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RESEARCH ARTICLE

AGGRESSIVE ADENOID CYSTIC CARCINOMA OF THE NASAL CAVITY AND SOFT PALATE WITH EXTENSIVE SKULL BASE AND INTRACRANIAL EXTENSION: A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Adenoid cystic carcinoma (ACC) is a rare salivary gland malignancy characterized by slow growth, perineural invasion, and a propensity for local recurrence. We report an unusual case of an aggressive, unresectable ACC of the nasal cavity and soft palate with extensive skull base and intracranial extension in a 74-year-old female presenting with progressive dysphagia and right ear symptoms. Imaging revealed a large locally advanced lesion involving the soft palate, nasal cavity, maxillary sinus, skull base, orbit, infratemporal fossa, and temporal fossa. Histopathology confirmed ACC with perineural invasion. Owing to extensive disease, surgery was not feasible. The patient received multiple lines of systemic therapy, including CAP chemotherapy, vinorelbine-cisplatin, albumin-bound paclitaxel, methotrexate-gefitinib, and cisplatin-capecitabine, followed by palliative radiotherapy (30 Gy in 10 fractions). Despite multimodality treatment, the disease demonstrated relentless progression with poor response to therapy. The patient ultimately required hospice care and succumbed to the disease. This case highlights the aggressive biological behavior and therapeutic challenges associated with advanced ACC.

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INTRODUCTION

Adenoid cystic carcinoma (ACC) is a rare malignant epithelial tumor that most commonly arises from the major and minor salivary glands. It accounts for approximately 1% of all head and neck malignancies and nearly 10% of salivary gland cancers, with an estimated annual incidence of 3–4.5 cases per million population worldwide.(1,2) The tumor most frequently affects individuals in the fifth to seventh decades of life and demonstrates a slight female predominance(2,3). Although ACC is often characterized by a relatively slow growth pattern, it exhibits a distinctive tendency for perineural invasion, local tissue infiltration, delayed recurrence, and distant metastasis (4). Minor salivary glands of the palate constitute one of the most common sites of origin in the oral cavity(1,4). Owing to its neurotropic behavior, the tumor may extend along cranial nerves and reach the skull base, resulting in involvement of critical neurovascular structures and occasionally intracranial extension (5). Complete surgical resection followed by adjuvant radiotherapy remains the standard treatment for localized disease. However, management becomes particularly challenging in patients with unresectable tumors involving the skull base or intracranial compartment, where systemic therapy and palliative radiotherapy often provide limited benefit (5,6).

We report a rare case of advanced ACC of the nasal cavity and soft palate with extensive skull base destruction and intracranial extension that demonstrated resistance to multiple lines of chemotherapy and palliative radiotherapy, ultimately resulting in a fatal outcome.

CASE PRESENTATION

A 74-year-old female presented to our Radiation Oncology outpatient department with complaints of progressive dysphagia predominantly to solid foods for three months, which was insidious in onset and gradually progressed to involve semisolid foods associated with progressive anosmia and nasal obstruction. She also reported intermittent vibratory sensations in the right ear for the preceding two months. There was no history of tobacco or alcohol consumption. Her medical history was significant for hypertension for three years, controlled on antihypertensive medications, and hypothyroidism for which she had been receiving levothyroxine replacement therapy for three months. There was no significant family history of malignancy. Clinical examination revealed an ulceroproliferative lesion involving

the soft palate measuring approximately 4 × 5 cm with erythematous and irregular margins. No clinically palpable cervical lymphadenopathy was identified. Histopathological examination of the biopsy specimen obtained from the nasal cavity lesion was consistent with adenoid cystic carcinoma (ACC), demonstrating characteristic features along with evidence of perineural invasion. Contrast-enhanced computed tomography (CECT) at presentation revealed a large heterogeneously enhancing soft tissue mass measuring approximately 5.1 × 4.4 × 4.6 cm arising from the right soft palate. The lesion extended into the nasal cavity and maxillary sinus with destruction of the right hard palate, maxillary alveolar arch, and the posterolateral and posteromedial walls of the maxillary sinus. Lateral extension involved the medial surface of the right mandibular ramus, while superiorly the tumor infiltrated the skull base with erosion of the body and greater wing of the sphenoid bone and extension into the sphenoid and posterior ethmoid sinuses. Intracranial extension into the right temporal fossa and posterior inferior extraconal compartment of the right orbit was noted. The lesion also infiltrated the pterygoid musculature and temporalis muscle with erosion of the pterygoid plates. Medially, it extended into the right lateral nasopharyngeal wall, causing partial obliteration of the right choana and nasopharyngeal airway, with involvement of the tensor veli palatini and levator veli palatini muscles.

The patient reported receiving Ayurvedic treatment for approximately two months prior to seeking oncological consultation. Following multidisciplinary evaluation, a repeat CECT was obtained for treatment planning. This demonstrated a large ill-defined heterogeneously enhancing lesion measuring approximately 4.6 × 4.0 × 4.5 cm involving the right soft palate and infratemporal fossa. The lesion infiltrated the retromaxillary fat pad and caused osteolytic destruction of the posterolateral and medial walls of the right maxillary sinus with diffuse circumferential thickening of the maxillary antrum. Additional involvement included erosion of the medial surface of the right mandibular ramus, infiltration of the right lateral pharyngeal wall, extension into the oral cavity with destruction of the hard palate, invasion of the right nasal cavity with erosion of the inferior turbinate and bony nasal septum, and destruction of the greater wing of the sphenoid bone with extension into the sphenoid sinus and dural infiltration along the right temporal lobe. These findings have been supported by Figure 1.

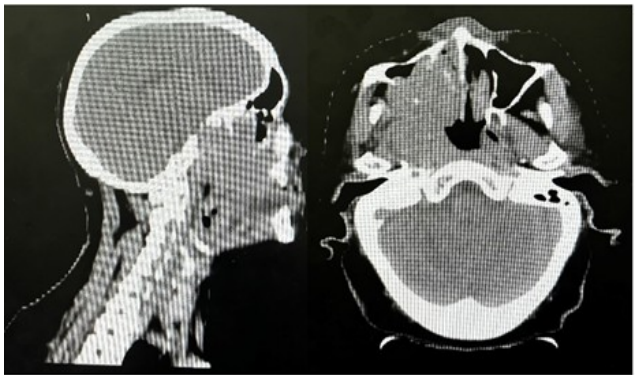


Figure 1. CECT Face and Neck demonstrating Radiological findings

The case was discussed in a multidisciplinary tumor board. Surgical evaluation deemed the tumor unresectable because of extensive local invasion, skull base involvement, and intracranial extension. Following cardiology clearance, the patient was initiated on systemic chemotherapy with the CAP regimen consisting of cyclophosphamide (1 g intravenously on Day 1), doxorubicin (60 mg intravenously on Day 1), and cisplatin (80 mg intravenously on Day 2), administered every three weeks with pegfilgrastim support. A total of six cycles were delivered. Post-chemotherapy radiological assessment demonstrated a modest response with mild reduction in tumor burden. The lesion measured approximately 4.5 × 3.1 × 5.1 cm. However, persistent disease was evident with continued infiltration of the right pterygomaxillary fissure and superior orbital fissure, osteolytic destruction involving the sphenoid body and greater wing, floor of the orbit, and petrous temporal bone. Tumor extension into the sphenoid sinus, middle cranial fossa, foramen ovale, foramen lacerum, and carotid canal was noted. The lesion was abutting the right cavernous sinus and anteromedial aspect of the temporal lobe and extended posteriorly into the nasopharynx involving the torus tubarius and Eustachian tube. The remaining radiological findings were largely unchanged. These findings have been shown in Figure 2.

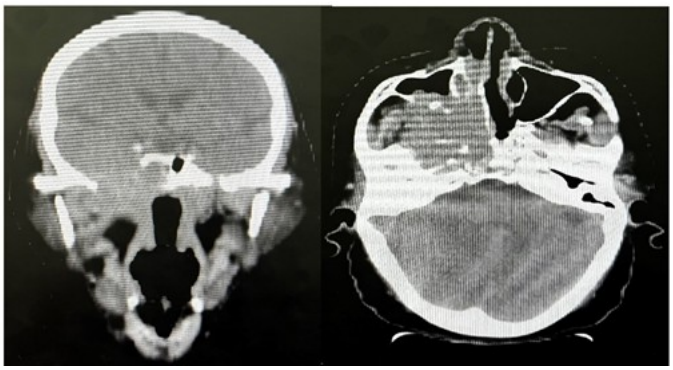


Figure 2. Post chemotherapy radiological status of disease

Given the suboptimal response, 2 cycles of second-line systemic therapy was initiated with vinorelbine (25 mg/m² intravenously on Days 1 and 8) combined with cisplatin (80 mg intravenously on Day 1). Subsequently, the patient received three cycles of albumin-bound paclitaxel administered weekly at a dose of 100 mg. Response assessment with positron emission tomography-computed tomography (PET-CT) unfortunately demonstrated disease progression, with the lesion increasing in size to approximately 5.3 × 4.6 × 7.0 cm while maintaining previously documented patterns of local invasion. In view of progressive unresectable disease and persistent symptom burden, the patient was treated with palliative external beam radiotherapy using an Elekta Versa HD linear accelerator. A dose of 30 Gy in 10 fractions (3 Gy per fraction) was delivered over two weeks. Despite completion of radiotherapy, significant residual disease measuring approximately 4 cm remained visible in the soft palate on clinical examination. At one-month follow-up, the patient reported worsening headache and nasal pain. Considering the substantial residual disease despite multiple lines of chemotherapy and palliative radiotherapy, further palliative systemic therapy was initiated with weekly methotrexate (50 mg intravenously) in combination with oral gefitinib (250 mg once daily). However, after four cycles, clinical and radiological progression was observed, accompanied by painful swelling over the root of the nose. Subsequently, the patient was switched to a combination regimen consisting of cisplatin (80 mg intravenously on Day 1)

and oral capecitabine (2000 mg daily in divided doses from Days 1–21). Four cycles were administered; however, no meaningful clinical or radiological response was achieved. Owing to continued disease progression and progressive decline in performance status, further active oncological treatment was deemed inappropriate. The patient was referred for hospice and best supportive care. Unfortunately, she succumbed to her disease approximately one month later, reflecting the highly aggressive and treatment-refractory nature of her malignancy.

DISCUSSION

ACC is generally regarded as an indolent malignancy; however, its biological behavior is frequently deceptive. Despite slow initial growth, the tumor demonstrates relentless local infiltration and a marked propensity for perineural spread, leading some authors to describe it as a "wolf in sheep's clothing".(1) The present case exemplifies this paradoxical behavior, as the patient presented with extensive locoregional disease involving the nasal cavity, maxillary sinus, infratemporal fossa, orbit, skull base, and intracranial compartment. Palatal ACCs commonly present with nonspecific symptoms such as pain, dysphagia, nasal obstruction, or a slowly enlarging mass, often resulting in delayed diagnosis.(2,3) In our patient, symptoms were initially limited to progressive dysphagia and ear-related sensations despite the presence of extensive underlying disease. Furthermore, the patient had received alternative therapy before oncological consultation, which may have contributed to further disease progression before definitive treatment could be initiated. Perineural invasion is one of the most characteristic pathological features of ACC and has been reported in approximately 30–60% of cases.(4) This phenomenon is strongly associated with local recurrence, skull base involvement, and poor prognosis. Histopathological evidence of perineural invasion in our patient correlated with the extensive spread to the sphenoid bone, skull base foramina, temporal fossa, and intracranial structures. Such patterns of spread highlight the unique neurotropic nature of ACC and its ability to traverse anatomical barriers that typically restrict local tumorextension (4,5). Surgery with negative margins remains the cornerstone of treatment and offers the best chance of long-term disease control (6). Unfortunately, the extensive skull base destruction, orbital involvement, and intracranial extension in our patient rendered the disease surgically unresectable. Similar challenges have been described in reports of skull base ACC, where complete excision is often impossible because of the proximity of critical neurovascular structures (5). The role of chemotherapy in ACC remains poorly defined. Several regimens, including cyclophosphamide, doxorubicin, cisplatin, vinorelbine, taxanes, and targeted agents, have been investigated, but objective response rates are generally modest and durable responses are uncommon.(6,7) In the present case, multiple lines of systemic therapy produced only transient disease stabilization followed by progression, underscoring the relative chemoresistance of advanced ACC. Radiotherapy plays an established role as an adjuvant modality and is frequently employed for unresectable disease or palliation.(6) Nevertheless, advanced tumors with extensive skull base invasion often remain difficult to control. Despite receiving palliative radiotherapy, our patient continued to demonstrate persistent residual disease and subsequent progression. This finding is consistent with previous reports showing suboptimal outcomes in patients presenting with advanced locoregional extension (5). Another remarkable aspect of the present case was the aggressive clinical course. Although ACC is classically associated with prolonged survival and late recurrence, certain tumors display highly aggressive behavior with rapid progression and treatment failure (1,7). The extensive skull base involvement, intracranial extension, and resistance to multiple systemic therapies likely contributed to the poor outcome observed in our patient. The case therefore emphasizes the biological heterogeneity of ACC and the need for improved systemic treatment strategies, including molecularly targeted therapies currently under investigation (7,8).

CONCLUSION

This case highlights an uncommon presentation of soft palate ACC with extensive skull base destruction and intracranial extension. Despite multimodality treatment including multiple chemotherapy regimens and palliative radiotherapy, the disease exhibited relentless progression and ultimately proved fatal. Early diagnosis and prompt multidisciplinary management remain critical, while advances in targeted therapies may offer future therapeutic opportunities for patients with unresectable and treatment-resistant ACC.

Ethics Statement: Written informed consent for publication was obtained from the patient. Ethical approval was not required according to institutional policy because no experimental intervention was performed. The study was conducted in accordance with the principles of the Declaration of Helsinki.

Author Contributions

Shivani Malik: Manuscript writing, Conceptualization, criticalrevision, Literature review, Supervision and final approval

The authorhave reviewed and approved the final manuscript.

Conflicts of Interest: The authors declare no conflict of interest.

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