Journal of
Medical Case Reports

CASE REPORT Open Access

Salivary gland adenoid cystic carcinoma presenting as a large metastatic hepatic mass: a case report

Matthew Walker[1], Alisa Dewald[1], Abdelrhman Refaey[2], Ivan Berezowski[3], Jacob Newman[5], Mamoun Younes[4] and Stephen Gray[2*]

Abstract

Background Salivary gland tumors are rare, representing 6–8% of head and neck tumors. Adenoid cystic carcinoma represents approximately 10% of salivary gland tumors. The preferential sites of metastases are the lung and bone, followed by the brain and liver. Liver metastasis as the initial clinical manifestation of parotid gland adenoid cystic carcinoma is very rare.

Case presentation We report the case of a 29-year-old African American male patient with a metastatic salivary gland tumor who presented initially with a large hepatic mass. He complained of right upper quadrant pain and early satiety. Abdominal computed tomography revealed hepatomegaly with a large mass centered in the left lobe measuring 14 cm. A computed tomography scan revealed innumerable bilateral pulmonary nodules measuring up to 8 mm, favoring metastases. Initial histopathological examination of the liver biopsy specimens revealed a well-differentiated carcinoma characterized by a distinctive myxoid stroma, consistent with metastatic adenoid cystic carcinoma. The patient underwent transcatheter arterial chemoembolization and was discharged with a hematology–oncology follow-up. A presumptive diagnosis of stage IV adenoid cystic carcinoma with lung and liver metastases was made. The patient was given cisplatin and vinorelbine.

Conclusion Metastatic parotid gland adenoid cystic carcinoma with initial clinical manifestation as a liver mass is very rare and was pathologically confirmed in this patient by its histological appearance. This case emphasizes the need for clinicians to consider salivary gland tumors in the differential diagnosis of unexplained hepatic lesions.

Keywords Salivary gland tumor, Liver metastasis, Lung metastasis, Adenoid cystic carcinoma, Basal cell adenocarcinoma, Cytopathology, Case report

*Correspondence:
Stephen Gray
stgray@mfa.gwu.edu
[1] The George Washington University School of Medicine and Health Sciences, Washington, DC, USA
[2] The George Washington Transplant Institute, Department of Surgery, The George Washington University Hospital, Washington, DC, USA
[3] Department of Medicine, The George Washington University School of Medicine and Health Sciences, Washington, DC, USA
[4] Department of Pathology, The George Washington University School of Medicine and Health Sciences, Washington, DC, USA
[5] Division of Gastroenterology, Department of Medicine, Allegheny Health Network, Pittsburgh, PA, USA

Introduction

Salivary gland tumors are rare, representing 6–8% of head and neck tumors [1]. Among these, adenoid cystic carcinoma (ACC) is a significant malignant subset, accounting for approximately 10% of all salivary gland neoplasms and about 1% of head and neck malignancies [2, 3]. ACC affects both major and minor salivary glands, with a slight predilection for the minor glands. The incidence of ACC is estimated to be 0.4–13.5 cases per 100,000 person-years, highlighting its rarity [4].

ACCs are characterized by slow growth, high rates of distant metastasis, and difficulty in resection due to multiple recurrences [3, 4]. The 5-year survival rate for ACC is relatively high at 89%, but this drops significantly to 40% at 15 years, underscoring the importance of long-term follow-up [5]. The preferential sites of metastases are the lungs (occurring in up to 70% of metastatic cases) and bone, followed by the brain and liver [6]. Most liver metastases are derived from non-parotid ACCs and are often related to local recurrence or metastases to other organs, particularly the lung [7]. Notably, isolated liver metastases from ACC are extremely rare, with only a handful of cases reported in literature. In contrast, basal cell adenocarcinoma (BCACs), another malignant subset of salivary gland tumors, accounts for less than 1% of salivary gland tumors and are generally considered low-grade carcinomas that are locally destructive and tend to recur [2]. Unlike ACC, BCACs only occasionally metastasize, and very rarely to a distant site [8].

We herein report the case of a patient with a metastatic salivary gland tumor who presented initially with a significant hepatic mass originally believed to be a primary tumor. This case highlights the importance of considering rare metastatic presentations of ACC in the differential diagnosis of liver masses, even in the absence of a known primary tumor.

Case report

A 29-year-old African American male individual presented to the emergency department (ED) for right upper quadrant abdominal pain and early satiety. Physical examination was unrevealing except for abdominal tenderness. His past medical history included 5 years of daily alcohol use and occasional marijuana use. A computed tomography (CT) scan revealed hepatomegaly with a large mass centered in the left lobe measuring 14 cm (Fig. 1). Laboratory values showed mildly elevated transaminases, with an alanine transaminase (ALT) of 51, aspartate aminotransferase (AST) of 74, and an alkaline phosphatase of 93 units/L (Table 1). The patient was then discharged with a planned outpatient hepatology follow-up.

A total of 1 month later, the patient presented again to the ED with the same symptoms; magnetic resonance imaging (MRI) revealed a 19 cm enhancing left hepatic mass. Laboratory tests demonstrated a mild elevation in transaminases, a negative hepatitis viral panel, and unremarkable tumor marker levels, including alpha-fetoprotein (AFP), carcinoembryonic antigen (CEA), and carbohydrate antigen 19-9 (CA19.9). After appropriate pain control, the patient was discharged again with plans for outpatient follow-up.

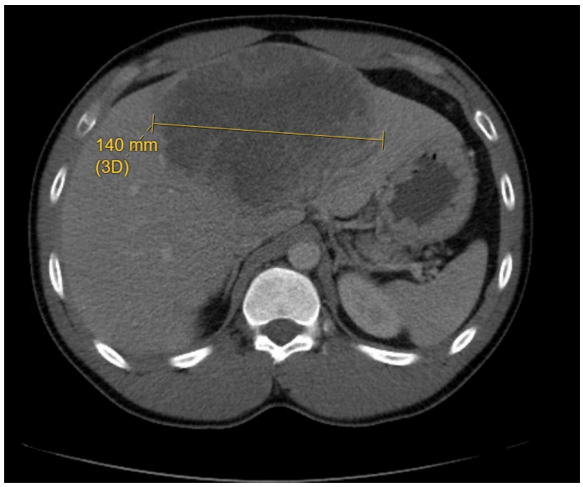

Fig. 1 Initial computed tomography scan demonstrates hepatomegaly with a large 14 cm mass in the left lobe

The patient was lost to follow-up owing to insurance concerns, until he presented to a different ED the following month for abdominal pain, vomiting, early satiety, and unintentional 10 lb weight loss. Abdominal CT angiography with contrast showed a 21 cm hepatic mass centered in the left hepatic lobe (Fig. 2), and given the concern for hepatocellular carcinoma, the patient was admitted for biopsy and staging imaging. Laboratory tests revealed alkaline phosphatase elevation (451 IU/L) and mildly elevated transaminases, with an AST level of 93 units/L and an ALT of 45 units/L. His tumor marker laboratory results were within normal ranges (CA 19.9 < 14.0; AFP: 1.97; and CEA: 5.28). A chest CT revealed innumerable bilateral pulmonary nodules measuring up to 8 mm, favoring metastases. Initial histopathological examination of the liver biopsy specimens revealed a well-differentiated carcinoma characterized by a distinctive myxoid stroma with tubular and cribriform structures, consistent with metastatic adenoid cystic carcinoma. The patient underwent transcatheter arterial chemoembolization (TACE) and was discharged with a hematology–oncology follow-up.

He was readmitted 2 weeks later for a planned repeat TACE procedure, as well as a repeat liver biopsy for further testing. Initial laboratory values on admission were an AST of 44 units/L, ALT of 24 units/L, and alkaline phosphatase of 282 units/L. During this admission, a computed tomography (CT) scan of the neck soft tissue with contrast revealed a complex bilobed mass in the right parotid gland measuring approximately 6.5 cm (Fig. 3). Its progression into the parapharyngeal space caused notable deformity of the oropharynx, raising further concerns about potential malignancy.

Table 1 Clinical course summary, with pertinent laboratory values and imaging results; several laboratory values were not available owing to presentation at an outside hospital

Presentation	Lab values	Mass findings	Treatment	Disposition
Initial ED visit	AST: 74 units/L ALT: 51 units/L Alk Phos: 93 units/L	14 cm liver mass	Symptom management	Discharged with outpatient follow-up
Return ED visit, 1 month after initial visit	Mildly elevated AST and ALT	19 cm liver mass	Symptom management	Discharged, then lost to follow-up
Third ED visit, 2 months after initial visit	AST: 93 units/L ALT: 45 units/L Alk Phos: 451 units/L	21 cm liver mass with bilateral 8 mm pulmonary nodules	Transcatheter arterial chemoembolization (TACE)	Admission
Readmission 10 weeks after the initial visit	AST: 44 units/L ALT: 24 units/L Alk Phos: 282 units/L	New 6.5 cm right parotid mass	TACE, cisplatin, and vinorelbine	Continued outpatient follow-up

AST, aspartate aminotransferase; ALT, alanine aminotransferase; Alk Phos, alkaline phosphatase; ED, emergency department; TACE, transarterial chemoembolization

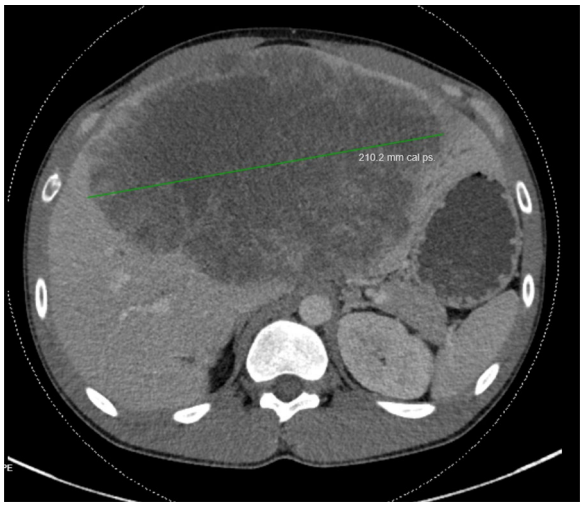

Fig. 2 Follow-up abdominal computed tomography angiography with contrast demonstrates hepatomegaly with a 21 cm hepatic mass centered in the left hepatic lobe

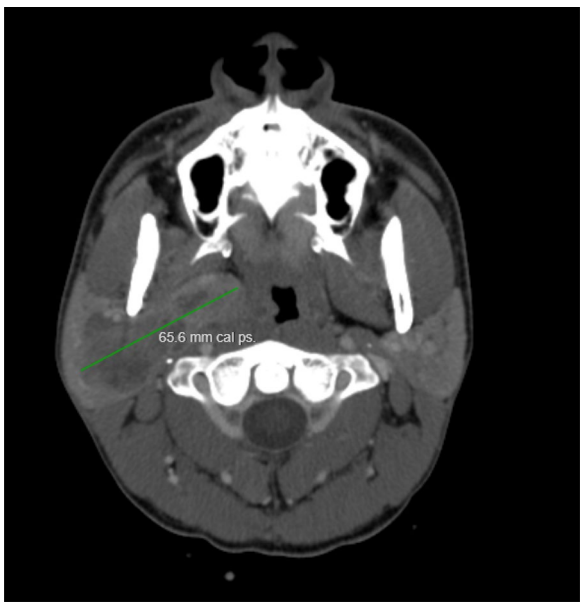

Fig. 3 A computed tomography scan of the neck soft tissue with contrast demonstrates a bilobed heterogeneously enhancing mass measuring 6.5 cm involving the superficial and deep lobe of the right parotid gland as well as the right parapharyngeal space

A biopsy was taken with immunohistochemistry findings suggestive of ACC (Fig. 4); however, BCAC could not be completely excluded without examination of a completely removed salivary gland tumor. Therefore, a presumptive diagnosis of stage IV adenoid cystic carcinoma with metastases to the liver and lung was made. Given the tremendous tumor burden and poor prognosis, the treatment goals were palliative, and the patient was started on a chemotherapeutic regimen with cisplatin and vinorelbine.

A total of 5 months later, the patient presented to the emergency department with neurological symptoms, including lightheadedness and left-sided weakness. A CT scan revealed new right frontal hyperdensities, consistent with brain metastasis. During this admission, he received one dose of radiotherapy with a plan to finish five doses. He was then discharged with a planned outpatient follow-up.

Discussion

This case presents a rare instance of ACC initially manifested as a single hepatic mass. ACC is known for its slow growth and tendency for delayed metastasis, typically to the lungs and bones before affecting the liver [6]. Our patient's presentation with a large liver mass as the primary finding is very rare.

The diagnostic journey in this case was complex, highlighting the challenges in identifying metastatic

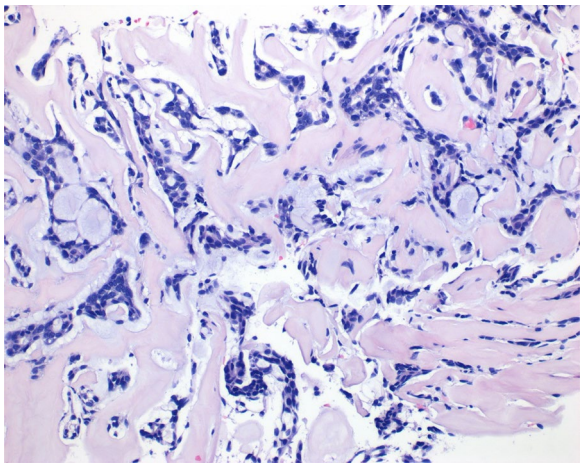

Fig. 4 Microscopic appearance of the salivary gland showing tubular and cribriform structures in a mucinous background (hematoxylin–eosin, 20×). Immunoperoxidase staining showing a dual-cell population with the majority of cells positive for cytokeratin 7 and a lesser population of cells positive for S100 and tumor protein 63

ACC without a known primary tumor. The differential diagnosis of a large hepatic mass without a known primary tumor includes hepatocellular carcinoma, metastatic disease from an unknown primary source, or benign hepatic tumors (for example, hepatic adenoma and focal nodular hyperplasia) [9]. The rapid growth of the hepatic mass and the discovery of pulmonary nodules raised suspicion for a metastatic process. The distinctive histopathological findings observed in the liver biopsy, showing a well-differentiated carcinoma with myxoid stroma, were crucial in guiding the diagnosis towards a salivary gland origin, which was confirmed later with imaging and biopsy of the parotid gland. Previous literature associates solid structures on pathologic examination with higher metastatic potential in ACC rather than tubular or cribriform structures [10]. Our patient, however, presented with distant metastases and primarily tubular and cribriform features on pathologic analysis (Fig. 3).

The unique behavior of adenoid cystic carcinoma (ACC) is closely linked to its genetic profile. Unlike many other cancers, ACC has a specific genetic pattern that helps explain its slow, but persistent spread. Researchers have discovered a particular gene fusion *(MYB-NFIB)* in about half of ACC cases, which appears to drive the tumor's ability to metastasize [11–14]. These genetic characteristics help explain why ACC can develop metastases years after the initial tumor is treated, and why it tends to spread to specific organs like the lungs and liver [15].

Liver metastasis in ACC is rare and typically occurs in conjunction with other organ involvement. Our case is notable, as isolated liver metastasis is uncommon, with only a few previous reports in literature [3, 5, 16–19]. The median survival time for patients with ACC liver metastases is approximately 14 months, with 1-year survival rates at around 55.8% [4].

The treatment modalities for these patients are poorly understood because of their rarity, high degree of resistance to chemotherapeutic agents, and recurrence following surgical resection [3]. In this case, given the extensive tumor burden, a palliative approach combining local therapy (TACE) with systemic chemotherapy was chosen. This aligns with current practices for advanced ACC, where treatment aims to control symptoms and potentially slow disease progression [4].

Our case contributes to the limited understanding of ACC's atypical presentations, emphasizing the importance of considering salivary gland tumors in the differential diagnosis of unexplained hepatic lesions. The treatment remains individualized, with a focus on palliative care to maximize patient comfort.

Conclusion

This case is an example of the atypical presentation of a metastatic salivary gland tumor as a single large hepatic mass. This emphasizes the need for clinicians to maintain a broad differential diagnosis when encountering hepatic masses, particularly in patients without a known primary malignancy. Adenoid cystic carcinoma presenting as a liver metastasis is exceedingly rare, highlighting the importance of thorough histopathological examination and consideration of atypical presentations of salivary gland tumors. We recommend comprehensive imaging studies, including head and neck regions, along with regular follow-up imaging in similar cases. This report contributes to the limited literature on ACC liver metastasis and addresses the necessity for individualized diagnostic and treatment approaches in managing rare salivary gland malignancies.

Acknowledgements
Not applicable.

Author contributions
MW and AD drafted the hospital course and contributed to the initial manuscript. AR drafted the conclusion and discussion sections, collaborated in editing and finalizing the manuscript, and handled the submission process. IB and JN reviewed the manuscript draft to ensure scientific accuracy. MY, as the pathologist, obtained the pathology images and authored the pathology section of the report. SG is the corresponding author and supervised the overall project.

Funding
No funding was received for this study.

Availability of data and materials
The datasets analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate
The need for ethical approval was waived. Written consent was obtained from the patient for participation in this case report.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare that they have no competing interests related to this study.

Received: 1 November 2024 Accepted: 6 February 2025
Published online: 19 March 2025

References

1. El-Naggar AK, Chan JKC, Grandis JR, *et al*. World Health Organization classification of tumours of head and neck. Lyon: IAARC; 2017.
2. Tian Z, Li L, Wang L, Hu Y, Li J. Salivary gland neoplasms in oral and maxillofacial regions: a 23-year retrospective study of 6982 cases in an eastern Chinese population. Int J Oral Maxillofac Surg. 2010;39(3):235–42. https://doi.org/10.1016/j.ijom.2009.10.016.
3. Li JY, Liu P, Lei YL, Mao ZQ, Hu F, Liu JB. Liver metastasis from adenoid cystic carcinoma of the submandibular gland: a case report. Int J Clin Exp Pathol. 2020;13(3):550–5.
4. Zheng Y, He Y, Wu F, Liu M, Wang L, Wu J. Possible local treatment for liver metastases of adenoid cystic carcinoma (ACC): single-centre experience and literature review. Transl Cancer Res. 2020;9(8):4573–82. https://doi.org/10.21037/tcr-20-1028.
5. Li XH, Zhang YT, Feng H. Liver metastasis as the initial clinical manifestation of sublingual gland adenoid cystic carcinoma: a case report. World J Clin Cases. 2021;9(19):5238–44. https://doi.org/10.12998/wjcc.v9.i19.5238.
6. Spiro RH. Distant metastasis in adenoid cystic carcinoma of salivary origin. Am J Surg. 1997;174(5):495–8. https://doi.org/10.1016/s0002-9610(97)00153-06.
7. Harish K, Mangala Gouri SR. Adenoid cystic carcinoma of the parotid metastasizing to liver: case report. BMC Cancer. 2004;4:41. https://doi.org/10.1186/1471-2407-4-41.Published2004Jul30.
8. Cuthbertson DW, Raol N, Hicks J, Green L, Parke R. Minor salivary gland basal cell adenocarcinoma: a systematic review and report of a new case. JAMA Otolaryngol Head Neck Surg. 2015;141(3):276–83. https://doi.org/10.1001/jamaoto.2014.3344.
9. Rodríguez de Lope C, Reig ME, Darnell A, Forner A. Approach of the patient with a liver mass. Frontline Gastroenterol. 2012;3(4):252–62. https://doi.org/10.1136/flgastro-2012-100146. (**Epub 2012 Aug 11**).
10. Sung M, Kim KH, Kim J, *et al*. Clinicopathologic predictors and impact of distant metastasis from adenoid cystic carcinoma of the head and neck. Arch Otolaryngol Head Neck Surg. 2003;129(11):1193–7. https://doi.org/10.1001/archotol.129.11.1193.
11. Humtsoe JO, Kim HS, Jones L, Cevallos J, Boileau P, Kuo F, Morris LGT, Ha P. Development and characterization of MYB-NFIB fusion expression in adenoid cystic carcinoma. Cancers (Basel). 2022;14(9):2263. https://doi.org/10.3390/cancers14092263.
12. Wysocki PT, Izumchenko E, Meir J, Ha PK, Sidransky D, Brait M. Adenoid cystic carcinoma: emerging role of translocations and gene fusions. Oncotarget. 2016;7:66239–54.
13. Mitani Y, Li J, Rao PH, Zhao YJ, Bell D, Lippman SM, Weber RS, Caulin C, El-Naggar AK. Comprehensive analysis of the *MYB-NFIB* gene fusion in salivary adenoid cystic carcinoma: incidence, variability, and clinicopathologic significance. Clin Cancer Res. 2010;16(19):4722–31. https://doi.org/10.1158/1078-0432.CCR-10-0463.
14. Ho AS, Ochoa A, Jayakumaran G, Zehir A, Valero Mayor C, Tepe J, Makarov V, Dalin MG, He J, Bailey M, Montesion M, Ross JS, Miller VA, Chan L, Ganly I, Dogan S, Katabi N, Tsipouras P, Ha P, Agrawal N, Solit DB, Futreal PA, El Naggar AK, Reis-Filho JS, Weigelt B, Ho AL, Schultz N, Chan TA, Morris LG. Genetic hallmarks of recurrent/metastatic adenoid cystic carcinoma. J Clin Invest. 2019;129(10):4276–89. https://doi.org/10.1172/JCI128227.
15. Xu LH, Zhao F, Yang WW, Chen CW, Du ZH, Fu M, Ge XY, Li SL. MYB promotes the growth and metastasis of salivary adenoid cystic carcinoma. Int J Oncol. 2019;54(5):1579–90. https://doi.org/10.3892/ijo.2019.4754. (**Epub 2019 Mar 18**).
16. Garg N, Tomar R, Goyal S, Singh UR. Isolated liver metastases in an adenoid cystic carcinoma of the submandibular gland on fine needle aspiration cytology: an unusual presentation. Cytopathology. 2014;25:137–8 (**Crossref. PubMed**).
17. Deshpande AH, Kelkar AA. Hepatic metastasis as an initial manifestation of salivary adenoid cystic carcinoma: cytologic diagnosis. Diagn Cytopathol. 2009;37:45–7 (**Crossref. PubMed**).
18. Spolverato G, Fite J, Bishop J, Argani P, Pawlik TM. Liver metastasis as the initial presentation of adenoid cystic carcinoma. Dig Dis Sci. 2014;59:2004–6 (**Crossref. PubMed**).
19. Zemni I, Tounsi N, Bouraoui I, Slimene M, Sahraoui G, Ayadi MA, Chargui R, Rahal K. A single liver metastasis from adenoid cystic carcinoma of the parotid gland: case report. J Investig Med High Impact Case Rep. 2019;7:2324709619879631. https://doi.org/10.1177/2324709619879631.

Publisher's Note