

## CASE REPORT

## Advanced Adenoid Cystic Carcinoma with Widespread Visceral and Skeletal Metastases

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**ABSTRACT**

**Context:** Adenoid cystic carcinoma (ACC) is a rare malignancy of secretory glands, characterized by an indolent yet relentless course with high rates of local recurrence and distant metastases.

**Case:** We present a case of a 40-year-old male diagnosed in 2019 with nasal ACC, who developed widespread metastases to lungs, liver, bones, peritoneum, and lymph nodes despite multiple surgeries and targeted therapies. Progression continued on lenvatinib, lenalidomide, and axitinib. Patient received palliative radiotherapy for painful skeletal lesions.

**Conclusion:** This case highlights the aggressive nature of ACC in advanced stages and underscores the role of palliation when systemic therapies fail.

**KEYWORDS**

• Adenoid cystic carcinoma • Metastatic ACC • Targeted therapy • VEGF inhibitors • Palliative radiotherapy

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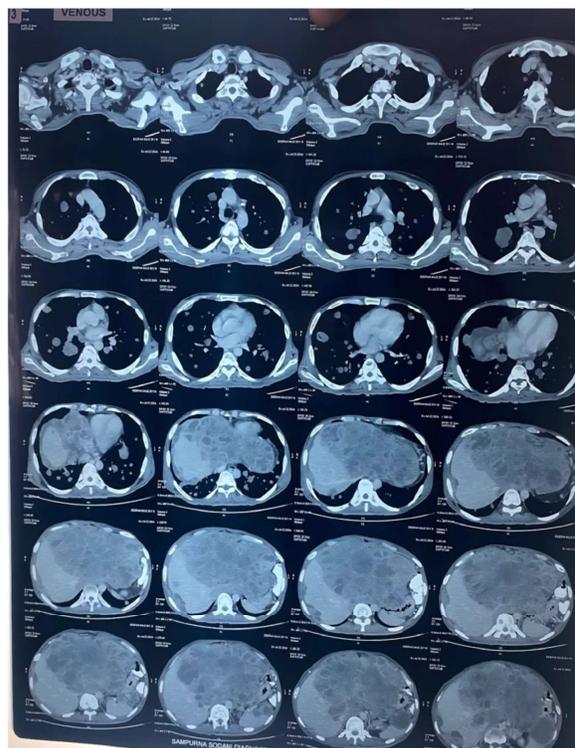
## INTRODUCTION

Adenoid cystic carcinoma (ACC) is an uncommon salivary gland tumor, accounting for less than 1% of head and neck malignancies. It typically arises in the minor salivary glands, including the nasal cavity and paranasal sinuses. Though its progression is often slow, ACC is infamous for perineural invasion, recurrence, and delayed metastasis.<sup>1</sup> Common sites of metastasis include the lungs, liver, bone, and, rarely, the peritoneum. Managing metastatic ACC is a challenge, with limited effective systemic options and an emphasis on palliation.<sup>2</sup>

## CASE REPORT

A 40-year-old non-smoking male presented in November 2019 with a left nasal mass. He underwent lateral rhinotomy, and histopathological examination revealed adenoid cystic carcinoma. There was perineural invasion but no evidence of distant metastasis. He did not receive any adjuvant treatment and between 2019 and 2023 had recurrences three times for which he underwent three additional lateral rhinotomies for locoregional recurrence. In early 2024, he reported systemic symptoms including loss of appetite, significant weight loss, generalized weakness, and right-sided facial swelling. On clinical examination, pallor was noted but no icterus, clubbing, or palpable lymphadenopathy. Respiratory examination revealed mild basal crepitation on both sides. A firm periumbilical mass was present on abdominal wall and tenderness was noted over the lumbar spine. His CT Scan abdomen on 23 February 2024 revealed a massively enlarged hepatic lesion in segments II, III, IVA, and V, measuring 21.7 x 17.2 x 14.1 cm. The lesion showed near-complete obliteration of the left portal vein and encasement of the inferior vena cava. There were multiple omental nodules and perisplenic peritoneal deposits (*Figure 1*). A biopsy taken from the liver confirmed metastatic adenoid cystic carcinoma. Immunohistochemistry showed weak androgen receptor expression (1-2%). He was initiated on oral lenvatinib (24 mg) targeting angiogenic pathways, continued for 10 months. In June 2025, repeat CT chest and whole abdomen imaging showed new pulmonary parenchymal nodules and pleural based lesions the largest being 3.6 x 3.6 cm in the right lower lobe. There was also evidence

of skeletal metastasis, including rib erosion and lytic lesions in ribs 2 to 5. Hepatic lesions remained large and invasive, with increased size of subcapsular and peritoneal deposits. After disease progression on lenvatinib, he was switched to a combination of lenalidomide and axitinib. Nevertheless, a PET-CT scan on 25 July 2025 revealed florid, widespread FDG-avid disease. FDG uptake was observed in a recurrent nasal cavity lesion measuring 2.7 x 2.2 cm (SUVmax 3.0), multiple bilateral pulmonary nodules (SUVmax up to 7.1), mediastinal lymphadenopathy, and an exophytic hepatic mass compressing adjacent viscera (SUVmax 4.4). Discrete omental and peritoneal lesions infiltrated the abdominal and pelvic cavities, while subcutaneous FDG-avid deposits were noted in the anterior abdominal wall (4.3 x 4.1 cm; SUVmax 5.8) and large mass in liver (*Figure 2 & 3*). A biopsy from the periumbilical deposit reconfirmed metastatic ACC. Due to progressive bone pain, he received palliative radiotherapy (30 Gy in 10 fractions) to lumbar spine with 6MV photons on Linear accelerator. As per multidisciplinary tumor board decision, doxorubicin-based chemotherapy was commenced, given extensive disease and poor response to targeted agents. Presently patient is on chemotherapy without any response.



**Figure 1:** Ct Showing Massively Enlarged Hepatic Lesion



Figure 2: Petct Showing Diffuse Disseminated Mets

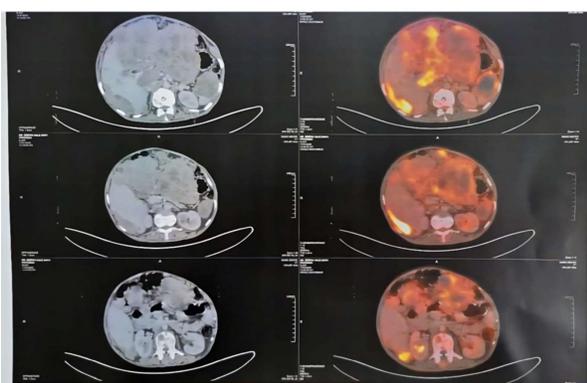


Figure 3: Petct Showing Fdg Uptake In Liver

## DISCUSSION

Adenoid cystic carcinoma of the nasal cavity is rare but known for aggressive local behavior and delayed but recurrent distant metastasis.<sup>3</sup> Despite complete surgical excision and clear margins, recurrence and distant spread are common. Lung metastases are the hallmark of disseminated disease and may remain indolent for years. However, involvement of the liver, peritoneum, bones, and subcutaneous tissue as in this case is very unusual and indicates rapid tumor progression.<sup>4</sup> The diagnostic challenge in this patient was not in identifying primary disease, but rather in controlling metastatic spread in organ systems rarely affected by ACC.<sup>5</sup> Prolonged survival in ACC often masks low-grade but widespread dissemination. The liver was significantly affected in our patient with multifocal, partially necrotic regions causing vascular compromise and visceral compression. Concurrent omental nodules, mesenteric masses, and subcutaneous deposits contributed to a complex clinical picture, mimicking carcinomatosis from

gastrointestinal sources. Targeted agents like lenvatinib (a VEGF receptor tyrosine kinase inhibitor) and axitinib (primarily used in renal cancers) have shown modest benefits in salivary gland tumors including ACC.<sup>6</sup> Lenvatinib-based studies show stabilization in some ACC subtypes, but responses are rarely durable. After a brief stabilization, this patient developed further metastases, highlighting the limitations of current systemic therapy.

Given the weak androgen receptor positivity in this case, hormonal manipulation was not considered promising. Cytotoxic chemotherapy, such as doxorubicin, although associated with limited survival benefit, remains a reasonable choice in rapidly progressing cases where targeted therapy has failed.<sup>7</sup>

Palliative radiotherapy addressed pain and improved quality of life. The patient's extensive radiographic disease burden including PET-avid foci in visceral, musculoskeletal, and dermal tissues attests to the aggressive biology of this otherwise indolent tumor.<sup>8</sup>

## CONCLUSION

This case describes a rare and unusually aggressive pattern of disseminated adenoid cystic carcinoma originating in the nasal cavity, with metastasis to the liver, lungs, peritoneum, bones, and subcutaneous tissue. Despite initial surgical control, the disease rapidly evolved with widespread FDG-avid metastasis and resistance to molecular-targeted treatment. This underscores the unpredictable nature of ACC, timely adjuvant treatment and the importance of lifelong surveillance, early detection of recurrence, and the need for novel, more effective therapeutic strategies. A multidisciplinary palliative approach is essential to optimize outcomes and manage symptoms in advanced disease stages.

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