CASE REPORT

Simultaneous radicular cyst and mucoepidermoid carcinoma in the maxilla: a diagnostic nightmare

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SUMMARY

We present a case of a 20-year-old woman presenting initially with an asymptomatic palatal swelling. Radiographic examination showed a cyst at the right maxilla with bucco-lingual expansion and perforation of palatal bone. Incisional biopsy was carried out via a buccal approach and the result revealed a benign odontogenic cyst, in keeping with radicular cyst. The patient was then scheduled for cyst enucleation. During the procedure, it was found that the palatal lesion was unrelated to the maxillary cyst. Incisional biopsy of the palatal mass was carried out and revealed a low-grade mucoepidermoid carcinoma. The patient then had a partial maxillectomy with fibula flap reconstruction. There was no recurrence at postoperative 1 year follow-up and she was rehabilitated with dental implants.

BACKGROUND

Mucoepidermoid carcinoma (MEC) is the most common malignant tumour of the salivary gland.^{1 2} It accounts for 35–52% of malignant tumours in the minor salivary glands where the palate being the site of the highest frequency.^{1–3} It has a variable histopathological feature which is predictive of its highly variable clinical course.³ A low-grade MEC with features of prominent intracystic formation, minimal cell atypia and higher ratio of mucous cell has the best prognosis. High-grade counterpart meanwhile has minimal intracystic formation, prominent cell atypia and higher ratio of epidermoid cells. Surgery is the treatment of choice with adjunctive radiotherapy as indicated.³

Radicular cyst (RC) meanwhile is a benign inflammatory odontogenic cyst that is always related to at least one non-vital tooth.⁴ It will appear radiographically as a radiolucent lesion surrounding the root apex of affected teeth with the loss of lamina dura along the root. Root resorption and bone expansion with cortical perforation is not uncommon. A typical RC has a fibrous capsule lined by stratified squamous epithelium with inflammatory cell occasionally seen in the walls of the cyst.⁴ Enucleation combined with removal of the source by extraction or endodontic treatment usually will cure the disease.

The simultaneous occurrence of different pathology in an anatomically similar region of the jaws is rare. It is even more exceptional when one exhibits malignant growth while the other is benign. We report a case where two synchronous pathologies, one being a large simple cyst at the left maxilla and the other being an MEC at the left

palate, with highlights on achieving diagnosis in this difficult situation.

CASE PRESENTATION

A 20-year-old woman was referred to us with a swelling at her left palate for 1 month. She was previously seen by a general dentist and completed a course of antibiotics. She was later referred to us for the persistent swelling. Her general health was good and her medical history was unremarkable. Social history revealed that she had no smoking or drinking habit. General and extraoral examination did not reveal any abnormalities. On intraoral examination, there was bucco-palatal expansion of the alveolus in the upper left quadrant. The expansion extended from teeth 24 to 28. A soft palatal swelling with normal looking overlying mucosa was also noted adjacent to the alveolus at teeth 24–28 region and extended to the midline. There was mild tenderness on palpation. Teeth 24–28 were slightly mobile and were unresponsive to electric pulp test.

INVESTIGATIONS

Panoramic radiograph (figure 1) showed a well-circumscribed unilocular radiolucent lesion involving teeth 24–28. The sinus floor was displaced superiorly with a thin cortical bone separating the lesion and the antrum. The roots of teeth 24, 25, 26 and 27 were partially resorbed. Coronal view of the CT scan showed bucco-palatal cortical expansion with perforation of the palatal bone (figure 2). The findings from the radiographic examination seemed to suggest that a large cystic lesion in the maxilla had expanded the alveolar bone and perforated the palatal bone and caused the swelling in the palate.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses were cystic ameloblastoma, keratocystic odontogenic tumour and RC.

Figure 1 Preoperative panoramic radiograph showing a large radiolucent lesion at the left maxilla.

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Figure 2 Coronal section of the CT scan showed buccal and palatal bony perforation in left maxilla and the left maxillary sinus floor was elevated.

TREATMENT

Incisional biopsy was performed via the buccal approach (figures 3 and 4). Yellowish serous cystic fluid was noted (figure 5). A specimen of 10×10×3 mm was sent for histopathological examination. Microscopically, it was found that the cyst was lined by an attenuated simple-to-low stratified epithelium showing no ameloblastic features. Mild inflammation was noted beneath the epithelium (figure 6). The appearances were those of a benign odontogenic cyst, in keeping with RC. We concluded that the lesion was an RC related to the non-vital teeth with bucco-palatal expansion and palatal bone perforation as the cause of the soft palatal swelling.

Enucleation of the cyst was later performed under general anaesthesia. The cyst lining was thick and fibrous which was easily enucleated from the surrounding bone in one piece (figure 7). Further examination of the bony cavity confirmed the palatal perforation but the palatal swelling was found to be a separate lesion. Furthermore, an ulcer of 1 cm × 1 cm had been developed at the posterior part of the swelling. An incisional biopsy was performed immediately. Microscopic examination of the cyst lining revealed similar picture found during the incisional biopsy. The cyst was walled by fibrous tissue and bony rim of tissue. The inner surface was lined by stratified squamous epithelium focally. Moderately dense lymphoplasmacytic infiltrate was present in the granulation tissue where the inner surface was denuded of epithelial lining. Scanty goblet cells

Figure 3 Clinical photo of the palatal swelling during the first incisional biopsy.

Figure 4 Incisional biopsy of the cyst. Buccal bony perforation was noted. There were occasionally found at the squamous epithelium. There was no evidence of ameloblastoma or malignancy. The overall features were consistent with RC.

Microscopic examination of the palatal swelling revealed stratified squamous epithelium covered tissue with the stroma infiltrated by sheets and small nest of tumour cells formed by

Figure 5 Cystic fluid aspirated at the biopsy.

Figure 6 The cystic lesion was lined by stratified squamous epithelium. The subepithelial stroma consisted of inflammatory infiltrates (H&E, $\times 100$).

mucous cells, clear cells, intermediate cells and squamous cells (figures 8 and 9). A few small cysts lined by mucous cells were also noted (figure 10). The tumour cells were non-immunoreactive for myoepithelial markers (calponin, actin and S100 protein). Scattered p63+ cells were present. The stroma was sclerotic. Rare mitosis (less than 1/10 high power fields) was noted. There was no significant tumour necrosis and no perineural infiltration. The overall features were those of low-grade MEC. A second surgery was organised and she underwent left maxillectomy with fibula free flap reconstruction of left maxilla (figures 11 and 12). Final histopathological examination revealed a low-grade MEC showing focally infiltrative margin and comprised islands and sheets of tumour cells which included mucous cells, intermediate cells and epidermoid cells (figures 13–16). Dilated cysts that were lined by similar malignant cells were observed. The cystic component accounted for more than 20% of the tumour. Necrosis was absent. Cytologically, the nuclear morphology was quite uniform and featured open chromatin. Mitoses were very occasionally seen (less than 1/10 high power fields). Neural and lymphovascular invasion were not identified. Decalcification sections showed that the MEC permeated through the palatine bone but did not reach the superior cortical part. The maxillary antral mucosa showed patchy ulceration with proliferation of granulation tissue and marked active chronic inflammation but there was no evidence of malignancy.

Figure 7 Cyst enucleation in total.

Figure 8 The tumour comprised nests of epithelial cells of various morphologies (H&E, $\times 100$).

OUTCOME AND FOLLOW-UP

The patient recovered well and was discharged from the hospital 10 days after the surgery. She received implant rehabilitation at postoperative 1 year (figures 17–20).

DISCUSSION

The hard palate is a highly specialised region of the mouth. It is composed of an orthokeratinised epithelial lining and it is firmly adhered to the underlying bone. The postero-lateral part is slightly thicker and soft owing to the numerous nerves, vessels and minor salivary glands at the submucous level. Any pathology arising from the bone, salivary gland, vessels, nerves, connective tissue and epithelium can be the cause of the palatal swelling. In addition, pathology arising from adjacent dental tissue or antrum can cause palatal swelling by expansion of the bone. A prospective study of aetiopathology of maxillary swelling by Biswas and Cranck⁵ found that the most common non-neoplastic cause was odontogenic cyst while squamous cell carcinoma as the most common neoplastic cause. There are numerous possible causes of palatal swelling and it is difficult to make the diagnosis especially if two pathologies with same clinical signs occur simultaneously.

Both RC and MEC may present as a palatal swelling. In MEC, the swelling is usually soft and may appear bluish. Ulceration is common in high-grade variant. While in the RC,

Figure 9 Some tumour cells adopted a squamoid appearance (H&E, $\times 200$).

Figure 10 Mucous cells with abundant apical cytoplasmic mucin were present. Nests and cords of intermediate cells were observed at the periphery (H&E, ×200).

swelling is usually bony hard owing to expansion of the palatal bone. The bone can occasionally be perforated and results in a soft swelling in the hard palate. Ulceration due to the RC itself is uncommon but traumatic ulcer in the palatal swelling is possible due to mastication and swallowing. When different pathologies with shared clinical pictures occur synchronously, diagnosis could be difficult. In the presented case, the presence of a soft palatal swelling was a shared feature of RC and low-grade MEC. This caused difficulty in identifying a second pathology when bucco-palatal expansion and palatal perforation was shown on the CT scan. The ulcer only developed after the first biopsy. The initial diagnosis based on palatal swelling corresponds with the features of RC clinically and radiographically.

The occurrence of simultaneous malignant and benign pathology in the head and neck region is uncommon. More commonly reported are malignant transformation of a benign lesion such as thyroid carcinoma from thyroglossal cyst,⁶ squamous cell carcinoma from brachial cyst,⁷ carcinomas arising from odontogenic cyst or tumour.⁸⁻⁹ It is even more exceptional to find occurrences of benign and malignant pathologies within the same anatomic region synchronously. Previously reported synchronous occurring malignant and benign pathology within the same region mostly involved the major salivary glands.¹⁰ The rarity for the occurrence of cases as reported here warranted a literature review as to search for other possible explanation for such event. We searched in the literature in an attempt to identify any pathology that could possibly be indistinguishable clinically and histologically to a synchronously occurring RC and an

Figure 12 Immediate reconstruction of fibula free flap.

MEC in a similar site. It has been reported that both the controversial glandular odontogenic cyst (GOC) and central MEC (CMEC) could have clinical features similar to the reported case such as swelling or bony expansion, cortical perforation and also cystic radiological presentation.¹¹⁻¹² Histopathologically, if an RC presents with mucous metaplasia, it would be difficult to rule out GOC or CMEC because of the similar features.¹¹

However, the lesions reported here were unlikely to be the presentation of a single pathology such as GOC or CMEC because of the following reasons. First, both lesions were evidently not connected because of the ease in separating the cyst lining from the bony cavity and palatal tissue at the perforated area thus excluding it as a single lesion. Second, the final histopathological examination of the MEC confirmed the absence of bony perforation at the posterior palate where the tumour was located confirming it to arise from the minor salivary gland within the palatal mucosa but not the bone thus ruling out CMEC. Most importantly, the microscopic examination of the cyst lining following the enucleation did not demonstrate the characteristics of GOC or CMEC.¹¹ No mucous metaplasia was seen making diagnosis of RC straightforward.

In retrospect, we believe a number of elements that could have helped us in differentiation both lesion early on in this case. By highlighting these factors, it is hoped that clinician encountering such difficulties would be able to rule out such occurrences. First, we should be aware of the possibility of having synchronous lesions with similar clinical and radiological features although it is extremely rare. There has been no similar occurrence reported

Figure 11 Partial maxillectomy.

Figure 13 The tumour formed sheets of cohesive cells with a prominent cystic component. Necrosis or neural invasion was absent (H&E, ×40).

Figure 14 Areas showing squamoid cells along the cystic lining (H&E, ×100).

Figure 15 Mucous cells were observed displaying abundant apical mucin (H&E, ×200).

in the literature; hence, this report would hopefully be able to raise the index of suspicion among fellow surgeons and clinicians. From the clinical aspect, thorough and careful evaluation of CT scan should be carried out to assess the thickness of the

Figure 16 Tumour nests with mucous cells, squamoid cells and intermediate admixing with one another. Anaplasia was not observed. The mitotic count was less than 1/10 high power fields (H&E, ×200).

Figure 17 Panoramic radiograph taken at postoperative 1 year.

Figure 18 Postoperative 1 year.

Figure 19 Implant rehabilitation at postoperative 1 year.

Figure 20 After implant placement.

palatal mucosa to identify any unusual soft tissue growth. Thin slices (1 mm) of CT scan should be used for evaluation. Any increase in thickness should raise the suspicion of a second pathological lesion. By relying on clinical examination, based on our experience, it was almost impossible to differentiate the posterior part of the palatal swelling was of salivary gland origin due to the diffuse margin between both causes of swelling.

Learning points

- ▶ Clinicians should be aware of the possibility of having synchronous lesions with similar clinical and radiological features although it is extremely rare.
- ▶ When investigating a palatal swelling using a CT scan, thin slices (1 mm) should be used for evaluation to assess the thickness of the palatal mucosa to identify any unusual soft tissue growth.
- ▶ Any increase in thickness should raise the suspicion that a pathology originated from the palate, rather than from an expansile lesion in the maxillary sinus.

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