Unusual association of diseases/symptoms

Oral ranula in an HIV-positive patient: case report and literature review

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We describe the presentation and treatment of an HIV-positive patient with an oral ranula, and review the literature. Ranulas are mucocèles or retention cysts formed by the extravasation of mucus from the sublingual gland, presumably due to continued production of saliva in the presence of ductal obstruction. Oral ranulas in children are rare and the overall prevalence of mucocèles has been reported as 0.08% in children aged 0–12 years. However, there has been a documented increased occurrence in HIV-positive patients. This has been attributed to a blockage of the salivary gland by inflammation and peri-ductal fibrosis following HIV-associated salivary gland disease. Oral lesions may indicate infection with HIV and can also predict progression of HIV to AIDS. The most common oral manifestation is oral candidiasis occurring in 67% of children with HIV. Following this salivary gland disease, periodontal and gingival disease and herpes simplex are the next most common. The exact prevalence of ranulas in an HIV population is not known but the occurrence of a paediatric patient with HIV having at least one oral lesion has been documented as high as 63% and salivary gland disease at 50%. The true extent of the relationship between HIV and ranula is as yet unknown. This represents the only reported case of oral ranula in an HIV-positive patient in the UK.

BACKGROUND

Rarely seen in the UK. Not a well-known association between oral ranulas and HIV.^{1–4}

CASE PRESENTATION

A 12-year-old HIV-positive girl was referred for ear, nose and throat (ENT) assessment with a 2-year history of intermittent bilateral parotid pain and swelling, causing dysphagia. Occasionally the symptoms were exacerbated by gustatory stimulation, but could occur at almost any time of the day. There had been one episode of infective swelling which had been treated effectively with antibiotics. The patient, who moved to the UK from Zimbabwe in 2004, had been diagnosed and treated for HIV in the UK from 2005. She had been started on highly active anti-retroviral therapy 4 months prior to ENT assessment due to low CD4 count, though poor compliance with medication had been suspected.

On oral examination, a right-sided sublingual swelling was visible (figure 1). It was bluish in colour and was pushing the tongue forward. There were no other oral lesions visible.

INVESTIGATIONS

A magnetic resonance scan of the head and neck was requested and confirmed a well-defined lesion within the floor of the mouth on the right side (figure 2A,B). The scan signal intensities were consistent with a ranula and there was no evidence of it extending into the neck.

DIFFERENTIAL DIAGNOSIS

- ▶ Oral ranula
- ▶ Cyst

- ▶ Sialoadenitis
- ▶ Abscess.

TREATMENT

Following the scan, surgery was planned. The cyst was excised perorally with a small amount of sublingual salivary tissue. The ranula was limited to the oral cavity and did not descend into the neck. There was minimal bleeding and the postoperative period was uneventful. The tissue was submitted for histological examination.

The mass was an elongated, collapsed cyst measuring 2.6×1×0.6 cm, with a short, tubular segment of soft tissue. Microscopically, it consisted of hypocellular fibrotic and chronically inflamed tissue with numerous hyalinised granulomata (figure 3). These showed no necrosis or

Figure 1 The photograph demonstrates the sublingual swelling as seen in the clinic.

Figure 2 (A, B) MR scans (axial and sagittal T1-weighted images) showing a right-sided oral ranula.

casation. Occasional isolated multinucleate giant cells were only seen. Other areas showed fibrosis and extensive chronic inflammation of salivary gland tissue with marked atrophy of the acini and dilated excretory ducts. Occasional preformed cysts were also present which were filled by mucoid material and scattered desquamated cells. The appearances were concluded to be consistent with a cyst/mucocele with chronic and fibrotic sialoadenitis.

OUTCOME AND FOLLOW-UP

In this case, excision of the ranula with a small amount of salivary gland tissue was successful. There was no disease recurrence after 1 year of follow-up. The patient continues to be reviewed in the HIV clinic and with

improved compliance of medication her CD4 count has remained stable.

DISCUSSION

Ranula commonly presents as a gradually enlarging, unilateral, round or oval, bluish, soft tissue mass that is usually painless. The ranulas if large may cause medial and superior deviation of the tongue and can affect swallowing. Larger masses may cause obstruction of Wharton's duct leading to salivary outlet obstruction and submandibular swelling during eating. In this case, the patient presented with intermittent, unilateral swelling which was frequently painful.

There are two main types of ranula; simple (intraoral) and plunging (cervical). The simple ranula is confined to the floor of the mouth and is much more common. Diagnosis of oral ranula is usually clinical due to its characteristic appearance and location. The plunging ranula is in the infra mylohyoid compartment of the neck and may be confused with other causes of cervical swelling, particularly if there is no oral component.

Ranulas are most common in the second decade of life and slightly more common in women than in men.⁵ They are rare in children and overall prevalence of mucoceles has been reported as 0.08% in children aged 0–12-years.¹ In one study of paediatric patients in Zimbabwe, the number of patients with ranulas increased dramatically between 1999 and 2005.² Interestingly, 88.5% of children presenting with ranula were HIV seropositive. The authors implicated HIV-salivary gland disease as a predisposing factor for ranula in the paediatric population and proposed that ranula be considered another HIV/AIDS-associated lesion in Zimbabwe. This increase may be due to extensive fibrosis in mucus salivary glands or inflammatory tissue,⁶ or both, causing obstruction and

Figure 3 Heavy chronic inflammation of glandular tissue (right) with a wall of a cyst/ranula (left) showing multiple small pale hyalinised granulomata (inset).

hence mucocoele.⁷ Alternatively, the apparent increased incidence may instead be attributed to a greater number of patients seeking care at the study hospital during this time.⁸ The possible association between oral ranula and HIV has only been reported in one case outside of Zimbabwe. This single case was documented of a 7-year-old HIV-positive child with an oral midline ranula in North America in 1992.⁹ A number of other studies have reported the wide range of other oral manifestations of HIV/AIDS in different developing countries. These include oral Kaposi's sarcoma, also seen in Zimbabwe,¹⁰ cancrum oris in Zimbabwe,¹¹ South Africa,¹² Lesotho,¹³ Mali¹⁴ and Nigeria,¹⁵ and *Penicilliosis marneffei* in South East Asia.¹⁶ There is as yet no explanation for these regional differences.

Oral manifestations are often the earliest manifestations of HIV infection can also predict progression of HIV to AIDS. The WHO describe seven cardinal lesions: oral candidiasis, hairy leukoplakia, Kaposi's sarcoma, linear gingival erythema, necrotising ulcerative gingivitis, necrotising ulcerative periodontitis and non-Hodgkin's lymphoma which are all strongly associated with HIV infection.¹⁷ However, in children with HIV the most common oral manifestation is oral candidiasis occurring in 67% of children.⁵ Salivary gland disease, periodontal and gingival disease and herpes simplex are the next most common. The exact prevalence of ranulas in an HIV population is not known, but the occurrence of a paediatric patient with HIV having at least one oral lesion has been documented as high as 63% and salivary gland disease at 50%.⁴

There are a variety of methods used to treat ranula but surgery is the established treatment. Recommendations for the preferred approach and technique are variable and include marsupialisation, and excision of the ranula with or without excision of the sublingual and/or submandibular gland. Despite these treatments, many patients experience recurrence.⁵ In this case, excision of the ranula with a small amount of salivary gland tissue was successful and no disease recurrence was observed. A retrospective review of 864 cases by Patel *et al*,¹⁸ suggested that transoral excision of the ipsilateral sublingual gland with ranula evacuation yields the lowest recurrence and complication rates (13%). There is no reason to suspect that patients with HIV with ranula would not have a similar perioperative and postoperative course as a patient without HIV.¹⁹

Competing interests None.

Patient consent Obtained.

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Learning points

- What is an oral ranula?
- Potential association of ranulas and HIV.
- We report the first case of an oral ranula in an HIV-positive patient in the UK.
- Further work is needed to determine the link between these pathologies.

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