associated osteonecrosis of the jaw: evaluation of 85 ONJ patients

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From January 2004 to March 2008, 85 patients affected by ONJ-BP were observed at Sezione di Odontostomatologia, University of Parma, Italy, for a total of 101 ONJ-sites recorded. Of these 101, 79 sites were treated at our Unit. Strategy treatment was performed in accordance with the 'available guidelines' (present at the time of the first examination of each lesion), as well as systemic status of the patient and his compliance. Characteristic of the lesion and its response to antibiotics was also considered. The use of low level laser therapy (LLLT) in order to improve healing process of these lesions was added for the management of ONJ. 'Clinical success' has been evaluated for ONJ-BP lesions as a treatment able to give a positive result that may be maintained over at least 3 months free from signs and symptoms: transition from a higher stage to a lower one or complete mucosal healing (here classified as Stage '0'). The treatments performed on the 79 ONJ-BP sites have retrospectively been subdivided in four different groups: G1, 13/85 ONJ-BP sites treated only with medical therapy; G2, 17/85 ONJ-BP sites treated with medical therapy and LLLT; G3, 13/85 ONJ-BP sites treated with medical and surgical therapy; G4, 36/79 ONJ-BP sites treated with medical, surgical therapy and LLLT. For G1 no clinical success was recorded, 6/13 (46%) of ONJ sites in G2 had a transition to Stage 0 as well as 7/17 (41%) in G3. In addition, an improvement was also recorded for 28/36 ONJ sites (80.5%) treated in G4 (21/36 Stage 0). The follow-up was of from 3 to 60 months (mean follow-up 18 months). In our experience, an early conservative surgical approach (possibly associated to laser treatment) for BP-induced ONJ could be considered as more efficacious in comparison to medical therapy for the management and quality of life of these patients.