JCOMS eISSN:2783-3127 https://journalofcoms.com

Journal of

Current Oncology and Medical Sciences

Vol. 5, No. 3

Case report Free Access

Metastatic carcinoma-ex pleomorphic adenoma of the pharynx with carotid space invasion: a rare case report

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Abstract

Introduction: Carcinoma ex pleomorphic adenoma (CXPA) is a rare, aggressive malignancy of the salivary glands that arises from a pre-existing pleomorphic adenoma. Although pleomorphic adenomas are benign, their potential for malignant transformation necessitates timely diagnosis and management. Pathological assessment remains the gold standard for diagnosis, with surgery followed by radiotherapy being the standard of care.

Case Presentation: A 38-year-old male presented with a metastatic carcinoma ex pleomorphic adenoma of the pharynx, with invasion into the carotid space. Diagnosis was confirmed histopathologically. The patient underwent palliative radiotherapy.

Discussion: CXPA is often difficult to diagnose due to its overlapping features with benign tumors, especially in atypical locations such as the pharynx. Malignant transformation typically indicates a more aggressive clinical course, including local invasion and distant spread. In this case, carotid space involvement further complicated management, highlighting the importance of early detection and comprehensive treatment.

Conclusion: Early recognition of pleomorphic adenomas and their potential for malignant transformation is critical. This case emphasizes the need for a multidisciplinary approach in diagnosing and managing rare presentations of CXPA to improve patient outcomes.

Keywords: Carcinoma ex-pleomorphic adenoma, Recurrent pleomorphic adenoma, Carotid space invasion

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Received: 2025.7.24, Accepted: 2025.9.25

Introduction

Salivary gland tumors are uncommon, accounting for only 3 %–10 % of head and neck neoplasms. They can arise in the major salivary glands (parotid, submandibular, and sublingual) or in the minor salivary glands (small, predominantly mucus-secreting glands located beneath the mucosal lining of the upper aerodigestive tract, such as labial, lingual, palatal, buccal, glosso-palatal, and retromolar glands) (1, 2).

Carcinoma ex-pleomorphic adenoma (CXPA) is a carcinomatous transformation within a primary (de novo) or recurrent pleomorphic adenoma of a salivary gland, most commonly found in the parotid gland. However, it can also originate in the submandibular gland and rarely in minor salivary glands located in the hard and soft palate. Tumors arising from minor salivary glands are typically smaller in size compared to those from the major salivary glands, and the incidence is less than 7%. In addition to salivary glands, CXPA has also been identified in lacrimal glands, nasal cavities, trachea, and breast (3, 4). Here, we present a case of recurrent pleomorphic adenoma of the soft palate that ultimately transformed into CXPA of the pharynx with carotid space invasion and distant metastasis.

Case presentation

A 38-year-old male patient initially presented in his native country, Zambia, in June 2006 with a swelling on the left side of the soft palate. The swelling was excised, and histopathology revealed pleomorphic adenoma (PA). However, he developed recurrences in 2012, 2015, and 2016, each of which was surgically re-excised, with histopathology confirming PA in all instances. In December 2018, he presented with an extensive and inoperable local recurrence, and he underwent palliative radiation therapy (30 Gy in 10 fractions over 2 weeks) in Zambia. He was later referred to our center for further evaluation and treatment.

On local examination, the patient exhibited a large, irregular lesion in the left oral cavity, measuring approximately 6 × 5 cm. The mass extended into the right side of the oral cavity, including the right tonsillar pillar, and inferiorly involved the base of the tongue. Superiorly, the lesion extended to the soft and hard palate. Laterally, it involved the retromolar space and left tonsillar pillar. Palpation revealed enlarged left level IA and IB lymph nodes. The patient had difficulty closing his mouth and complained of mid-back pain, which required analgesic medication. Neurological examination was unremarkable.

A whole-body PET-CT was performed, revealing a large, necrotic mass involving the left pharyngeal space, obliterating the posterior nasopharynx, and extending into the supraglottic region with luminal obstruction. Superiorly, the mass extended into the left infratemporal fossa and carotid space. Posteriorly, the lesion thinned the wall of the left maxillary sinus, medially indenting the soft palate and the posterior one-third of the tongue. The lesion showed moderate to intense heterogeneous uptake (SUVmax: 14.6). Bilateral sub-centimeter nodules were seen in the lungs with minimal FDG uptake (SUVmax: 3.8), consistent with bilateral lung metastasis. Additionally, FDG-avid lytic lesions were observed in the D11 and L3 vertebrae with soft tissue involvement (SUVmax: 9.5 in L3 vertebra) (Figure 1).

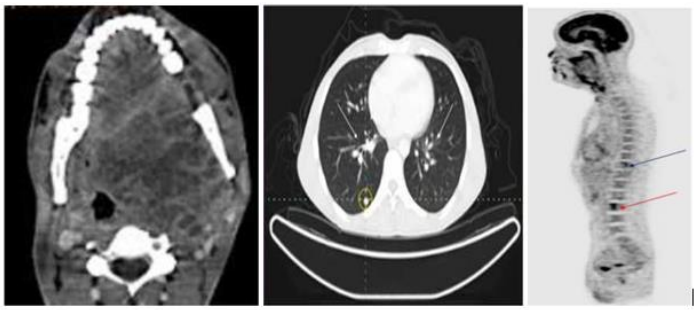

Figure 1. PET CT: A: Axial view of PET scan showing extent of the primary tumor; B: Axial view of PET scan showing distant metastasis to bilateral lungs; C: Sagittal view of PET scan showing lytic lesions at D11 (blue arrow) and L3 (red arrow).

Histopathological findings from a biopsy of the left retromolar trigone were consistent with CXPA (Figure 2).

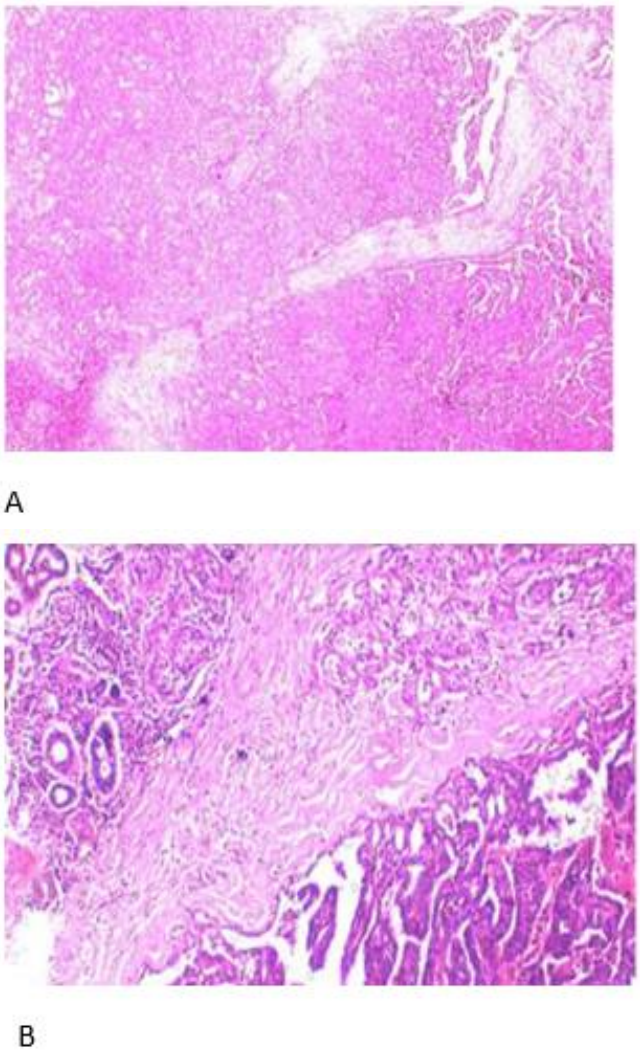

Figure 2. Hematoxylin and Eosin:A: Low-power view of salivary gland tissue infiltration by atypical ductal cells forming glands. Cribriform pattern present; B: High-power view showing capsular invasion by atypical ductal cells.

Immunohistochemistry (IHC) findings showed that GFAP was positive in tubules with scattered focal loss. HMWCK, p63, vimentin, and CK7 were diffusely positive. S100 stained myoepithelial cells. The MIB1 index was 15-20% on hot spots. Her2-neu was negative. myoepithelial cells. The MIB-1 index was 15-20 % on hot spots. Her2/neu was negative. Additionally, the left level IB lymph node biopsy showed features consistent with a chondroid-forming myoepithelial-rich lesion, favoring a recurrence of pleomorphic adenoma with extensive myoepithelial areas.

Given the extensive local recurrence, metastasis, and overall disease burden, surgery was not considered feasible. Radiation therapy to the primary site (oropharynx) was not offered due to the short intervals between prior treatments. The patient was therefore planned for stereotactic body radiotherapy (SBRT) to treat the symptomatic metastatic vertebral lesions at D11 and L3. SBRT with 6 MV photons was administered to the D11 vertebra to a total dose of 27 Gy in 3 fractions (9 Gy per fraction) and to the L3 vertebra to a total dose of 18 Gy in a single fraction. After three cycles of chemotherapy with paclitaxel and carboplatin and SBRT, the patient showed stable disease on follow-up PET-CT imaging. He reported symptomatic improvement, including pain relief, during a short 6-month follow-up period. However, the patient did not return for further follow-up after this period.

Discussion

CXPA accounts for approximately 12 % of all salivary carcinomas. The peak incidence occurs in the 6th to 7th decade of life (about 1-2 decades later than pleomorphic adenoma). While most are now recognized as salivary duct carcinomas, other morphological subtypes, including myoepithelial carcinoma and epithelial-myoepithelial carcinoma, are also seen (5). Microscopically, CXPA presents with varying degrees of invasion, including intracapsular, minor extracapsular (<5 mm beyond the capsule), and wide extracapsular (>5 mm beyond the capsule) invasion (6). Metastatic salivary gland tumors are rare clinical findings, with only 20 % of patients with parotid gland malignancy developing metastatic disease. Common sites for distant metastasis include the lungs, bones, liver, and central nervous system, although metastasis to the breast, ileum, spleen, and iliac crest has also been described (7). Tumors with wide extracapsular invasion (beyond 5 mm of the capsule) have a high risk of recurrence and distant metastasis, as seen in our case, where symptomatic vertebral metastatic lesions developed at the D11 and L3 vertebrae.

The transformation of PA to CXPA has been widely recognized, with previous reports detailing similar patterns of progression. In our case, PA occurred in a male patient at 38 years of age and transformed into CXPA at 50 years of age. This was a recurrent PA of

the soft palate that ultimately transformed into myoepithelial CXPA of the pharynx over a 12-year period. A case report of soft palate CXPA presenting as direct cavernous sinus invasion has been reported in the literature (4). However, in our patient, the CXPA of the pharynx invaded the carotid space, which, to our knowledge, has not been reported before. The carotid space is a paired area confined by the carotid sheath, a connective tissue boundary in the neck, which extends from the jugular foramen at the skull base to the aortic arch at the thoracic inlet. Lesions in the carotid space can arise from various structures within the space, including the carotid artery, cranial nerves IX, X, and XII, the ansa cervicalis, or the sympathetic plexus. These lesions include paragangliomas, carotid body tumors, glomus jugulare, glomus vagale, nerve sheath tumors, neurofibromas, schwannomas, lipomas, and carotid sheath meningiomas (8).

Surgical resection is the most common treatment for salivary gland tumors, and the extent of surgery is mainly determined by anatomical and clinical factors. However, for some lesions, the correct diagnosis should guide therapeutic decisions. In the case of PA, the risk of recurrence is about 2–3 %, with the highest risk seen in the myxoid subtype and in the presence of an incomplete tumor capsule, pseudopodia, and satellite nodules. Thus, more extended surgical techniques are preferred. Another factor necessitating extended surgery is recurrence (9). Complete resection with an intact capsule results in a lower recurrence rate. A higher number of recurrences significantly increases the risk of subsequent recurrence. PA is not associated with age or gender, and unlike Warthin tumor, it is not associated with tobacco use (10).

For patients with recurrent PA, radiotherapy is associated with a significant reduction in the risk of recurrence compared to surgery alone, regardless of the completeness of resection. Although concerns about radiation-associated toxicities exist, they are generally limited and do not outweigh the potential benefits of radiotherapy for recurrence. In this case, the patient did not receive adjuvant radiotherapy when he was operated on for recurrent PA, leading to multiple recurrences. This ultimately resulted in transformation into CXPA, with the disease becoming unresectable and distant metastasis developing. The patient was subsequently offered SBRT to symptomatic bony metastatic sites and palliative chemotherapy. However, the patient was lost to follow-up after 6 months, a key limitation of this case report. This early loss limits the ability to assess the long-term effectiveness of these therapies and their impact on the patient's quality of life.

The available literature on the role of chemotherapy in this context is limited. However, some chemotherapeutic agents—such as platinum compounds (cisplatin, carboplatin), taxanes (paclitaxel), and alkylating agents (cyclophosphamide, fluorouracil)—have been used in metastatic settings with varying results. Adjuvant concurrent chemotherapy is generally recommended in cases with high-risk features, including unresectability, positive surgical margins, or lymph node involvement.

There is growing interest in exploring novel treatment strategies. Several investigational studies have focused on molecular markers, particularly BRCA1 and BRCA2 mutations. In addition, researchers have investigated a range of other genetic alterations, such as HMGA2**,** CASP8, MLH1**,** RARB**,** KLK3**,** AI69125**,** EWSR1 rearrangement**,** and EGFR amplification. Despite these efforts, no specific genetic alteration has yet been definitively linked to CXPA, and targeted therapies for this condition remain under investigation.

Conclusion

Proper diagnosis of salivary gland pathology is crucial in making the right therapeutic decisions, determining the patient's prognosis, and increasing survival rates through a multi-modality approach. Tumors with wide extracapsular invasion have a high risk of recurrence and distant metastasis. Significant challenges are seen from its clinical presentation to diagnosis and treatment. This case highlights the aggressive potential of recurrent pleomorphic adenoma and underscores the importance of adjuvant radiotherapy and vigilant follow-up. The rare occurrence of carotid space invasion and vertebral metastases in this patient adds a unique aspect to the clinical picture.

Author contribution

Conceptualization, SA, KG and KM; writing original draft preparation, SA, KG, KM and MD; writing, review and editing, SA, MD and SR; supervision, KM and SA; Figures: KG and MD All authors have read the final manuscript

Funding

There is no funding.

Conflicts of interest

There are no conflicts of interest.

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