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International Journal of Surgery Case Reports

journal homepage: www.casereports.com

A rare case of lacrimal adenoid cystic carcinoma with large hepatic and multiple pulmonary metastases with successful surgical treatment



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ARTICLE INFO

Article history:

Received 20 January 2016

Accepted 22 January 2016

Available online 2 February 2016

Keywords:

Adenoid cystic
Lacrimal gland
Hepatic metastases
Lung metastases

ABSTRACT

INTRODUCTION: Adenoid cystic carcinoma (ACC) of the lacrimal gland is rare but with high recurrence rates and late metastases. They occasionally metastasise via haematogenous spread to lungs, brain and bone. Liver is a rare site of metastasis and is usually present with disseminated disease.

PRESENTATION OF CASE: A 42 year old lady, a known case of adenoid cystic carcinoma of the lacrimal gland who had been operated 4 years ago for the same followed by chemotherapy and radiotherapy. She was detected with large hepatic and bilateral multiple pulmonary metastases. A right hepatectomy was done. After six months when her general condition improved we did a staged pulmonary metastatectomy. Histology confirmed the diagnosis. She is presently doing well with no recurrence since 18 months.

DISCUSSION: Usually asymptomatic, the commonest symptom is pain. It is a locally invasive disease with a poor prognosis if detected late. Surgery is the only proven therapy. Isolated hepatic metastases being managed surgically has been reported. However, extensive, staged, surgical resection of hepatic and pulmonary metastases has not been reported.

CONCLUSION: Lacrimal ACC is an aggressive tumour with known late metastases with the liver being affected seldomly. Our case highlights that even disseminated metastases to the liver and the lungs from lacrimal ACC can be managed surgically with a good outcome. To our knowledge, no such report with distant metastases to the liver and the lung along with their successful surgical management has been reported.

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1. Introduction

Adenoid cystic carcinoma (ACC) is a malignant neoplasm of the secretory glands. It is commonly seen in middle aged females. Lacrimal gland ACC though a rare tumour, is the commonest malignant epithelial neoplasm of the lacrimal gland [1].

However, the related poor prognosis is well known. Despite having a slow growth, late distant metastases to the lung and bones have been reported [2]. Metastases to the liver is seldom encountered, and when found is usually beyond surgical cure due to wide spread disease dissemination.

We present a case of lacrimal ACC that was surgically treated followed by chemoradiotherapy. After 5 years she came with distant multiple liver and pulmonary metastases. To our knowledge, such

simultaneous hepatic and pulmonary metastases from a lacrimal ACC has not been reported so far. We surgically managed the metastases by a right hepatectomy followed by a staged bilateral pulmonary metastatectomy.

2. Presentation of case

A 42 year old lady presented with complaints of vague right upper abdominal pain. She had undergone a left orbitotomy and excision of lacrimal gland for lacrimal adenoid cystic carcinoma five years ago. She received 5 cycles of chemotherapy with leucovorin, cisplatin and 5-FU along with 27 fractions of radiotherapy (5600 rad). On examination she had mild tenderness in the right hypochondrium with hepatomegaly. She had a normal liver profile and tumour markers. Her orbital imaging studies were normal. The computed tomography (CT) of the chest and abdomen revealed a large 10 × 8 × 5 cm lesion in segment VI and another lesion in segment IVB about 4 × 3 × 3 cm in segment IV superficially with multiple bilateral pulmonary irregular nodules **Images 1 and 2**. FNAC of the liver lesion confirmed metastatic adenoid cystic disease

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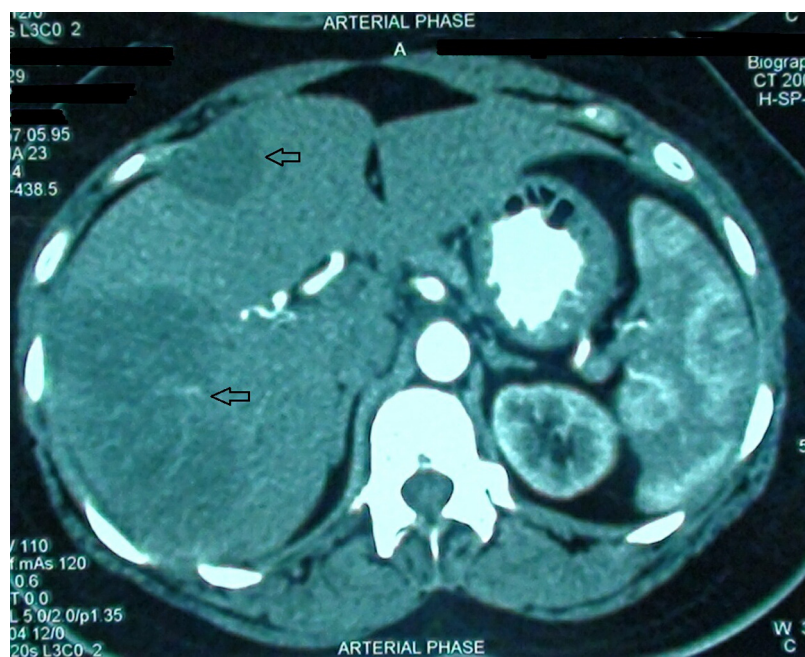


Image 1. CT abdomen showing a large $10 \times 8 \times 5$ cm lesion in segment VI and another lesion in segment IVB about $4 \times 3 \times 3$ cm in segment IV superficially.

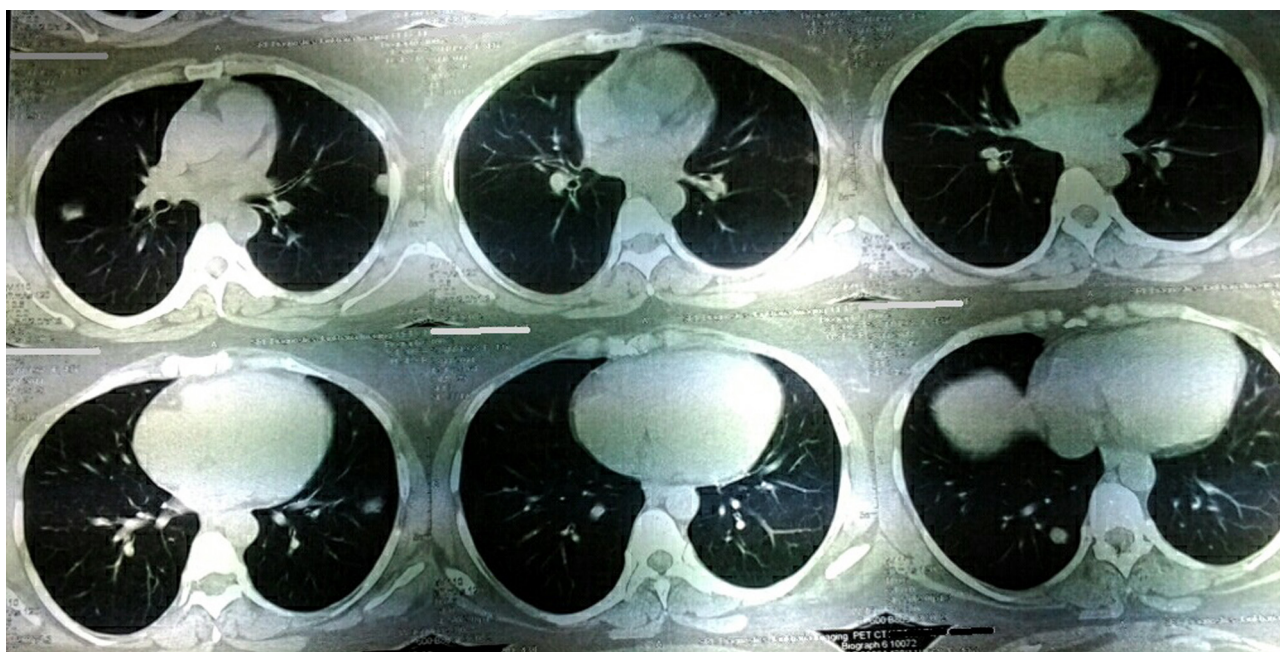


Image 2. CT chest showing multiple bilateral pulmonary irregular nodules.

of cribriform type. We further evaluated her with a Positron Emission Tomography that confirmed the metastatic disease confined to the liver and lungs only. After discussion with our multidisciplinary team, we decided to do staged resection as patients overall general and physical condition did not allow to undergo upfront extensive surgical resection of both liver and pulmonary metastases in the same sitting. We planned resection of the liver disease first due to a higher standardized uptake value (SUV) followed by the pulmonary lesions as they had a lower SUV. Intra-operatively, we found a lesion in segment VII that was stuck to the anterior abdominal wall and also to the right hemidiaphragm and another separate smaller lesion in segment IVB. The rest of the liver looked normal. Right hemi-hepatectomy with segment IVB resection was

done. Another single 1.5×1 cm metastatic nodule was excised from the posterior peritoneal surface over the diaphragm (**Image 3A**). Postoperatively the patient recovered well. Histopathology confirmed metastases of adenoid cystic carcinoma of cribriform type to the liver with clear margins (R0 resection) (**Image 4**).

On regular follow up, the multiple pulmonary nodules had marginally increased in size. After six months, we planned a staged pulmonary metastatectomy due to reduced pulmonary function. First, we performed muscle sparing left thoracotomy with metastatectomy for 9 pulmonary nodules (**Image 3B**). We repeated the procedure on the right side after a month and 8 pulmonary nodules were removed. These also showed metastatic nodules of classical adenoid cystic carcinoma of cribriform type. She tolerated the

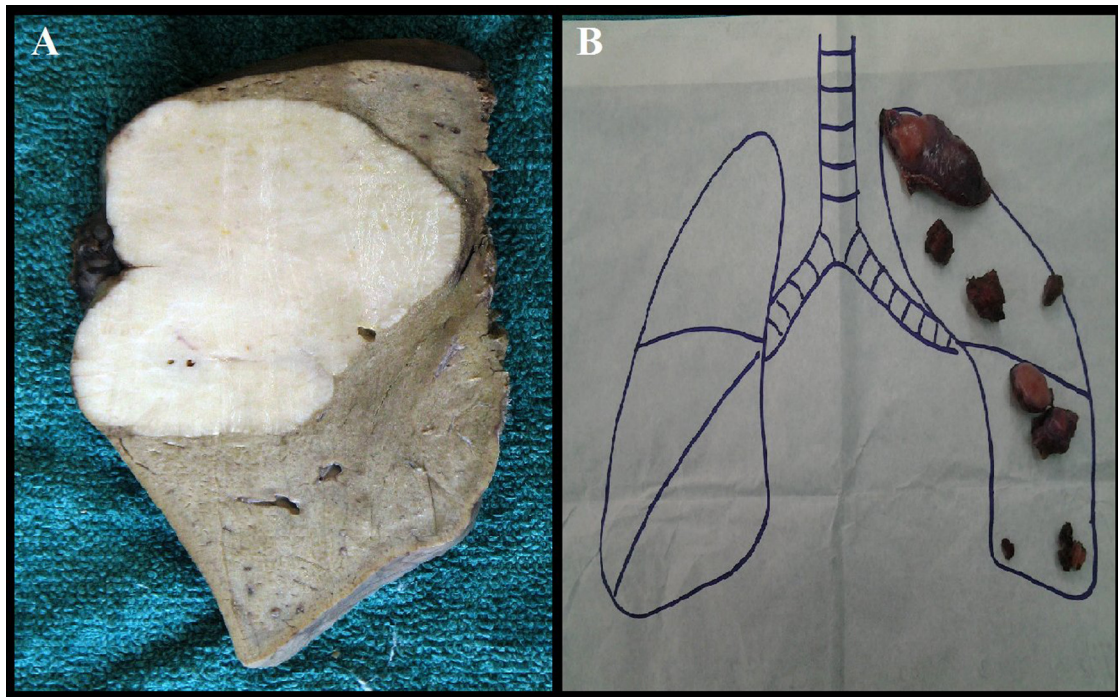


Image 3. (A) Cut section of right hepatectomy showing greyish white tumour (9.8 × 5.5) cm reaching the capsular surface, closest margin of resection 0.5 cm, (B) pulmonary metastatic lesions following left muscle sparing thoracotomy.

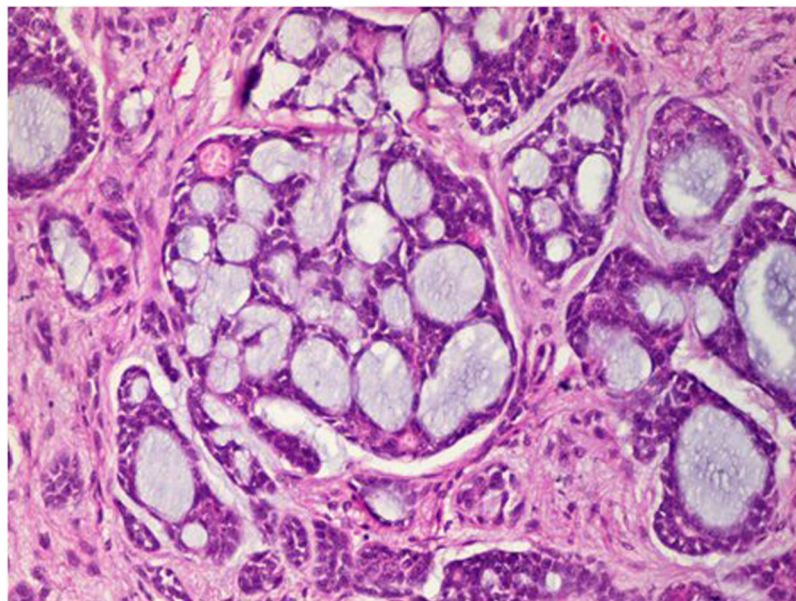


Image 4. H&E 20X microphotograph typical adenoid cystic pattern with small cells arranged in lobules.

procedure well. There is no recurrence of disease after 18 months of second thoracotomy.

3. Discussion

Lacrimal ACC accounts for 1.6% of all orbital tumours [2,3]. Usually asymptomatic, the commonest symptom is pain in the region of the lacrimal gland. ACC is also found in the skin, breast, cervix, prostate gland, external auditory canal, nasopharynx, lacrimal glands, vulva, esophagus, and trachea. It is a locally invasive disease with a poor prognosis if detected late [4,5].

Local recurrence is generally the way of progression of disease. Lacrimal ACC rarely shows lymphatic spread. However, perineural spread is commonly seen [6,11]. The incidence of distant metastases of overall ACC is 25–55%. The five year survival rate of these patients is 20% [1]. The lungs, brain and bones are the common sites of distant metastases [7–9]. Our patient had liver metastases along with pulmonary metastases, with the former being a rare site of metastases. Late diagnosis, perineural spread, solid growth pattern, positive margins and local recurrence despite radiotherapy account for the poor outcome of the disease [10,12].

The only proven mode of therapy in lacrimal ACC is surgical resection of the tumour. Post operative radiotherapy may be useful

in cases of close or positive margins. Our patient probably represents the first case of lacrimal ACC that presented with hepatic and pulmonary metastases simultaneously five years after primary resection and was still managed surgically with a good outcome.

Surgical cure for lacrimal ACC is challenging due to complex orbital anatomy as well as its aggressive behavior. Isolated hepatic metastases from lacrimal ACC being managed surgically has been reported [13]. However, extensive, staged, surgical resection of hepatic and pulmonary metastases has not been reported.

There is no substantial evidence regarding the benefit of chemotherapy and radiotherapy in the management of primary as well as metastatic ACC. Intra Arterial Cytoreductive Chemotherapy is now being considered as adjuvant therapy [14].

4. Conclusion

Lacrimal ACC, though rare is an aggressive tumour with distant metastases occurring late. Surgery is the only effective mode of therapy for both primary and metastatic disease. Lifelong follow-up is necessary for early detection and management.

Conflicts of interest

None.

Funding

None.

Ethical Approval

Approval given from Lilavati Hospital And Research Centre.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contribution

Sali Priyanka A. and Yadav Kamal S. wrote the paper Bushan Kirti designed the study Rajpurohit Virendra, Varty Paresh and Sharma Sanjay analysed the same.

Guarantor

Sali Priyanka A., Sharma Sanjay.

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