Mucoepidermoid Carcinoma of a Nasal Cavity – a Rare Tumour

Mukoepidermoidní karcinom dutiny nosní – vzácný nádor

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Summary

Backgrounds: Mucoepidermoid tumours arise from the ductal cells of the salivary glands, most commonly the parotid. The occurrence of these tumours in the mucus glands of the air passages is extremely rare. They are very aggressive tumours with poor prognosis. ***Case:*** A case of nasal mucoepidermoid carcinoma with probable origin from the middle turbinate is reported. ***Conclusion:*** Nasal mucoepidermoid carcinomas are extremely rare. They usually present with symptoms similar to sinusitis. When dealing with a recurrent/aggressive tumour in the ethmoids, the possibility of mucoepidermoid carcinoma can be considered in the differential diagnosis. They are aggressive tumours with a poor prognosis.

Key words

mucoepidermoid carcinoma – nasal cavity – turbinates – epistaxis

Souhrn

Východiska: Mukoepidermoidní nádory vznikají z duktálních buněk slinných žláz, nejčastěji parotis. Výskyt těchto nádorů v mukózních žlázách dýchacích cest je velmi vzácný. Jedná se o agresivní nádory se špatnou prognózou. ***Případ:*** Je uváděn případ mukoepidermoidního karcinomu dutiny nosní, který pravděpodobně vznikl ze střední skořepy. ***Závěr:*** Mukoepidermoidní karcinomy dutiny nosní jsou velmi vzácné. Obvykle mají obdobné příznaky jako sinusitida. Objeví-li se recidivující/agresivní nádor v kosti čichové, lze v diferenciální diagnostice uvažovat o možnosti mukoepidermoidního karcinomu. Jedná se o agresivní nádory se špatnou prognózou.

Klíčová slova

mukoepidermoidní karcinom – dutina nosní – střední skořepa – epistaxe

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Introduction

Although the existence of mucoepidermoid tumours of the salivary glands has been known since 1895, it became a well defined entity only in 1945. The original authors felt mucoepidermoid tumours could be either benign or malignant [1]. In the light of later evidence, it became evident that so-called benign tumours exhibited aggressive clinical behaviour. So the more appropriate name of mucoepidermoid carcinoma was widely accepted [2].

Mucoepidermoid tumours arise from the ductal cells of the salivary glands [3]. They represent 5–8% of all salivary gland tumours [2,4]. Of all anatomical sites, the parotid is the most common, followed by a much lower incidence in the palate [4]. Glands of similar architecture are found in the nasal cavity, paranasal sinuses, mouth and pharynx, but occurrence of this tumour in them is rare [2–4]. They have been variously reported in the nasal cavity [5], ethmoid [2,3], larynx [5], maxilla [6,7] and mandible [8]. Only a handful of such reports exist in literature. A probable origin from the middle turbinate (as in our case) has not been previously reported.

Case Report

A 48-year-old male patient presented to the outpatient department with complaints of headache, facial pain and nasal obstruction that had lasted for 10 days. Clinical examination showed a purulent discharge in the left middle meatus and pain over the left maxillary sinus. Radiological examination suggested left maxillary sinusitis. He improved with antibiotics and nasal decongestants but came back with complaints of increasing nasal obstruction two weeks later. Rhinoscopy showed an intranasal polypoidal mass arising from the middle meatus of the left side. The patient underwent endoscopic intranasal polypectomy. Histopathology of the excised mass was reported as a polyp lined with respiratory epithelium with dense eosinophilic infiltrate. The patient continued to be symptomatic for nasal obstruction and rhinorrhoea, which persisted despite therapy. Five months later, the patient was re-evaluated due to the development of bloodstained nasal discharge along with persistence of the previous symptoms. Examination showed a mucosa-covered swelling arising from the middle turbinate. A provisional diagnosis of a left inverted papilloma of the middle turbinate was made. He underwent repeat endoscopic removal of the intranasal mass. Histopathology showed an infiltrating neoplasm with a glandular and nest-like pattern. The glands had a signet ring cell lining with mild to moderate dysplastic nucleus. The stroma showed necrosis and inflammatory infiltrates. Stromal infiltrate showed polygonal to spindle cells with hyperchromatic, anaplastic nuclei with many abnormal mitotic figures and scanty cytoplasm (Fig. 1). Histopathological features were suggestive of a high-grade mucoepidermoid carcinoma.

The patient did not appear for follow-up for six months. He presented again with a history of recurrent bouts of epistaxis and was admitted with a severe nasal bleed. Clinical examination showed a residual tumour only around the middle turbinate. CT evaluation of the Ostio Meatal Complex (OMC) showed a residual soft tissue density around the region of the left middle turbinate and the adjoining part of the septum (Fig. 2, 3). The maxillary sinuses showed bilateral mucosal thickening. The rest of the nose was normal. The patient refused the option of surgery as he did not want a postoperative facial scar. Other modalities of treatment were also refused despite counselling.

He again did not appear for follow-up for 6 months till he presented with massive epistaxis, proptosis and headache. Contrast-enhanced CT scan of the OMC showed a heterogeneous enhancing

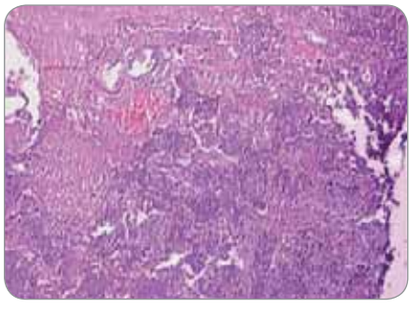

Fig. 1. Microphotograph showing mucous glands and malignant squamoid elements. H & E × 120.

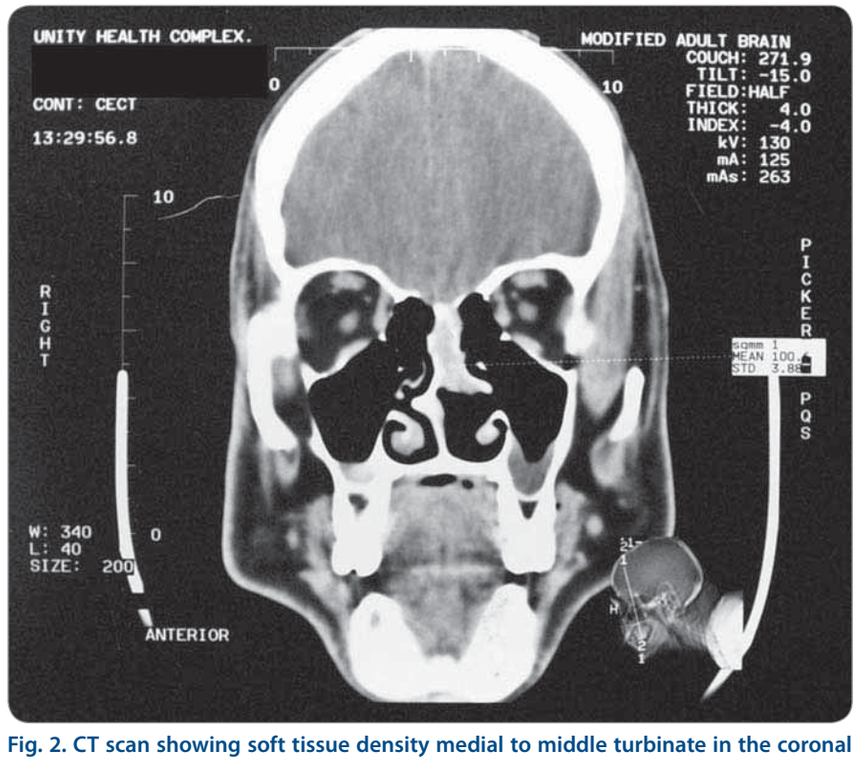

Fig. 2. CT scan showing soft tissue density medial to middle turbinate in the coronal plane.

tumour completely occupying the left nasal cavity with extension into the left frontal, ethmoidal, maxillary and sphenoid sinuses. There was an extension into the medial half of the left orbit, cribriform plate, left frontal lobe, septum, right nasal cavity, right ethmoids and the medial wall of the right orbit (Fig. 4). MRI confirmed intracranial and orbital involvement (Fig. 5). Oncological treatment was commenced with 200 cGy of radiotherapy with lateral opposed fields. After the first dose, the patient developed cerebral oedema and convulsions of a generalized tonic clonic type. He expired of cardio-respiratory failure two days later.

Discussion

Of the malignancies affecting the sinonasal region, the vast majority are squamous cell carcinomas. Malignancies of mucous gland origin constitute 4% of all tumours, of which adenoid cystic carcinoma is the most frequent [9,10].

Although mucoepidermoid carcinoma is the commonest malignant tumour in the salivary glands, the reason why the respiratory tract is immune to these tumours is unknown [11]. Fewer than 40 cases have been described to date [12]. In the sinonasal tract, it is most common in the maxilla, followed by decreasing incidence in the nose, nasopharynx and ethmoids [12].

No definite etiology/risk factors have been identified for sinonasal mucoepidermoid carcinoma. The most common symptoms are nasal obstruction and epistaxis [3,5,12]. Radiological investigations like CT and MRI are useful in detecting early tumours and possible intracranial extension.

One common feature which has been seen in previously reported data and in our case is the highly aggressive nature of the lesion. One peculiarity noted in our case was the behaviour of the tumour. The tumour was fairly quiescent for 6 months from the time of diagnosis, as was seen by the CT scan done 6 months after the diagnosis. Over the next 6 months, the tumour turned very aggressive and grew to massive proportions.

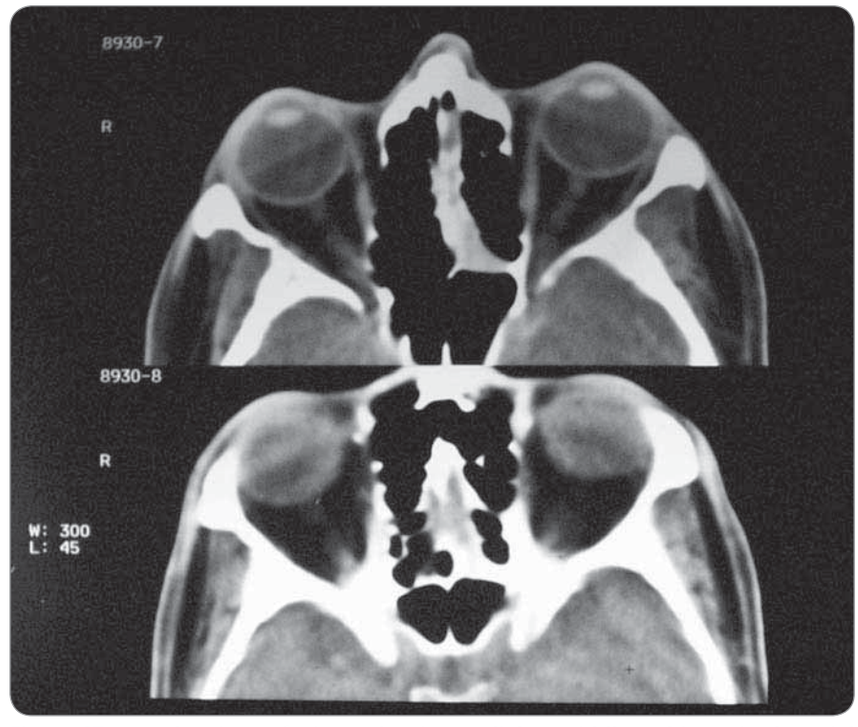

Fig. 3. CT scan showing soft tissue density medial to middle turbinate in the axial plane.

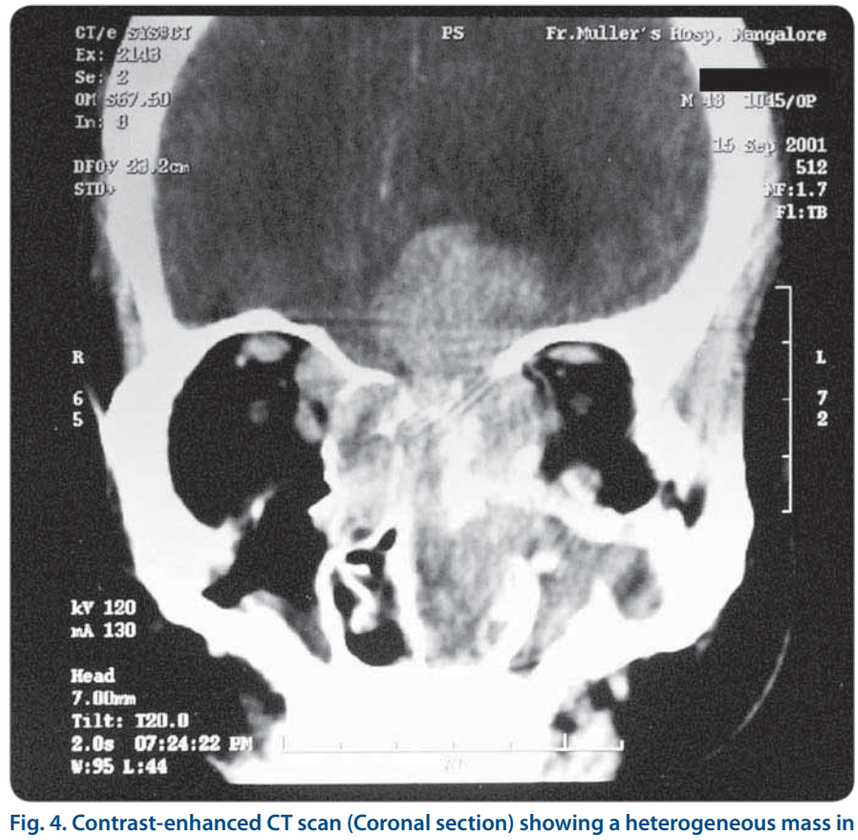

Fig. 4. Contrast-enhanced CT scan (Coronal section) showing a heterogeneous mass in the entire left nasal cavity and part of the right nasal cavity, with intracranial and orbital invasion.

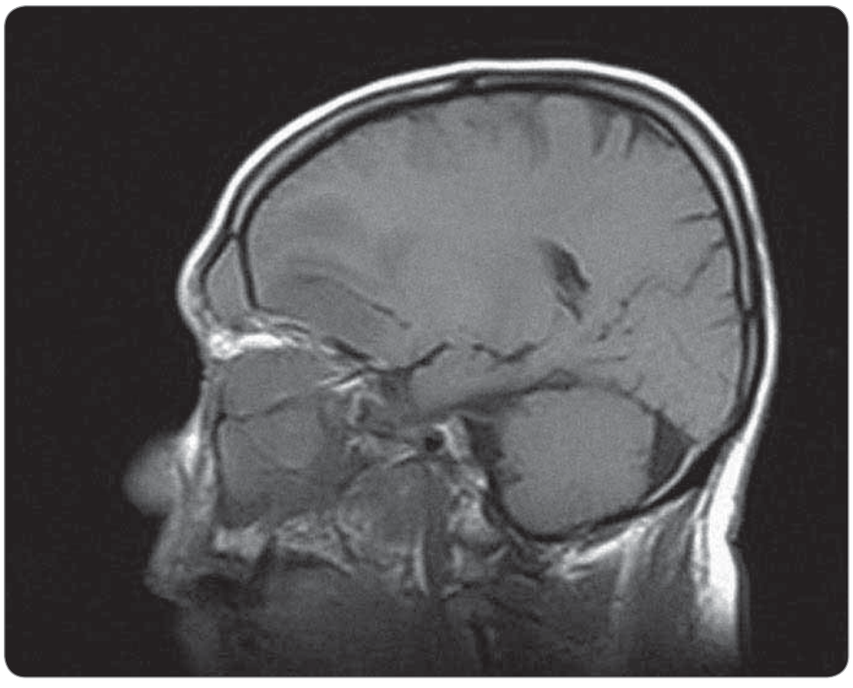

Fig. 5. MRI scan in the sagittal plane show-ing intranasal and intracranial spread of the tumour.

In the sinonasal region, where other pathologies are expected, mucoepidermoid carcinoma can be missed by the pathologist [2,5,13]. Extensive spread before diagnosis appears to be the rule rather than the exception [3]. An early lesion confined to the middle turbinate has not been described before, to the best of our knowledge.

The treatment depends on the tumour grade, extent of tumour invasion and the condition of the patient [12]. Low-grade tumours can be managed by surgery alone. Combined therapy with surgery and radiotherapy are needed for intermediate and high grade ones. High-grade tumours generally indicate poor prognosis.

Even with radical attempts at treatment, recurrence is common [5,7,12] and in several cases death also [5,12]. The one previous report which did not have recurrences/death [3] failed to mention the grading of the tumour. The author may have dealt with a low-grade tumour variant. Considering the absence of a large series on this particular tumour and the rarity of its occurrence, we are still far from understanding and managing it with best results.

Conclusion

Nasal mucoepidermoid carcinomas are extremely rare. They usually present with symptoms similar to sinusitis. When dealing with a recurrent/aggressive tumour in the ethmoids, the possibility of mucoepidermoid carcinoma can be considered in the differential diagnosis. They are aggressive tumours with a poor prognosis.

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