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Cervical Spine Metastasis from Recurrent Parotid Adenoid Cystic Carcinoma (ACC) With Long Term Neurological Recovery

Suresh S Pillai*

Department of Spine Surgery, Sree Gokulam Medical College Hospital, India

***Corresponding author:** Dr. Suresh S Pillai, Department of Spine Surgery, Sree Gokulam Medical College Hospital, Venjaramood, Trivandrum, India

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Abstract

Background: Adenoid Cystic Carcinoma (ACC) of the parotid gland is a rare malignant salivary tumor with distinctive biological behavior including perineural invasion, indolent progression, and delayed distant metastasis. Skeletal metastases, particularly to the cervical spine, are uncommon and may cause severe neurological deficits.

Case presentation: We report a case of 42-years-old woman with a history of left parotidectomy and adjuvant radiotherapy for ACC, who presented five years later with progressive quadriparesis due to metastasis at the C7 vertebra. She underwent anterior cervical decompression with C7 corpectomy, iliac crest strut grafting, and anterior cervical plating. Postoperatively, she achieved excellent neurological recovery and independent ambulation. Two years later, she developed local recurrence at C7 and again underwent anterior decompression and stabilization through the contra lateral approach, with full neurological recovery. Another recurrence after two years manifested with widespread systemic metastases, including lung, liver, brain, and vertebral lesions, leading to her demise within four months.

Conclusion: Spinal metastasis from ACC of the parotid gland is exceedingly rare. This case is notable for its repeated cervical spine recurrence, delayed distant spread, and remarkable neurological recovery after two major spinal decompressions. The case highlights the importance of long-term vigilance, multidisciplinary timely management, and a thorough understanding of ACC's unique perineural and hematogenous spread.

Keywords: Adenoid Cystic Carcinoma; Parotid Gland; Cervical Spine Metastasis; Corpectomy; Quadriparesis; Recurrence; Perineural Spread

Introduction

Adenoid Cystic Carcinoma (ACC) is an uncommon malignant neoplasm arising from the secretory glands, accounting for approximately 1% of head and neck malignancies and 10% to 15% of all salivary gland tumors [1,2]. ACC exhibits a distinctive biological course, characterized by slow but relentless progression, perineural invasion, and late distant metastasis-often many years after initial treatment [3,4].While the lungs and bones are the most common sites of metastasis, spinal involvement, especially of the cervical spine, is rare but clinically significant due to potential for spinal cord compression and resultant neurological deficits [5,6].We report an unusual

case of recurrent C7 vertebral metastasis from parotid ACC, managed successfully twice with anterior cervical corpectomy and reconstruction, followed by eventual widespread dissemination. The patient's long-term survival and neurological recovery underscore the unique behavior and management challenges of this tumor.

Adenoid Cystic Carcinoma (ACC) is a rare malignant neoplasm arising from secretary glands, most commonly those of the salivary system. It constitutes approximately 1% of all head and neck malignancies, yet represents the most common tumor of the minor salivary glands and the second most common tumor of the major salivary glands, accounting overall for 10% of all salivary gland tumours [7]. ACC is typically slow-growing compared with other carcinomas and is well known for its propensity for perineural invasion and hematogenous spread, most frequently affecting elderly individuals [8]. Due to its rarity data on predisposing risk factors and optimal management strategies for advanced disease remain limited. ACC comprises only 2% to 3% of all parotid gland tumors, underscoring its uncommon occurrence at this site [9].

Case Report

A 42-years-old woman presented in 2011 with progressive weakness of all four limbs and gait difficulty over several weeks. She was wheelchair-bound at admission. Neurological examination revealed spastic quadriparesis (MRC Grade 3/5 upper limbs, 2/5 lower limbs), hyperreflexia, and bilateral Babinski positivity. Her history was significant for left parotidectomy for adenoid cystic carcinoma performed five years earlier, followed by adjuvant radiotherapy. Surveillance imaging had shown no local recurrence. Her mother had a history of carcinoma breast.

Imaging Findings

MRI of the cervical spine revealed a destructive lesion at the C7 vertebral body with collapse and cord compression (Figure 1). The lesion was hypointense on T1-weighted and hyperintense on T2-weighted images with paravertebral soft-tissue extension (Figure 2). A smaller lesion was also noted at the anterosuperior corner of S1, suggestive of metastasis.

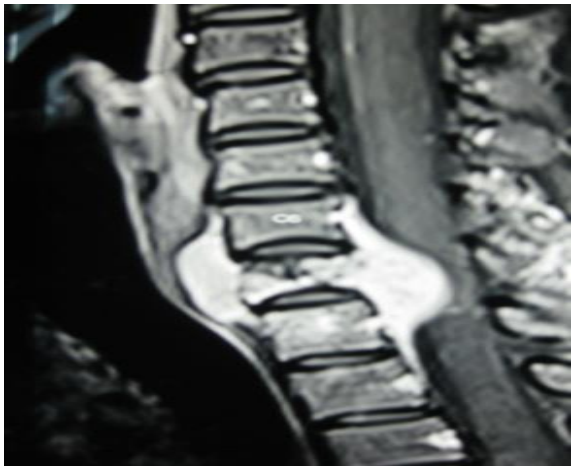

Figure 1: T2 weighted MRI scan of the cervical spine showing collapsed C7 vertebra with soft tissue component compressing the spinal cord.

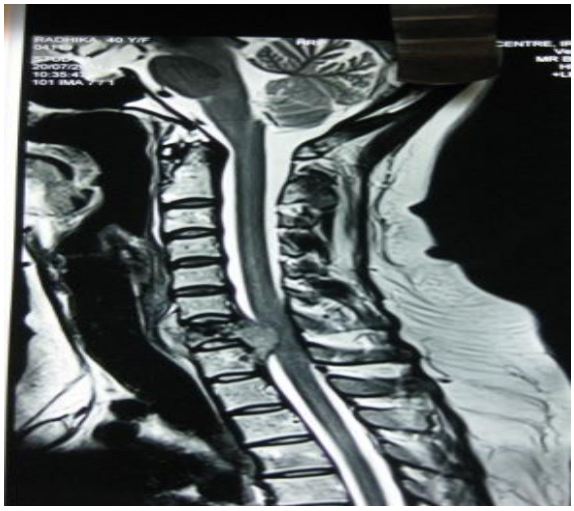

Figure 2: T1 weighted MRI scan of the cervical spine showing collapsed C7 vertebra with soft tissue component compressing the spinal cord.

Surgical Management (First Surgery, 2011)

The patient underwent C7 corpectomy, iliac crest strut grafting, and anterior cervical plating through a right-sided anterior cervical approach (Figure 3). Intraoperatively, a friable vascular lesion involving the C7 vertebral body was

excised. Reconstruction was performed using a tricortical iliac crest graft and cervical locking plate.

Histopathology confirmed metastatic adenoid cystic carcinoma, with a cribriform pattern and perineural invasion consistent with the parotid primary.

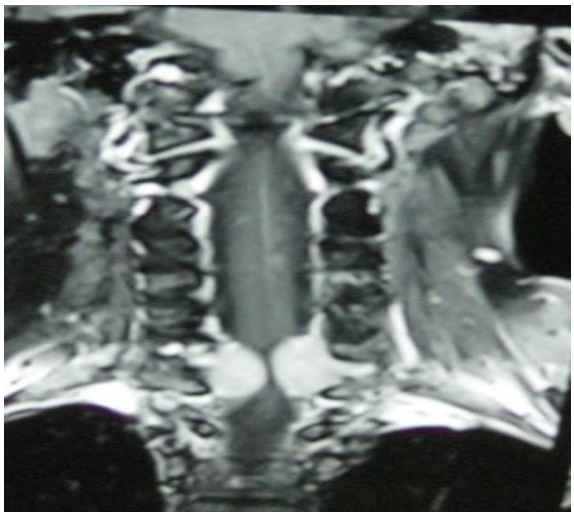

Figure 3: Coronal section of T2 weighted MRI scan of the cervical spine showing collapsed C7 vertebra with soft tissue component compressing the spinal cord.

Postoperative Course

Neurological improvement was dramatic-motor power improved to 4+/5, and she regained independent ambulation within three months. She was referred for medical oncology evaluation, and systemic therapy was continued.

Recurrence and Second Surgery (2013)

Two years later, she again developed progressive quadriplegia. MRI demonstrated recurrent tumor at the C7 level compressing the spinal cord. She underwent revision anterior cervical decompression and reconstruction via a left-sided approach, excising the recurrent tumor and replacing the structural graft. Postoperatively, she achieved near-complete neurological recovery, regaining independent walking ability.

Final Recurrence and Disease Progression (2015)

In 2015, the patient presented with quadriplegia and respiratory difficulty. MRI and CT revealed diffuse recurrence at the previous site, with metastases involving the lungs, brain, liver, and multiple vertebrae (Figure 4). Despite supportive care and palliative oncology intervention, she succumbed to disseminated disease within four months.

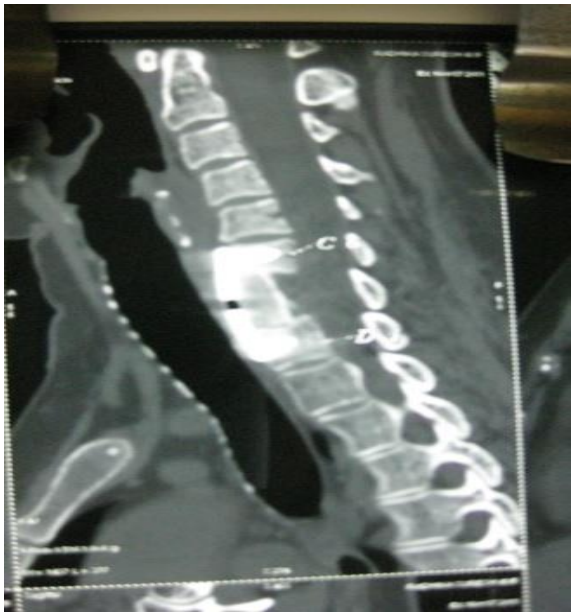

Figure 4: CT scan showing tricortical iliac crest grafting and fixation after the tumour removal.

Discussion

Epidemiology and Patterns of Spread

ACC of the parotid gland is well known for its slow but relentless progression, and its hallmark feature-perineural invasion-permits spread along cranial nerves and deep fascial planes [10]. Distant metastases may occur years or decades after initial treatment, even in the absence of local recurrence [11].

The most common metastatic sites are the lungs (≈40% to 60%), followed by bones (≈15%), liver, and brain [9]. However, spinal metastases from ACC are extremely rare, with only a few cases involving the cervical region reported in the literature [12-17].

A recent systemic review of 141 multicentric, multicountry clinical studies that included more than 25,800 patients found that adenoid cystic carcinoma was the second most common tumor after pleomorphic adenoma and the most common malignancy in the salivary gland [18].

Despite the slow development of the SACC, it is considered an aggressive tumor that can easily invade the surrounding structures. Perineural Invasion (PNI) is a distinctive feature of SACC by which the tumor cells travel along the nerves causing distant metastasis, especially intracranial.

Pathophysiology of Spinal Involvement

Bone metastasis in ACC is typically hematogenous, but perineural spread via the sympathetic chain and paravertebral plexus has been proposed, explaining the propensity for cervical involvement [19].

In this case, the C7 vertebra was repeatedly affected-possibly due to venous dissemination through Batson's plexus or direct perineural extension from the original parotid region.

Moreover, SACC can spread via the conventional perivascular route, most commonly to the lungs, followed by bone, liver, skin, and breasts, and rarely intracranially. However, intracranial metastasis is likely to occur in other ways, such as PNI, or via direct invasion of the base of the skull by an adjacent primary lesion [20].

Clinical Features

Neurological manifestations depend on the level of cord compression and degree of canal compromise. The index case presented with progressive spastic quadriparesis, a classical presentation of compressive myelopathy due to metastatic destruction.

SACC of the parotid has been reportedly associated with an odontogenic-like pain referring to the maxillary sinus and sialolithiasis [21,22].

Genetics

Most of the reported findings have confirmed that genetic factors are potential initiators of SACC. However, there have not been firm findings on which specific gene type or mutation causes SACC. Dai et al. [23], have investigated the possible link between beta-calcitonin gene-related peptide β CGRP of rs2839222 T/T genotype and SACC occurrence, and the study findings have concluded that this gene could be a high-risk factor for SACC because the serum levels of CGRP and β CGRP peptides were significantly high in SACC patients. It has also been reported that the gene mutation **KDM6A** could play a role in the SACC disease process [24]. The study has also reported a new gene mutation **KRAS** in two cases of SACC of Bartholin's salivary glands in the lip. Xie et al. [25], have detected evidence of upregulation of the NOTCH signaling cascade, a well-known signaling pathway that has been proven to contribute to the development of some human cancers, and its genetic receptor **NOTCH1**, as well as its downstream gene **HES1**, in the carcinogenesis, invasion, and metastasis of SACC, potentially by promoting the Epithelial-Mesenchymal Transition (EMT) related genes.

Biomarkers

Several studies have investigated the involvement of specific peptides and proteins in the SACC etiology. In a quantitative spectrometry-based study to analyse the protein expression profile in SACC and peritumoral tissue samples, more than 40,000 specific peptides and 4,454 Differentially Expressed Proteins (DEPs) were identified [26]. Of which, HAPLN1 was the most upregulated protein and BPIFB1 was the most downregulated. The study emphasized the importance of investigating the effects of these biomarkers on the SACC occurrence and progression. Kerr et al. [27].

Management

The management of spinal metastasis from ACC requires a multidisciplinary approach involving spine surgery, oncology, and radiotherapy. The primary goals are neurological preservation, pain relief, and spinal stability [28]. Surgical decompression is warranted when there is neurological deficit or spinal instability [29]. In our case, two anterior corpectomy and reconstruction procedures achieved complete neurological recovery on both occasions, demonstrating the effectiveness of timely surgical intervention even in recurrent disease.

Prognosis

Although systemic metastases from ACC are generally associated with poor prognosis, the disease course is notably indolent, and patients may survive for several years after metastasis [30]. In the present case, the patient survived nearly nine years after initial metastasis, with excellent functional recovery after two decompressive surgeries, highlighting the benefit of aggressive management and close follow-up [31,32].

Conclusion

Spinal metastasis from adenoid cystic carcinoma of the parotid gland is exceedingly rare and often presents long after primary treatment. This case is remarkable for its recurrent cervical metastasis, bilateral surgical approaches, and repeated full neurological recovery despite eventual widespread dissemination. Long-term surveillance is essential for ACC patients, given its unpredictable metastatic potential and delayed recurrences.

Future Trends in Diagnosis and Treatment

Many studies have investigated the viability of using specific biological markers, proteins, and signalling pathways that have been shown to promote or suppress SACC as a potential target therapy or as an early hallmark for the diagnosis of SACC.

The future trends in the diagnosis and management of SACC depend on the discoveries of certain elements attributable to the disease oncogenesis. However, the rarity of the disease hampers the striving for further research and clinical trials to explore new approaches and novel therapies.

Take-Home Message

- Adenoid cystic carcinoma may metastasize to the spine even years after primary surgery.
- Cervical involvement is rare but can lead to severe neurological deficits.
- Early surgical decompression can yield excellent neurological recovery, even in recurrent cases.
- Long-term follow-up is mandatory due to the tumor's potential for late and repeated metastasis.

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