A REPORT OF MAXILLARY SINUS ACINIC CELL CARCINOMA

Jessica Filbertine a, Ni Putu Setiawathi b and I Ketut Suanda c

a General Practitioner, Wangaya Regional General Hospital, Denpasar, Bali, Indonesia, b Otorhinolaryngologist, Department of Otorhinolaryngology-Head and Neck Surgery, Wangaya Regional General Hospital, Denpasar, Bali, Indonesia, c Otorhinolaryngologist – Oncology Consultant, Department of Otorhinolaryngology-Head and Neck Surgery, Medical Faculty of Udayana University/ Sanglah Central General Hospital, Denpasar, Bali, Indonesia

ABSTRACT

Introduction: Acinic cell carcinoma (ACC) is a rare malignant tumour of the salivary gland. It usually shows in the parotid gland and rarely shows in another site. Case Report: A 33 years old female from East Nusa Tenggara came to our outpatient clinic with a complaint of nasal obstruction and recurrent epistaxis on her right nose 1 year ago. The biopsy for the tumor was done by the previous Ear-Nose-Throat (ENT)-specialist and resulted in suggestive pleomorphic adenoma. During her visit, we found a red-coloured mass occupying the right nasal cavity emerging from the middle meatus. The patient had not received medical nor non-medical therapy after the biopsy and declined to have epistaxis episodes during the time. Nasal obstruction was her current complaint and what brought her to seek medical care. We continued with a midfacial degloving surgery and removed the tumour, which aroused from the maxillary sinus. Histopathology of the tumour shows an acinic cell carcinoma feature. Discussion: We searched all English and Non-English previously ACC reports in the nasal cavity to compare our case and treatment plan. Conclusion: Sinonasal ACC is a rare, slow-growing, low-grade malignancy tumour with an excellent outcome, despite indistinct symptoms.

KEYWORDS acinic cell carcinoma, sinonasal tumour, rare case

Introduction

Acinic cell carcinoma (ACC) is an uncommon malignant tumour of the salivary gland which may rarely present in ectopic sites. It usually arises in the major salivary glands and minor salivary glands, commonly in the parotid gland of more than 80% of ACC cases, with an incidence of around 2-4% of all parotid gland tumours. ACC can also arise in lips, palate, neck, buccal mucosa and other ectopic sites, with only around 17% arising intraorally, 4% from the submandibular gland and less than 1% from the sublingual gland. [1-4]

This tumour has a slow-growing, low-grade behaviour. It is known to have an excellent long-term survival outcome amongst any salivary malignancy.[1,5-6] Acinic cell carcinoma (ACC) develops from tumorigenesis of cells responsible for acinar development. Histopathological patterns include solid, papillary-cystic, follicular, and microcystic, with solid being the most common. Although acinic cell carcinoma is usually considered a low-grade malignancy, it can transform into high-grade adenocarcinoma or undifferentiated carcinoma. ACC may show a complete loss of acinar differentiation, desmoplasia, tumour necrosis, frequent mitosis, and cervical or distant metastasis. All acinic cell carcinomas show lymph node and distant metastases in about 50% of cases and require radical surgery, treatment of the lymph nodes and adjuvant radio-chemotherapy. [4,7]

Case Report

A female patient, 33 years old from East Nusa Tenggara, came to our outpatient clinic with a complaint of nasal obstruction and recurrent epistaxis on her right nose. She first had an episode of epistaxis 1 year ago. A total of 8 episodes with the two had caused her to seek medical care caused to profuse bleeding. Two months later, she visited an Ear-Nose-Throat (ENT)-a specialist in Kupang, East Nusa Tenggara. The ENT specialist realised
that she had a tumour on her right nose. She was then planned for a biopsy for the tumour. The specimen was grey and brown-black coloured, microscopically shown ductal epithelial cell with an oval-round nucleus, relatively monotone, hyperchromatic, cytoplasm partly vacuolated. Seen forming a microacini structure. The stroma consists of fibrohyaline tissue. Seen myxoid focus area. No signs of malignancy. Biopsy result suggestive of pleomorphic adenoma. No previous medical history nor family history contributory to the disease.

Figure 1 Patient’s appearance from front and tumour side of the face. Tumors nor destruction can’t be seen from inspection of the face.

During her visit, we did not find anything peculiar during the inspection (figure 1). However, we found a blood-red coloured mass occupying the right nasal cavity, which emerges from the middle meatus as shown in figure 2. The patient did not visit any ENTs during these 10 months for personal reasons, had no medical nor non-medical therapy and declined epistaxis episodes during the time. Nasal obstruction was her current complaint and what brought her to seek medical care.

We then conducted a contrast-enhanced soft tissue setting computed tomography (CT) scan of the Paranasal Sinuses. CT found solid mass with a clear margin size estimation 4 x 4.24 x 3.89 cm occluding the right maxillary sinus cavity, with strong contrast enhancement (87 HU density). The mass erodes the medial wall of the right maxillary sinus, expanding to the right nasal cavity, eroding the right middle conchae, suppressing the nasal septum to the left, and expanding to the ethmoid and right frontal sinus. In addition, the right sphenoethmoid recess is impedied by the mass. No enlarged lymphoid gland was seen on the CT result (figure 3).

To elaborate on the mass, we plan on having lateral rhinotomy with a midfacial degloving approach and plan for complete excision of the tumours. Therefore surgical preparation was done, which were blood laboratories of haemoglobin, leukocyte, erythrocyte, blood sugar, kidney function, liver function, prothrombin time (PT), and activated partial thromboplastin time (aPTT) and urinalysis. All her laboratory test results are within normal limits, except her PT and aPTT prolonged results. PT was 13.0 seconds (normal = 9.5 – 11.7 seconds) and aPTT was 85.4 seconds (normal = 25.5 – 42.1 seconds). Therefore we consulted with an internist specialist, and she was given three times daily Vitamin K intravenously for five days before her surgery permitted. On the sixth day, we conducted another PT and aPTT test, resulting in normal results. Chest radiography is also done with within the normal result.

During surgery, we found a reddish-white coloured tumour mass occupying the right maxillary sinus cavity destructing the anterior and medial walls of the maxillary sinus wall through the nasal cavity. The tumour was also found from the nasal cavity up to the ethmoid sinuses. Ethmoid sinuses were filled with a mucous secret. Specimen then sent to pathology anatomy.

Tissue preparation consists of surface epithelia, connective tissue stroma, compact bone tissue and tumour mass. Tumour mass consists of atypic oncocytic epithelial cell proliferation, hobnail cell, and vacuolated/foamy cell. Cells are composed, forming solid/lobular and microcystic. The nucleus appears enlarged with increasing nucleus/cytoplasmic (N/C) ratio, eccentric, hyperchromatic up to vesicular; some have nucleolus. Mitosis can be found. The cytoplasm is partly granular and eosinophilic. Intratumoral stroma appears to be hyalinised and infiltrated by lymphoid cells (figure 4).

Conventional morphology implies a malignant epithelial salivary gland neoplasm with acinar features. Therefore, the diagnosis is sinonasal ACC, with a differential diagnosis of secondary ACC.

Figure 2 Anterior rhinoscopy (left) and nasal endoscopic (right) of the right nasal cavity show bloody-red coloured mass occupying the nasal cavity, emerging from the middle meatus.

Jessica Filbertine  et al./ International Journal of Medical Reviews and Case Reports (2022) 6(11):64-68
We examined lung and bone surveys during follow-up, and no metastatic lesion was visualized. Liver and para-aorta ultrasonography showed no metastatic nodule. Blood tests are within normal results.

Building upon current findings, the patient is now in T3N0M0, stage III, according to the 8th edition of the American Joint Committee on Cancer (AJCC) TNM Staging System. The patient was referred to Sanglah Central General Hospital for the next treatment plan for radiotherapy and will be given chemotherapy while waiting for the radiotherapy schedule.

**Discussion**

ACC is a rare, slow-growing, low-grade malignancy tumour of the salivary gland.[8] It represents nearly 6% of all salivary gland tumours and less than 17% of primary malignant salivary gland tumours. The tumour is most frequently (90-95%) detected in the parotid gland.[6] Sinonasal ACC is very rare globally. Sinonasal ACC usually arises in the lateral nasal wall and turbinates.[8] From the literature search, we found 33 other Sinonasal Acinic Cell Carcinoma reports in Pubmed, and Google Scholar databases and only 25 were written in English (table 1). To our knowledge, this is the first report in our country and the 13th report in Asia/Pacific.

Previous reports of ACC in the nasal cavity are predominantly in females (61-63%), as in our case. However, age range from the 40s to 70s with a mean age of 56-57,2 years and the majority of patients were of the white race (77,8%), unlike our patient who is 33 years ago and is Asian.[3,5,9] Symptom duration before diagnosis ranges from 2 months to 10 years (table 1).

Overall ACC 5-year survival rate is 75% to 96% and 56% at 20 years.[4] The 2014 meta-analysis of survival by Biron et al. in sinonasal ACC of 19 sinonasal ACC cases showed 92.9% estimated 5-year and 10-year disease-specific survival (DSS). Despite a high DSS rate, tumours in sinonasal had indistinct symptoms leading to larger tumour size and more advanced clinical T-stage during diagnosis.[5,10] Also, patient with an older age has been associated with a decreased survival rate.[2] In Biron et al.’s report, patients treated with only surgery were 72.2% of cases, and the rest of the cases were given radiation postoperatively.[5] Spiro et al. reported a 33% local recurrence rate with 12% distant metastasis for all ACC ranging from one month to 19 years after initial treatment (median 2 years), in contrast to a previous report by Menace & Goldman’s of a higher recurrence rate of as high as 50 to 60% and may occur as late as 30 years post-surgery. Long-term follow-up must be done in all ACC cases.[11,12]

Treatment for salivary gland malignancies is mainly surgical, in the absence of distant hematogenous metastases.[13] There are no adjuvant radiotherapy requirements in sinonasal ACC if the disease is localized, well-differentiated and the surgical margins are clear.[5,14] Postoperative radiation therapy in ACCs suggested in advanced-stage diseases with nodal metastasis, high-grade (poorly differentiated) pathology, and positive margins. However, no studies had reported the survival benefit or recurrence rate improvement with this treatment modality.[2,5] The use of systemic chemotherapy in all salivary gland malignancies is limited to an advanced and incurable diseases. It is indicated for palliative treatment of metastatic or locoregional recurrence uncontrolled by surgery or radiotherapy. Cisplatin and cisplatin-based regimens have been most frequently explored, with a minimum response. However, adjuvant radio-chemotherapy is supposed to increase overall survival in advanced tumour stages in all salivary gland carcinomas. The study shows death occurred within 3 to 10 months after a distant metastasis was revealed.[7,11,13]

Tissue biopsy is needed as a basis for diagnosing a tumour.[2] Our patient’s biopsy from a part of the tumour, done before she came to our centre, shows pleomorphic adenoma. It is most likely for the biopsy result to be different from the final diagnosis of a case, as the biopsy takes only a small part of the whole tumour, so it might not be representative enough. Small ACC tumours can also be easily missed as the acinar cells are so well differentiated that they blend into the surrounding normal gland.[15]
**Table 1** Review of previously reported Acinic Cell Carcinoma(s), with 8 articles written in Non-English language; therefore, we could only review them through abstracts.

| No. | Author | Report year | Age | Sex | Location | Symptoms | Duration from the first symptom(s) | Treatment | Recurrency |
|-----|--------|-------------|-----|-----|----------|----------|------------------------------------|-----------|------------|
| 1   | Kleinsasser | 1970 | N/S | N/S | N/S | Nasal obstruction | N/S | N/S | N/S |
| 2   | Menace  | 1971 | 47 | F  | Ethmoid + maxillary sinus | Nasal obstruction | 10 months | Surgery | - |
| 3   | McCarthy | 1973 | N/S | N/S | AC Adenocarcinoma Maxillary sinus | N/S | N/S | N/S |
| 4   | Spiro et al. | 1978 | N/S | N/S | N/S | Nasal obstruction | N/S | N/S | N/S |
| 5   | Perzin et al. | 1981 | 75 | F  | Inferior turbinate | Nasal obstruction, epistaxis | N/S | Surgery | - |
| 6   | Ordonez et al. | 1986 | 60 | F  | Superior meatus | Nasal polyp, epistaxis | N/S | Surgery | - |
| 7   | Finkelhor et al. | 1987 | 45 | F  | Septum | Nasal obstruction | N/S | Surgery | - |
| 8   | Hanada et al. | 1988 | 68 | M  | Inferior turbinate | Nasal obstruction | N/S | Surgery + RT | - |
| 9   | Takimoto et al. | 1989 | 60 | F  | Middle + Inferior turbinate | Nasal polyp, epistaxis | N/S | Surgery | - |
| 10  | Dimitrakopoulos et al. | 1992 | 65 | M  | Maxillary sinus | Swelling and intermittent pain, diplopia | 2 months | N/S | - |
| 11  | Valerdiz-Cassasola et al. | 1993 | 47 | M  | Nasal cavity | Nasal obstruction, epistaxis | N/S | Surgery + RT | - |
| 12  | Schmitt et al. | 1994 | 60 | M  | Inferior turbinate | Nasal obstruction | N/S | N/S | - |
| 13  | Sim et al. | 1995 | 60 | M  | Nasal cavity | N/S | N/S | Surgery + RT | - |
| 14  | Yoshihara et al. | 1995 | 41 | M  | Maxillary sinus + parotid | N/S | N/S | Surgery + RT + C | - |
| 15  | Fujii et al. | 1998 | 71 | F  | Maxillary sinus | N/S | N/S | Surgery x2 | 4 year recurrence |
| 16  | Von Biberstein et al. | 1999 | 76 | F  | Middle turbinate | Nasal polyp | N/S | Surgery | - |
| 17  | Jasin et al. | 1999 | 44 | F  | Nasal septum | N/S | N/S | Surgery | - |
| 18  | Son et al. | 2000 | 57 | F  | Nasal cavity | Nasal obstruction, post-nasal drip, hyposmia | 2 months | Surgery | - |
| 19  | Sapçi et al. | 2000 | 47 | M  | Nasal septum | Nasal obstruction, epistaxis | N/S | Surgery | - |
| 20  | Yoon et al. | 2002 | 46 | M  | Maxillary sinus | Palatal and buccal swelling, nasal obstruction, lacrimal, maxillary sinus, exophthalmos | 4 years | Surgery | 3 years recurrence |
| 21  | Neto et al. | 2005 | 50 | M  | Nasal cavity | Nasal obstruction | N/S | Surgery + RT | 1 year recurrence |
| 22  | Hammami et al. | 2009 | 47 | F  | Nasal septum | Nasal obstruction, Hyposmia | 1 year | Surgery | - |
| 23  | Manganaris et al. | 2010 | 51 | F  | Nasal vestibule | Localized pain | N/S | Surgery | - |
| 24  | Wong et al. | 2010 | 42 | F  | Ethmoid sinus | Nasal obstruction, yellow-to-bloodstained nasal discharge, 8 years of chronic supplicative otitis media | 1 year | Surgery | - |
| 25  | Triantafillidou et al. | 2010 | 65 | M  | Maxillary sinus | Swelling and intermittent pain at the left maxilla, maxillary sinus, nasal cavity, Diplopia and infraorbital nerve paresthesia | 2 months | Surgery + RT | 3 years follow-up. Died from the disease (endocranial spread) |
| 26  | Somayaji et al. | 2014 | 65 | F  | Nasal cavity | Epistaxis, Nasal obstruction | 1 year | Surgery | - |
| 27  | Tang et al. | 2016 | 67 | M  | Nasal septum | Nasal obstruction, bloodstained nasal discharge | 4 years | Surgery + RT | - |
| 28  | Hou et al. | 2017 | 63 | F  | Nasal septum | Nasal obstruction, hyposmia, epistaxis | 2 years | Surgery | - |
| 29  | Li et al. | 2018 | 72 | N/S | Nasal obstruction, bloody sputum, loss of olfaction | N/S | Surgery | - |
| 30  | Nasereddine et al. | 2019 | 84 | M  | Nasal septal | Epistaxis, Nasal obstruction | N/S | Surgery | - |
| 31  | Nasereddine et al. | 2020 | 58 | N/S | Nasal cavity | N/S | N/S | - |
| 32  | Küçük et al. | 2020 | 59 | M  | Maxillary sinus | Nasal obstruction | 1 year | Surgery | - |
| 33  | Dionisio et al. | 2021 | 62 | M  | Lateral nasal wall | Nasal mass, epistaxis | 10 years | Surgery | - |
| 34  | This report | 2021 | 33 | F  | Maxillary sinus | Nasal mass, epistaxis | 1 year | Surgery + planned for C and RT | - |

**Abbreviation:** AC, Acinic Cell; F, female; M, male; RT, radiotherapy; C, chemotherapy; N/S, not specified.

Jessica Filbertine et al./ International Journal of Medical Reviews and Case Reports (2022) 6(11):64-68
Conclusion

Sinonasal ACC is a rare, slow-growing, low-grade malignancy tumour, despite indistinct symptoms. Recurrence is between 33-60%, ranging from less than a month in our case up to 10 years post-operatively. A biopsy is important as a diagnosing basis. However, histopathology results may differ between a small part of the tumor or after complete resection. Adjuvant radio-chemotherapy may be added to the treatment to increase overall survival.

Acknowledgments

The author would like to thank dr. Novitasari, Sp.PA for helping us in pathological analysis. We would like to thank dr. Made Prani WindaSari and dr. Grace Filbertine, M.Sc., for the input in making this case report.

Ethical Approval and Consent

The patient has given consent for the case to be published.

Funding

This work did not receive any grant from funding agencies in the public, commercial, or not-for-profit sectors.

Conflict of Interest

There are no conflicts of interest to declare by any of the authors of this study.

References

1. Somayaji KSG, Ali ZS, Abdulla MN, Nambiar VR, Johns TK. Acinic cell carcinoma of the nasal cavity. Arch Med Health Sci. 2014 Jan 1;2(2):220–2.

2. Naserrudin NS, Wahab SA, Ramanna VR, Ahmad AR. Primary nasal septal acinic cell carcinoma. Romanian J Rhinol. 2019 Mar 1;9(33):39–42.

3. Iman W, Merung M, Aschorijanto A. Profil penderita tumor kelenjar liur di RSUP Prof. DR. R.D. Kandou Manado periode Juli 2012-Juni 2015. E-Clin [Internet]. 2016 [cited 2021 Nov 26];4(1).

4. Young A, Okuyemi OT. Malignant Salivary Gland Tumors. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2021 [cited 2021 Nov 21]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK 563022/

5. Biron VL, Lentsch EJ, Gerry DR, Bewley AF. Case-control analysis of survival outcomes in sinonasal acinic cell carcinoma. Int Forum Allergy Rhinol. 2014 Jun;4(6):507–11.

6. Küçük Ü, Ekmekçi S, Çukurova I. Psammoma Body–Rich Acinic Cell Carcinoma of Maxillary Sinus. Dokuz Eylül Üniversitesi Tip Fakültesi Derg. 2020 Aug 31;34(2):175–9.

7. Ettl T, Schwarz-Furlan S, Gosau M, Reichert TE. Salivary gland carcinomas. Oral Maxillofac Surg. 2012 Sep 1;16(3):267–83.

8. Hou S, Wei J, Tian Y. Primary acinic cell carcinoma of the nasal septum: a case report and review of the literature [Internet]. 2017 [cited 2021 Dec 30]. Available from: https://www.semanticscholar.org/paper/Primary-acinic-cell-carcinoma-of-the-nasal-septum-%3A-Hou-Wei/b0ff42dc0dfd668b86252681a84e8b5 53238a929

9. Dionísio S, Ventura E, Gonçalves J, Nobre R, Marques H. Acinic Cell Carcinoma of the Nasal Lateral Wall. Cureus [Internet]. 2021 Oct 13 [cited 2021 Nov 11];13(10).

10. Lee S-Y, Shin HA, Rho KJ, Chung HJ, Kim S-H, Choi EC. Characteristics, management of the neck, and oncological outcomes of malignant minor salivary gland tumours in the oral and sinonasal regions. Br J Oral Maxillofac Surg. 2013 Oct 1;51(7):e142–7.

11. Spiro RH, Huvos AG, Strong EW. Acinic cell carcinoma of salivary origin. A clinicopathologic study of 67 cases. Cancer. 1978 Mar;41(3):924–35.

12. Manace ED, Goldman JL. Acinic cell carcinoma of the paranasal sinuses. The Laryngoscope. 1971 Jul; 81(7):1074–82.

13. Adelstein DJ, Koyfman SA, El-Naggar AK, Hanna EY. Biology and Management of Salivary Gland Cancers. Semin Radiat Oncol. 2012 Jul 1;22(3):245–53.

14. Wong A, Leong JL, Ho B. Primary acinic cell carcinoma of the ethmoid sinus. Ear Nose Throat J. 2010 Jul;89(7):E40-41.

15. Sirjani DB, Lewis Jr JS, Beadle BM, Sunwoo JB. Malignant Neoplasms of the Salivary Glands. In: Flint PW, Francis HW, Haughey BH, Lesperance MM, Lund VJ, Robbins KT et al., editors. Cummings Otolaryngology Head and Neck Surgery. 7th ed. Philadelphia: Elsevier; 2021.p.1197-1198.