e-ISSN: 2957-6717

Case report

Hyalinizing Clear Cell Carcinoma of Soft Palate and Nasopharynx treated with wide local excision: a case report

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Abstract

Hyalinizing clear cell carcinoma is an uncommon tumour usually found near the base of the tongue and palate and manifests as a painless submucosal swelling. It has frequently been mistaken for other, more prevalent tumours with clear cytoplasm, like mucoepidermoid carcinoma, acinic cell carcinoma, or clear cell oncocytoma. According to reports, hyalinizing clear cell carcinoma is a low-grade malignant tumour that frequently spreads to the cervical region. There is no treatment strategy because it is so uncommon. Wide local excision is the preferred treatment, and neck dissection, radiation therapy, or both are used to treat the condition. However, because of the too-low incidence or underreporting and absence of long-term follow-up, there is no definitive result. Our case emphasizes the challenges associated with diagnosing such uncommon cases and the necessity of a long period of post-excision follow-up to assess the treatment's outcome. **Keywords**. Hyalinizing Clear Cell Carcinoma, Minor Salivary Glands, Nasopharynx, Palate.

Received: 04/01/25 **Accepted**: 03/03/25 **Published**: 13/03/25

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Introduction

Under ten percent of head and neck cancers are hyalinizing clear cell carcinomas, which are uncommon, malignant tumours. This type of tumour, especially in females, frequently affects the minor salivary glands. Typically appears in patients who are in their seventh decade. The palate and base of the tongue are the most common sites, followed by the floor of the mouth, buccal mucosa, lip, retromolar trigone, and other sites of minor salivary glands. It is a low-grade malignant salivary gland tumour with a good long-term prognosis. We describe a case of hyalinizing clear cell carcinoma of the soft palate and nasopharynx that was effectively treated with a wide local excision and has not shown any postoperative complications [1]. The optimum treatment strategy is not well supported by the published literature because of the tumour's rarity.

Case report

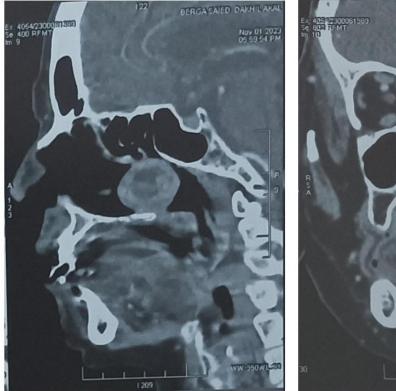
A 65-year-old female patient presented to the out-patient clinic complaining of swelling in her palate on the right side, causing her right nostril to be blocked and causing difficulty in breathing. Intraoral examination revealed the presence of a 2x2 cm mass lesion that began on the right side of the hard palate and extended to the soft palate, crossing the midline deeply into the left side (Figure 1). Physical examination and neck ultrasound did not reveal pathological lymphadenopathy.

Computed tomography scan study revealed the presence of an infiltrative soft tissue mass centered upon the lower part of the nasal cavity and inseparable from the lower nasal septum. The mass measures about 6x4x5 cm at its maximum CC and axial diameters, respectively, with a central cystic area of breakdown; there is heterogeneous enhancement. Both maxillary, frontal, right ethmoidal, and sphenoid sinuses (Figure 2). In addition, the computed tomography scan showed attenuated left and aerated right osteomeatal units. Patent aerated rest of nasopharynx and oropharynx.





Figure 1. Intraoral photograph of the patient, showing 2x2cm well defined focal lesion at the right side of the soft palate, at the junction of the hard and soft palate.



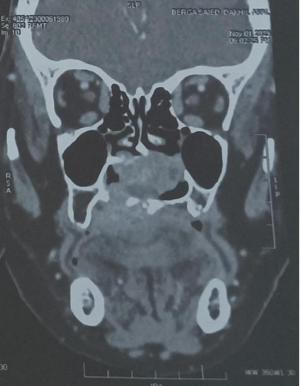


Fig. 2: Computed tomography scan showed presence of an infiltrative soft tissue mass centred upon the lower part of nasal cavity. Sagittal and coronal views.

Magnetic resonance imaging (MRI) showed a well-defined soft tissue intensity-enhanced mass centered on the posterior nasal septum, posterior nasal cavities, and anterior aspect of the nasopharynx with central necrosis. No invasion of maxillary antra. No intracranial perineural spread. It measured about (36x35x34 mm) (Figure 3). No enlarged nodes.



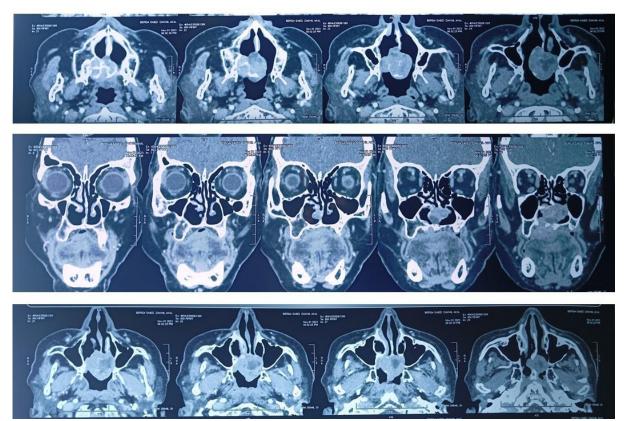


Figure 3: Magnetic resonance imaging showed a well-defied soft tissue intensity enhanced mass cantered on the posterior nasal septum, posterior nasal cavities and anterior aspect of nasopharynx with central necrosis.

Incisional biopsy histopathological examination showed multiple small fragments of respiratory epithelium with underlying fibrous stroma containing few small lobules of seromucinous salivary acini, with wide non-encapsulated malignant epithelial cell infiltration arranged in variable patterns composed of compact trabeculae, thin cords, some tubules, and solid sheets, surrounded by hyalinized stroma with focal mucoid-myxoid areas. The tumor cells displayed moderate pale-eosinophilic cytoplasm and oval monotonous vesicular nuclei with fine chromatin pattern and small nucleoli. No increased mitotic activity, cellular pleomorphism, or tumor necrosis seen. No perineural invasion or lymphovascular invasion is seen. The tumor infiltrates into the adjacent bone trabeculae. Diagnosis: right nasal fossa soft tissue mass incisional biopsy: malignant salivary gland tumour, consistent with polymorphous low-grade adenocarcinoma.

The diagnosis and surgical treatment were explained to the patient, but she refused to undergo the surgery and disappeared for a long time. She came back to our clinic complaining of nasal bleeding with increased difficulty in breathing. Nasal examination and laryngeal scope examination showed a protruding mass in the nasal floor (Figure 4).

The patient underwent surgery with a 1 cm safety surgical margin, the mass lesion was removed, and no flap was done to reconstruct the surgical defect. Briefly, under general anesthesia, in the strictest aseptic and sterile measures, the patient was readied for surgery. The Weber-Ferguson approach and right maxillary incision were performed. The palatal lesion was outlined, and the mucosa was excised with a safe margin of 1 cm. The palatal bone and the right lateral wall of the maxilla were resected using a surgical saw. With the aid of a pterygoid chisel, the pterygomaxillary disjunction was done, and the lesion was detached from the rest of the maxilla (Figure 5). A surgical acrylic stent was fixed on the palate with the help of surgical screws to facilitate post-operative healing and feeding (Figure 6).





Figure 4: Nasal examination showed protruding mass in the nasal floor

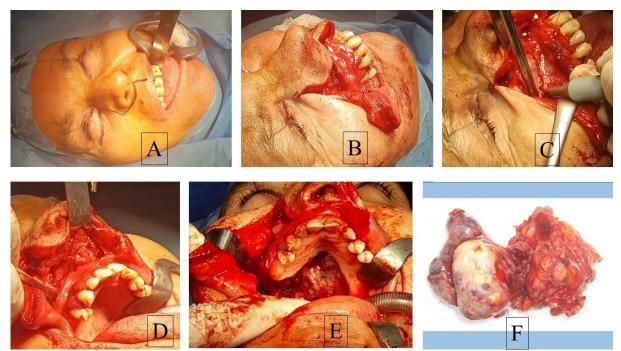


Figure 5: Intraoperative clinical pictures. A) Outlining of Weber Ferguson approach. B) Weber Ferguson approach done. C and D) Resection of lateral wall of maxilla.). E) The defect site after excision. F) The resected tumoral mass.





Figure 6: Intraoperative clinical picture showing an acrylic stent fixed in its place with screws.



Figure 7: Intraoperative clinical picture showing closure of operation field. The different incisions are stitched and secured in place by continuous subcutaneous interlaced suture technique.

Histopathological study of the excised tumoral mass showed tumor cells arising from minor salivary glandular tissue composed of solid sheets and trabecular cords of large, monomorphic clear cells infiltrating the surrounding skeletal muscle fibers. Numerous bands of intervening hyalinized stroma are seen, and there was glycogen within the tumor cells. Accordingly, the lesion diagnosed as hyalinizing clear cell carcinoma of minor salivary glands of the palate. All the margins of the resected palate and naso-sinus



maxillary tissues were tumor-free except the lateral maxillary margin that was invaded by the tumor. In addition, this histopathology examination revealed evidence of lymph vessel invasion but did not show evidence of blood vessel invasion or evidence of preneural invasion. Accordingly, chemoradiotherapy for the area of the lesion was recommended.





Figure 8: Clinical postoperative picture of the surgical site 2 months postoperatively, stent removed, showing good epithelization with no signs of infection or recurrence.

Discussion

The palate and base of the tongue are the primary areas affected by hyalinizing clear cell carcinoma, which most frequently develops in middle-aged women's minor salivary glands in the oral cavity [1]. Less often occurring locations include the parotid gland, subglottic larynx, nasopharynx, and hypopharynx [2].

The yearly incidence of all salivary gland tumours is between 0.4 and 13.5 cases per 100,000 people worldwide. In the United States, salivary gland tumours make up 0.3% of all malignancies and 6% of all head and neck cancers. (3) Malays are more likely than Chinese and Indians to have salivary gland tumours. (3) Minor salivary glands are the source of half of the most malignant salivary gland tumors [3].

Unless subsequent trauma occurs, the clinical appearance is a gradually expanding, submucosal bulge in the oral cavity without pain or ulcers. Additional symptoms include bleeding from the swelling, pain near the swelling after swallowing, or from dentures that are loose. Individuals who have lesions at the base of their tongues present with a feeling of a foreign body in their throat. According to Milchgrub et al., neck dissection revealed cervical node metastases in 2 out of 11 cases [1].

Hyalinizing clear cell carcinoma can metastasize to locoregional lymph nodes and is typically infiltrative. Perineural invasion is frequently observed [1]. This tumor's gradual growth is one of its key characteristics; as a result, it can be classified as a low-grade malignant tumor. It is distinguished histologically by a monomorphous population of polygonal to round cells with transparent cytoplasm when stained with conventional hematoxylin and eosin. Tumor cells lack ductal structures and have a high glycogen content, and they are organized in solid cell masses, sheets, nests, cords, or trabeculae [1,3]. Thick hyalinized collagen bands make up the tumor's stroma. Acinic cell carcinoma, clear cell oncocytoma, mucoepidermoid carcinoma, epithelial-myoepithelial carcinoma, sebaceous carcinoma, metastatic renal cell carcinoma, polymorphous low-grade adenocarcinoma, pleomorphic adenoma, and Pindborg tumor are among the other tumors with clear cytoplasm that are misidentified as hyalinizing clear cell carcinoma [1,3,4].

By assisting in the identification of the tumor's cell of origin, immunohistochemical staining facilitates diagnosis. PAS is positively stained; however, mucin is negatively stained. The cells react with antibodies against the antigens of the epithelial membrane and cytokeratins but not with the antigens of the S-100 protein or smooth muscle actin. Once additional distinct tumors with distinct cell morphology have been ruled out, the diagnosis is made by exclusion [4]. Immunoreactivity to vimentin and S-100 protein is seen in myoepithelial cells. Actin unique to muscles and the S-100 protein are both positive in epithelial-myoepithelial carcinomas.



Hyalinizing clear cell carcinoma is uncommon and lacks a well-defined treatment plan. Staging, however, adheres to the American Joint Committee on Cancer's (AJCC) eighth edition TNM Classification of Oral Cavity. There have been cases of metastases to the neck and lung, despite the tumor being classified as a low-grade malignancy. Wide local excision is the preferred treatment for hyalinizing clear cell carcinoma since it is a tumor with little potential for malignancy. Nevertheless, a high risk of cervical lymph node metastasis was discovered in a study of twenty-five-year-olds done by Solar AA et al., which underscored the significance of a comprehensive evaluation of cervical lymph nodes. Neck dissection ought to be taken into account in the treatment of hyalinizing clear cell carcinoma. There have been reports of radiotherapy before or after surgery, and some recurrences have been documented even after post-operative radiation (5). In a study of eight patients, Solar AA et al. found that three of them experienced recurrence, and one of them had received post-operative radiation [2]. Given the low incidence and the possibility of underreporting due to little follow-up, this may not be typical.

The authors discovered that the incidence is quite low when comparing the earlier articles; therefore, there is not any discernible trend. Even the age group has a median of 57 and ranges from 25 to 86. There is also a gender preference; according to the research, seventy-one percent of hyalinizing clear cell carcinoma cases occur in females [2].

Conclusion

Hyalinizing clear cell carcinoma Salivary gland is regarded as an uncommon malignant tumour. It is an asymptomatic, slowly developing tumour that is frequently observed in middle-aged women over 50. We reported a rare case of clear cell carcinoma of the palate in a woman aged 65 years that was treated with wide local excision only, with encouraging postoperative outcomes. This case report aims to contribute to the body of literature by presenting the presentation and long-term results of this uncommon entity. By raising awareness of this uncommon condition, fewer clear cell tumours will be misdiagnosed. All clinicians should be actively urged to perform thorough intraoral examinations regularly to screen for early asymptomatic developing lesions that may go undetected until they become larger, require extensive treatment, or provide a challenging prognosis. Clinically, this type of tumor can be mistaken for other benign neoplasms, making diagnosis difficult. However, histologically, there are multiple differential diagnoses for clear cell carcinoma.

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