Oral manifestations of systemic diseases

Brian Swinson, Helen Witherow, Paul Norris, Tim Lloyd

In the clinical examination for systemic diseases, inspection of the oral cavity is often overlooked. This area provides a wide array of clinical signs that may help the clinician establish a diagnosis. This article provides a summary of the more common oral manifestations of systemic disease.

Many systemic diseases may present with manifestations in the oral cavity. These may, in some cases, precede any other clinical signs of the disease. The ease of examination of this area makes it important for the clinician to be familiar with the more common oral presentations of various diseases. Familiarity with the various oral manifestations may also help the clinician differentiate between systemic and neoplastic change within the oral cavity. This article presents some of the more common oral findings that may alert the clinician to the possibility of systemic disease.

DERMATOLOGICAL

Lichen planus

Lichen planus is a common mucocutaneous disorder of unproven aetiology, although a few cases have an identifiable aetiology. Chronic liver disorders, graft *vs* host disease, hepatitis C, reaction to dental amalgam and several drugs have been shown to cause the oral lesions.

The main clinical features of the cutaneous form are pruritic, polygonal, purple papules occurring most commonly on the forearms and shins. Hair loss, ridging of the nails and plaques or erosions of the genital area may also be seen.

Oral lesions: The mucosal lesions are usually painless although pain may be a feature if there is mucosal breakdown. They are bilateral, usually affecting the buccal mucosa and lateral borders of the tongue. They form a reticular pattern of white mucosal striae (*Figure 1*) or white plaques. Desquamative gingivitis around the necks of the teeth may also be seen while areas of erosion and ulceration may cause pain (Eisenburg, 2000).

Management: The diagnosis is usually clinical but may require biopsy to exclude other pathologies. Topical corticosteroids are the mainstay of treatment, with non-responders requiring intralesional steroids, topical cyclosporin or in extreme cases systemic immunosuppression. Regular follow up is advised for the erosive form because of the potential for malignant transformation.

Pemphigus

This is an uncommon, autoimmune vesiculobullous disease occurring most commonly in older age groups. It has a higher incidence in Middle East and Mediterranean peoples and also in Ashkenazi Jews. It can also be induced by certain drugs, namely penicillamine and captopril.

Cutaneous lesions take the form of large flaccid bullae affecting any site, the genital areas being particularly prone.

Oral lesions: Bullae and vesicles affect the oral mucosa. These then burst to leave irregular, ragged erosions and ulcers, which are persistent in nature. A shearing force applied to the oral mucosa results in bullae formation (Nikolsky's sign) and is helpful in establishing the diagnosis.

Management: Immediate specialist referral is advised as the disease may prove fatal if untreated. Diagnosis is established by biopsy and immunostaining. High-dose corticosteroids are the treatment of choice, usually combined with steroid-sparing agents such as azathioprine or dapsone.

Pemphigoid

Mucous membrane pemphigoid is an autoimmune disease affecting the oral cavity, most commonly seen in middle-aged and elderly women. The mucous membrane pemphigoid variant is one of the most commonly encountered in the oral cavity but also affects nose, pharynx, skin and eyes. Ocular lesions are worrying and an ophthalmic opinion should be sought to prevent conjunctival scarring. Subepithelial bullae are seen as a result of damage to the epithelial basement membrane by antibodies (Anhalt, 1990).

Oral lesions: Tense vesicles, occasionally blood-filled, affect predominantly epithelialized mucosa, i.e. the palate and gingivae (desquama-

Mr Brian Swinson, Ms Helen Witherow and **Mr Paul Norris** are Specialist Registrars in Oral and Maxillofacial Surgery and **Mr Tim Lloyd** is Consultant in Oral and Maxillofacial Surgery, University College Hospitals, Mortimer Market, London WC1E 4AU

Correspondence to: Mr B Swinson

tive gingivitis). The lesions often heal with scarring, causing mucosal contractures.

Management: The diagnosis should be confirmed by mucosal biopsy and examined for evidence of immunoglobulin (Ig) G or C3 deposits at the basement membrane under direct immunofluorescence. Topical local corticosteroid treatment is usually adequate to control the lesions.

Erythema multiforme

This mucocutaneous condition affects predominantly young males. In half the cases the aetiology is unknown but it may be caused by several drugs, i.e. sulphonamides, penicillins or barbiturates. Herpes simplex and *Mycoplasma pneumoniae* have also been implicated. The major form is Steven–Johnson syndrome, which involves the mouth, eyes, skin and genitals, and presents with sudden fever and enlarged cervical lymph nodes (Wilkins et al, 1992).

Oral lesions: Large, erythematous, shallow ulcers are seen covered with haemorrhagic sloughs affecting any part of the oral mucosa. Cracked and crusted lips are also seen. A classical 'target lesion' occurring on the hands and feet may be seen, as well as ocular and genital lesions (*Figure 2*).

Management: Specialist referral is indicated and serology is required to check if herpes simplex or mycoplasma is the causative agent. Acyclovir should be started if herpes is suspected. Tetracycline mouthwashes promote ulcer healing and chlorhexidine resists super infection. Topical steroids are usually given but their benefit is unproven. Severe cases should be hospitalized.

Epidermolysis bullosa

There are three types of this inherited, blistering mucocutaneous disorder: simplex, junctional and dystrophic. The clinical picture ranges from relatively mild blistering to a severe fatal condition. The lesions commence as vesicles and bullae that affect areas of minor irritation. These rupture and form areas of erosion and ulceration which heal by scarring, causing distortion and contractures. The hands and feet are commonly affected while patients with the recessive dystrophic variant are predisposed to cutaneous squamous cell carcinoma.

Oral manifestations: Milder forms manifest as gingival erythema, tenderness and occasionally recession. More severe forms cause mucosal contracture with fibrous bands leading to obliteration of the labial groove, microstomia and ankyloglossia.

Management: This depends on the type and severity of the disease. A soft non-traumatic, non-cariogenic diet is encouraged and local anaesthetic can be administered slowly if required. No cure exists because of the genetic nature of the disease and patients and family members should receive genetic counselling.

Peutz–Jeghers syndrome

This rare condition is usually inherited as an autosomal dominant trait; it is characterized by freckle-like lesions of the hands, perioral skin and oral mucosa. This is seen in conjunction with intestinal polyposis, with a tendency to undergo malignant transformation (McGarrity et al, 2000). The intestinal polyps are hamartomatous in nature, usually affecting the small bowel, and may present with obstruction or intussusception. Ovarian and bladder tumours are also seen more frequently with this condition.

Oral lesions: These primarily affect the vermillion zone, labial and buccal mucosa and occasionally tongue. They are normally 1–3 mm in diameter and can be brown or bluish-grey in colour (*Figure 3*).

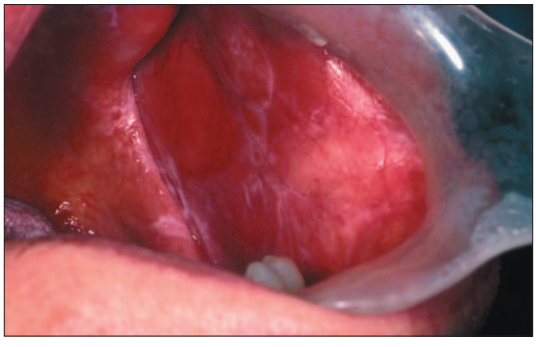


Figure 1. White mucosal striae, often seen bilaterally, in lichen planus.

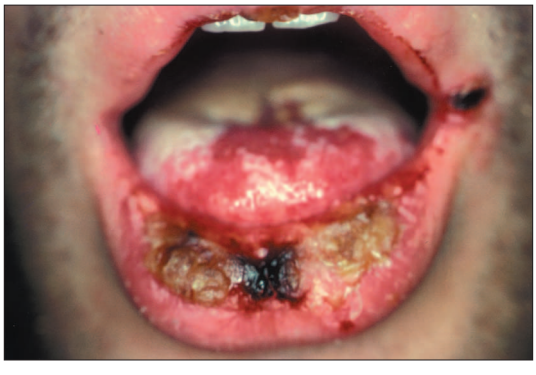


Figure 2. Lip and mucosal ulceration seen in erythema multiforme.

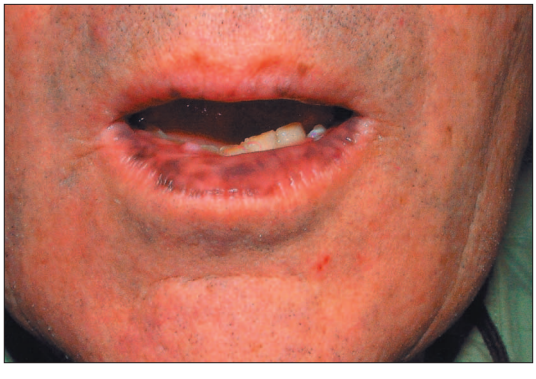


Figure 3. Patchy brown discolouration of the lips in Peutz–Jeghers syndrome.

Management: These patients should be monitored for any evidence of obstruction or intussusception and the clinician should have a low threshold for investigation of any abdominal complaint. Treatment for the perioral pigmentation is reassurance. Genetic counselling is also appropriate.

Several, more uncommon skin conditions have been seen to affect the oral cavity include linear IgA disease, dermatitis herpetiformis and lupus erythematosus and should be borne in mind in the differential diagnosis.

GASTROINTESTINAL

Crohn's disease

Along with ulcerative colitis, Crohn's disease comes under the umbrella of chronic inflammatory bowel disease. Affecting any part of the gastrointestinal tract, this disorder results in thickening and ulceration of the mucosa, most commonly in the ileocaecal region.

The main clinical symptoms include abdominal pain, constipation or diarrhoea, obstruction and malabsorption. Perianal fistulae or fissures may also be seen.

Oral lesions: Typical oral symptoms include aphthous-like ulceration and recurrent or persistent swelling of the lips. The oral signs may precede the bowel symptoms and commonly include a 'cobblestone' appearance of the buccal mucosa, angular stomatitis, hyperplastic and erythematous gingivae. Mucosal tags, linear ulceration with nodular swellings of the tongue and indurated fissures on the midline of the lower lip are occasionally seen (Kalmar, 1994). Oro-cutaneous fistulas have also been reported (*Figure 4)*.

Management: Crohn's disease may be confirmed by haematological investigation, sigmoidoscopy, rectal mucosal biopsy and radiography.

Nutritional deficiencies should be corrected appropriately and diet control is often applied. Good oral hygiene is recommended and the use of corticosteroids systemically, topically and intralesionally has been advocated. Fluoride supplementation may be of some use.

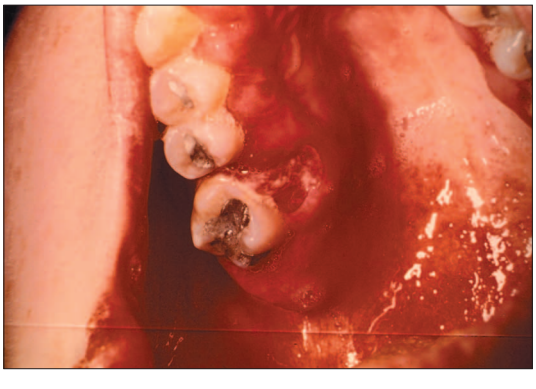


Figure 4. Crohn's disease showing deep mucosal ulceration adjacent to a molar tooth.

Ulcerative colitis

This is another inflammatory disorder that always affects the rectum and extends proximally for a variable distance in the large intestine. The main clinical features include bloody diarrhoea, weight loss, abdominal cramps and distention. Long-term complications of ulcerative colitis may include neoplastic change and amyloidosis.

Oral lesions: Recurrent oral aphthous ulceration, which may pre-date the bowel lesions, and haemorrhagic ulcers of the oral mucosa are commonly seen. Other ulcers, clinically resembling pyoderma gangrenosum, have been noted.

Oral pustules may also be recognized in patients with ulcerative colitis and have been termed 'pyostomatitis vegetans'. Spondyloarthropathy, causing erosion of the temporomandibular joint, may occasionally be seen.

Management: This is similar to Crohn's disease and diagnosis is confirmed by rectal biopsy. Treatment is systemic or local and the correction of any nutritional deficiencies is essential. Topical or intralesional steroids are useful for the management of oral lesions although there is a tendency for recurrence.

Coeliac disease

Coeliac disease is a malabsorptive disorder of the small intestine in gluten-sensitive individuals. Ingestion of gluten leads to atrophy and inflammation of the villi of the small intestine. Abdominal discomfort, steatorrhoea, diarrhoea and weight loss may be seen but symptoms can often be non-specific with only tiredness and malaise seen. Failure to thrive may be the presenting feature if it occurs in childhood.

Oral lesions: Oral manifestations of coeliac disease include minor aphthous oral ulceration, which may only be present in up to 5% of patients. Other signs may be glossitis, burning mouth and angular stomatitis – these are mainly in association with underlying nutritional deficiencies. Children presenting early with coeliac disease may show enamel hypoplasia.

Management: Coeliac disease is best managed by dietary control, which consists of a gluten-free diet. Any underlying nutritional deficiencies should be corrected. Oral ulceration can be managed with 2% lignocaine gel, Corsodyl mouthwash or topical steroid preparations.

Liver disease

Liver disease may affect the oral cavity in a number of ways depending on the original aetiology. The disease and also the medical treatment of the disease may affect the mouth.

Alcoholic liver disease: The underlying bleeding tendency may manifest itself as gingival bleeding, while bilateral parotid swelling (sialosis) may often be seen in cirrhotic patients.

Primary biliary cirrhosis: Many cases of primary biliary cirrhosis (PBC) can be complicated by Sjögren's syndrome. This involves the combination of xerophthalmia, xerostomia and a connective tissue disease. The dry mouth can cause difficulty in denture wearing and dental caries.

Lichen planus has also been shown to complicate some PBC cases with the features described above.

Many patients with liver disease are managed on systemic corticosteroid therapy, which again predisposes to oral candidiasis.

Pernicious anaemia

Destruction of the gastric parietal cells, via several possible causes, leads to a lack of intrinsic factor required for the absorption of vitamin B_{12}, causing a megaloblastic anaemia. This may lead clinically to lethargy and shortness of breath.

Oral manifestations: The tongue is commonly affected and becomes depapillated, red, beefy and painful. The pain is often described as 'burning' in nature. Focal patches of mucosal atrophy or ulceration may also be seen in conjunction with angular stomatitis and candidiasis (Greenberg, 1981).

Management: Treatment usually consists of monthly intramuscular injections of vitamin B_{12} (cyanocobalamin) causing rapid resolution of the oral symptoms.

Plummer–Vinson (Paterson–Kelly) syndrome

This relatively rare condition presents with iron-deficiency anaemia in conjunction with glossitis and dysphagia, usually in females from northern Europe.

The dysphagia is caused by the development of a thin diaphanous web across the upper third of the oesophagus and can be demonstrated by endoscopy or barium contrast radiographic studies. Koilonychia of the nails is also often seen.

The condition is significant in that it has been associated with a high frequency of both oral and oesophageal squamous cell carcinoma (Hoffman and Jaffe, 1995).

Oral manifestations: Patients often complain of a burning sensation of the mouth and tongue. Atrophy of the lingual papilla causes a red, smooth and shiny appearance of the tongue, which is seen in conjunction with angular cheilitis. Dysphagia and occasionally odynophagia may be noted.

Treatment: Correction of the iron deficiency anaemia usually relieves the oral symptoms but regular oesophageal dilatation may be required to improve the dysphagia.

Regular surveillance of the oral cavity, hypopharynx and oesophagus is needed to check for malignant change.

ENDOCRINE

Hypoadrenocorticism

Addison's disease may result from atrophy of the adrenal cortices, leading to insufficient release of cortisol and aldosterone. Destruction of the adrenal cortices may be a result of circulating autoantibodies, although Graves' disease, Hashimoto's disease, tuberculosis, acquired immunodeficiency syndrome (AIDS) and malignancy are less commonly associated with adrenal insufficiency.

The clinical features tend to present insidiously with abdominal upset, fatigue, weakness and depression. Hyperpigmentation of the skin, especially in scars, is seen with patches of vitiligo occasionally present. Adrenal crisis is the main complication and may lead to collapse, hypotension, bradycardia, shock and even death.

Oral lesions: Oral pigmentation, which is asymptomatic, is seen diffusely and usually bilaterally on the buccal mucosa or tongue and may precede the cutaneous pigmentation. The stress of dental treatment may induce a crisis and corticosteroid cover must be given preoperatively.

Management: Diagnosis is confirmed by performing a synacthen test. Radiography or computed tomography scanning may be helpful in finding the cause of the condition. Treatment in most cases is long-term corticosteroid replacement therapy.

Diabetes mellitus

The resultant hyperglycaemia, whether insulin dependent (type 1) or non-insulin dependent (type 2), affects many organ systems and may cause neuropathy, retinopathy, renal dysfunction, cardiovascular and metabolic complications that may have oral manifestations.

Oral manifestations: These are more commonly seen in type 1 diabetes. Xerostomia is present in one third of diabetic patients and may cause a tender burning mouth. This is often seen with candidiasis and is felt to be the result of hyperglycaemia (*Figure 5*). Periodontal disease, as a result of angiopathy, is seen more frequently and tends to progress more rapidly than in non-diabetic patients while susceptability to caries is also recognized in these patients. Diffuse non-

tender, bilateral enlargement of the parotid glands (diabetic sialadenosis) may be seen in either type 1 or 2. Zygomycosis is a fulminant fungal infection seen in poorly controlled, ketoacidotic type 1 patients and can cause extensive tissue destruction.

Management: The management of oral symptoms, as with all diabetic patients, is good glycaemic control.

Oral symptoms may be minimal in well-controlled diabetics. There is a great need for immaculate oral hygiene and regular dental visits. Dietary control is important as these patients are susceptable to caries. Oral candidiasis is best treated with oral or topical antifungals (Bell et al, 1999).

Acromegaly

Excessive growth hormone production, after fusion of the epiphyseal plates, results in this uncommon condition seen in adult patients. A pituitary adenoma is the usual cause and it may present as headaches, visual disturbances or irregularities of other pituitary hormones. Systemically heart disease, hypertension and peripheral neuropathy are seen. Growth of the small bones of the hands and feet is a feature, together with membranous bones of the skull and jaws (Cohen and Wilcox, 1993).

Oral manifestations: Hard and soft tissues are affected by this condition. Coarsening of the facial features is seen, together with soft palate hypertrophy, resulting in sleep apnoea. Macroglossia with scalloping of the lateral margins and thickening of the lips are noted.

Calvarial thickening and associated growth of the sinuses is present. Development of the frontal sinuses results in the appearance of frontal bossing. Mandibular prognathism with an increase in alveolar bone causes spacing of the teeth and an anterior open bite of the dentition.

Management: Surgical excision of the pituitary mass via the transsphenoidal approach is usually undertaken. This may be used in conjunction with somatostatin analogues while radiation therapy has a role in some cases. Surgical mandibular resection and frontal remodelling may be used to improve cosmesis in well-controlled cases (*Figures 6a* and *b*).

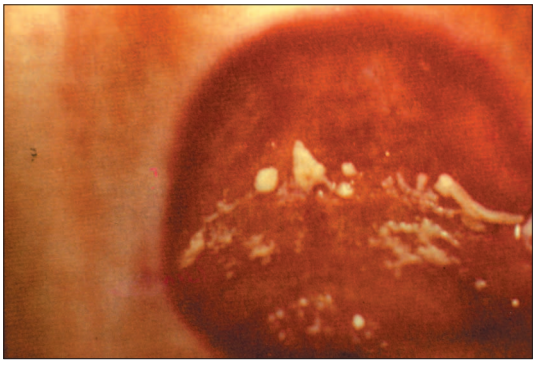


Figure 5. Candidiasis present on the hard palate.

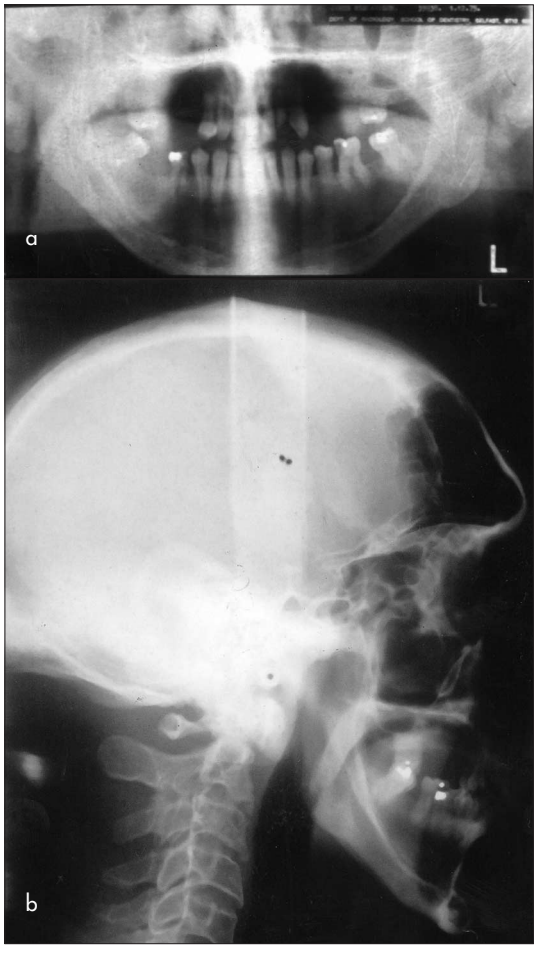
Hypothyroidism

Hypothyroidism in adults causes the deposition of a glycosaminoglycan ground substance in the soft tissues producing a non-pitting oedema, described as myxoedema. Clinically it presents with lethargy, dry coarse skin, alteration of the voice, constipation and fatigue.

Oral manifestations: The lips and tongue are thickened as a result of the glycosaminoglycan deposition. The resulting macroglossia may affect speech and cause a poor oral seal, resulting in 'mouth breathing'. Xerostomia and candidiasis may be a feature and in the infantile form delayed eruption of the dentition is seen.

Management: Thyroid replacement therapy is the treatment of choice. Meticulous oral hygiene should be encouraged and antifungals commenced if required.

Figure 6. a. Orthopantomogram showing proliferation of the lower alveolar bone and prognathism in acromegaly.
b. Mandibular and frontal sinus development in acromegaly.



HAEMATOLOGICAL DISORDERS

Anaemia

The term anaemia covers a wide spectrum and is not a disease per se, but a manifestation of many other diseases. Insufficient intake, impaired absorption, increased demands (e.g. pregnancy) or blood loss may result in deficiencies of elements necessary for haemopoesis, giving rise to differing clinical presentations.

Oral manifestations: These may range from a burning sensation of the mouth and tongue, to atrophy of the filiform and fungiform papillae giving the tongue the smooth dorsal surface seen in atrophic glossitis. Other features include oral pallor, oral ulceration, angular cheilitis and secondary candidiasis.

Management: The aim is to treat the underlying cause, and diagnosis is via haematological investigation. Oral administration of ferrous sulphate is simple and effective in many cases of iron deficiency anaemia and response is good. Oral signs of vitamin B_{12} deficiency respond well to intramuscular injection of hydroxocobalamin, while folic acid supplementation may also be needed.

Leukaemia

Neoplastic change in the haemopoetic stem cells results in differing forms of leukaemia. Classification into acute or chronic and then myeloid or lymphoblastic only gives a rough guide to clinical behaviour.

Clinical features include anaemia, bleeding, pallor, weakness, splenomegaly, hepatomegaly and lymphadenopathy.

Oral manifestations: Gingival and periodontal bleeding is often the first presentation of leukaemia. These may be present with petechiae or gingival swelling as a result of leukaemic infiltration. Ulceration of the oral mucosa are typically deep and punched-out lesions with a necrotic base. Opportunistic infections, such as candida or herpesviruses, are commonly seen (Heaney and Golde, 1999). Cervical lymph node enlargement may also be encountered. It should be remembered that oral signs may result from the leukaemia and also the chemotherapy used to treat it (*Figure 7*).

Management: Chemotherapeutic regimens form the main treatment of leukaemia although the individual protocols and prognosis depend greatly on the type of disease being treated. Control of secondary infection by acyclovir and antifungal medication should be commonplace and meticulous oral hygiene followed.

Hereditary haemorrhagic telangiectasia

This autosomal dominant trait results in vascular hamartomas affecting the skin and mucosae. Diagnosis is often as a result of several episodes of epistaxis, which reveals numerous scattered red blanching papules. These can also be seen on the brain, spinal cord and scattered throughout the respiratory, gastrointestinal and genitourinary tracts.

Oral manifestations: The telangiectatic vessels are most commonly seen on the lips, vermillion border and dorsal and ventral surfaces of the tongue, although any intraoral site may be affected (*Figure 8*). They have a tendency to bleed if traumatized, although this is less commonly experienced than epistaxis.

Management: Mild cases require no treatment while cryotherapy may be needed in more severe examples. Patients with hereditary haemorrhagic telangiectasia and evidence of pulmonary arteriovenous malformation will require antibiotic prophylaxis for dental treatment.

Lymphoma

Hodgkin's disease accounts for approximately 20% of cases of lymphomas, with non-Hodgkin's lymphomas providing the remaining 80%. The cervical and supraclavicular nodes are a common presentation of Hodgkin's lymphoma while non-Hodgkin's lymphomas has a higher percentage of extranodal disease. The prognosis is influenced by 'B symptoms' such as weight loss, fever and night sweats.

Oral manifestations: Waldeyer's ring is involved more commonly in non-Hodgkin's lymphomas and presents as a non-tender, slow-growing cervi-

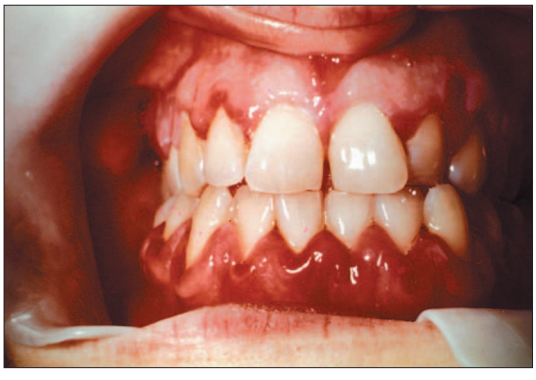


Figure 7. Gingival swelling, ulceration and bleeding may be the presenting features of leukaemia.

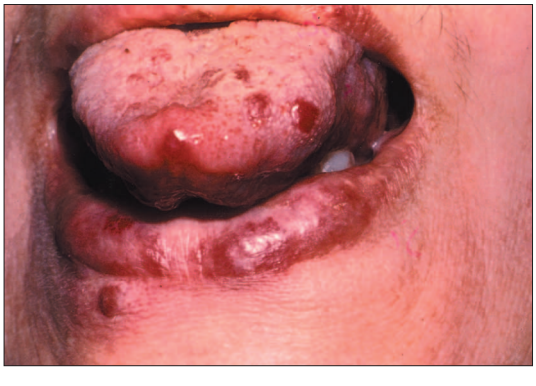


Figure 8. Lip, mucosal and tongue lesions present in hereditary haemorrhagic telangiectasia.

cal mass. Hodgkin's disease rarely affects the mouth, but if present usually appears as extranodal disease and may affect bone as well as soft tissues. These masses can present anywhere in the oral cavity and a soft or hard tissue mass should raise the suspicion of a lymphomatous infiltrate.

Infection with herpes zoster or candida may occur secondary to the disease process or the cytotoxic therapy.

Treatment: The combination of chemotherapy and radiotherapy is commonly used with the prognosis dependent on the disease stage.

BONE DISORDERS

Paget's disease

This rare condition principally affecting older patients is characterized by abnormal and uncoordinated deposition and resorption of bone. Most cases are polyostotic. The affected bones become thickened, enlarged and weakened causing kyphosis, tibial bowing and enlargement of the cranium. Radiographically the bone may show a 'cotton wool' appearance, although this depends on which phase the disease is in. Death may occur secondary to a high output cardiac failure in the osteoclastic phase while malignant change in the form of osteosarcoma is found in 1% of cases and carries a grave prognosis.

Oral manifestations: After calvarial, maxillary involvement is more common than mandibular and progressively gives rise to the striking appearance of 'leonine facies'. This may result in enlargement of the alveolar ridges, causing spacing of the teeth and obliteration of the maxillary sinuses. A common complaint is that dentures no longer fit. Hypercementosis of the roots is seen, causing difficulty in extractions, and this can occasionally be followed by osteomyelitis in the osteosclerotic phase or severe bleeding in the osteoclastic phase (Smith and Eveson, 1981) (*Figure 9*).

Management: The use of calcitonin and bisphosphonates can reduce bone turnover and restore the high levels of alkaline phosphatase to the normal range. Deafness and visual disturbances can prove difficult to treat, while regular denture replacement may be needed because of the increasing alveolar ridges. Regular surveillance for malignant change should be undertaken.

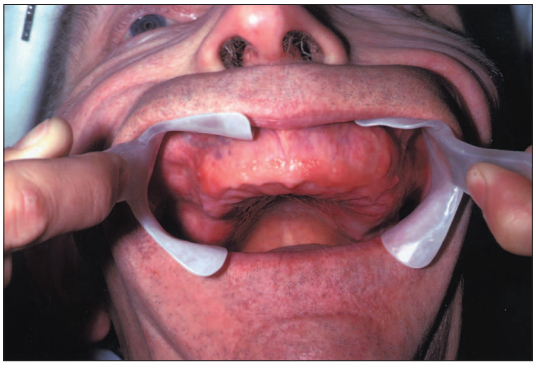


Figure 9. Alveolar and maxillary hyperplasia in Paget's disease.

Osteogenesis imperfecta

This inheritable, heterogenous group is caused by a defect in collagen maturation, resulting in poorer quality bone, dentine, sclera, skin and ligaments. This results in weakening of these structures, with the weight-bearing bones being most obviously affected. In its severest form babies are stillborn while in others the clinical picture, depending on the severity, is one of long bone and spinal deformities, blue sclera, hearing loss and joint hyperextensibility.

Oral manifestations: Mandibular prognathism, combined with an enlarged tongue, causes splaying of the lower anterior teeth. Mandibular fractures are rarely seen, in contrast with their long bone counterparts. Dentinogenesis imperfecta (as a result of defective dentine production) may be present in the dentition causing a blue translucence and fragility to the teeth. They have bulbous crowns with obliteration of the pulp space seen radiographically (Schwartz and Tsiporas, 1984).

Management: These patients pose a dental challenge as the teeth soon decay and fragment. Treatment includes strict oral hygiene, glass ionomer cement restorations and complete overdentures to restore the vertical occlusal height.

Hyperparathyroidism

The excessive production of parathyroid hormone can be classified as primary, secondary or tertiary. Altered calcium metabolism leads to the development of renal calculi and metastatic calcification. Loss of the normal trabecular pattern and decrease in density cause the 'ground glass' appearance on radiographs while the so-called 'brown tumours' are locally destructive lesions that may develop into osteitis fibrosa cystica (central fibrosis of longstanding brown tumours).

Oral manifestations: Loss of the lamina dura surrounding the teeth and blurring of the margins of the inferior dental canal is seen radiographically. The above named bony changes may be seen in the mandible and central giant cell lesions can be seen in the bone or soft tissues.

Management: Surgery, in the form of parathyroidectomy, is usually indicated in the presence of bone lesions. Medical management has a limited role to play.

Osteopetrosis (Albers–Schoenberg disease)

This rare group of hereditary skeletal diseases is characterized by increased bone density with a reduced medullary cavity as a result of a defect

in the normal osteoclastic bone resorption. The infantile (malignant) form is usually inherited as an autosomal recessive trait leading to diffuse sclerosis of the skeleton, marrow failure, multiple fractures and cranial nerve compressions. The adult form is usually less severe.

Oral manifestations: In the malignant form facial deformity is present with a broad face, hypertelorism, frontal bossing and flattening of the alveolar ridges. The teeth show delayed eruption and short roots. Cranial nerve compressions may result in facial paralysis.

Osteomyelitis of the jaw bones is a feared complication following tooth extraction. This is caused by poor blood supply causing areas of avascular necrosis which predispose to the osteomyelitis.

Management: In the infantile form, bone marrow transplantation offers the only hope of cure. Other therapies (interferon gamma with calcitriol) may help to lower bone mass. Osteomyelitis of the jaw bones requires aggressive management with drainage, surgical debridement and appropriate prolonged antibiotics. Hyperbaric oxygen therapy aids healing of recalcitrant areas but surgical reconstruction is occasionally required.

Fibrous dysplasia

This is probably the most common fibro-osseous lesion affecting the jaws. Excessive proliferation of cellular fibrous connective tissue is mixed with irregular, immature, non-lamellar bone. The monostotic form commonly affects the jaws and is usually seen in the second decade of life. Presentation is of an insidious, slow, painless growth of the maxilla more commonly than the mandible. If the maxilla is affected adjacent bones (zygoma or sphenoid) may also be involved. The polyostotic form more often affects the long bones.

One polyostotic form (McCune–Albright) is characterized by multiple affected sites coupled with café-au-lait pigmentation and multiple endocrinopathies, e.g. hyperthyroidism or pituitary adenoma. Rarely, sarcomatous change has been described but usually in patients who received radiotherapy for their lesions.

Oral manifestations: The monostotic variety predominates with the usual presentation as a slowly developing facial asymmetry more often affecting the upper jaw. If the tooth-bearing areas are affected there may be expansion of the buccal and palatal plates with splaying of the teeth. Radiographically the lesion appears poorly demarcated with a ground glass or 'peau d'orange' appearance (MacDonald-Jankowski, 1999) (*Figure 10*).

Management: These lesions tend to burn themselves out eventually and any surgical intervention is best reserved until then. Surgical cosmetic reshaping may be preceded with pamidronate to control osteoclast activity. **HM**

Conflict of interest: none.

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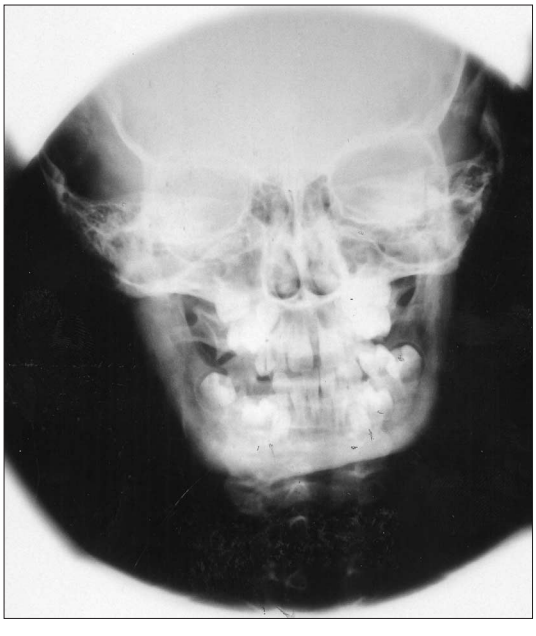

Figure 10. 'Ground glass' appearance of fibrous dysplasia in the right mandible.

KEY POINTS

- Many systemic diseases present with oral manifestations.
- Oral signs may precede other systemic manifestations.
- Both the disease and the treatment may affect the oral cavity.
- Treatment may require systemic therapy in addition to oral measures.
- Early referral to an oral specialist may hasten the diagnosis.