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SINONASAL ADENOID CYSTIC CARCINOMA: A CASE REPORT

Otorhinolaryngology

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ABSTRACT

Introduction: Adenoid cystic carcinoma (ACC) is an uncommon malignant tumour characterized by indolent growth, perineural spread, and a high likelihood of local recurrence and distant metastasis, particularly to the lungs and bones. **Case Report:** We describe a 28-year-old woman with an 8-year history of right-sided nasal swelling and 6-year history of nasal obstruction and discharge. Contrast-enhanced computed tomography revealed a heterogeneously enhancing sinonasal mass involving adjacent structures. Fine-needle aspiration cytology was suggestive of a benign salivary gland lesion. She underwent complete tumour excision via lateral rhinotomy with functional endoscopic sinus surgery. Histopathological examination of the excised specimen confirmed adenoid cystic carcinoma, staged as pT3NxMx, followed by adjuvant radiotherapy (60 Gy in 30 fractions).The prolonged symptom duration highlights the slow-growing nature of this tumour and the potential for delayed diagnosis **Conclusion:** A high degree of clinical suspicion is warranted for prolonged unilateral nasal complaints, as sinonasal ACC can mimic benign conditions on initial cytology. Radical surgical excision followed by postoperative radiotherapy remains the standard treatment.

KEYWORDS

Adenoid Cystic Carcinoma, Paranasal Sinus Neoplasms, Radiotherapy, Nasal Cavity

INTRODUCTION

Adenoid cystic carcinoma (ACC) was originally described by Billroth as "cylindroma" (Bavikar et al., 2024). It arises predominantly in the major and minor salivary glands and is only rarely encountered in the sino-nasal region (Apoorva et al., 2022; Rassam & Sood, 2022; Gurung et al., 2022). Sinonasal involvement shows a female predominance, with the maxillary sinus affected more frequently than the nasal cavity. Patients typically present with nasal discharge, obstruction, facial pain, headache, or cheek swelling (Bavikar et al., 2024). ACC follows an indolent course, with a marked predisposition for perineural invasion, and local recurrence and distant metastasis, particularly to the lungs and bones, occurring at later stages of disease (Gurung et al., 2022).

Because of its non-specific presentation, sinonasal ACC is easily mistaken for benign inflammatory conditions, delaying diagnosis. We report a case of sinonasal ACC in a young woman to emphasize the need for sustained clinical suspicion in patients with persistent unilateral nasal symptoms, and the value of timely, multidisciplinary management.

Case Report

A 28-year-old female presented in the outpatient department with complaints of right-sided nasal swelling over the maxillary region for 8 years, associated with right-sided nasal obstruction and discharge for 6 years. She had no history of nasal trauma or bleeding, and no relevant family history.

A single, non-tender swelling of soft-to-firm consistency was noted over the right dorsum of the nose, measuring 4x4x3 cm, with no local rise in temperature. The overlying skin was normal, with no fistula, sinus, or tract (Figure 1). The right nasal cavity was obliterated by a mass arising from the lateral wall of the nose. A deviated nasal septum was noted on the left side.

Figure 1. Pre op photo of patient front view

Contrast-enhanced computed tomography of the paranasal sinuses showed a well-defined, heterogeneously enhancing soft tissue density measuring 6.4x4x5.7 cm epicentred in the right nasal cavity, with extensions superiorly to the inferior and middle turbinates, medial thinning of the nasal septum, lateral involvement of the medial wall of the right orbit, and inferior thinning of the hard palate (Figure 2).

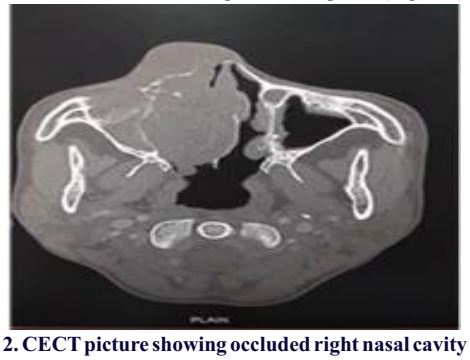

Figure 2. CECT picture showing occluded right nasal cavity

Fine-needle aspiration cytology showed highly cellular smears with ductal epithelial cells arranged in clusters, sheets, and groups, with round, regular nuclei and eosinophilic cytoplasm; a few cells showed mild anisonucleosis. Myoepithelial cells and myoid areas were noted, along with eosinophilic hyaline globules surrounded by ductal epithelial cells, against a haemorrhagic background. The findings were suggestive of a benign epithelial lesion, consistent with cellular pleomorphic adenoma.

Blood investigations were within normal limits (haemoglobin 9.9 g/dL, total leukocyte count 6700/microL, platelet count 5.48 lakh/microL, blood urea 15 mg/dL, serum creatinine 0.6 mg/dL, SGOT 18 U/L, SGPT 12 U/L, serum bilirubin 0.4 mg/dL, blood group B positive). Screening for HIV, Hepatitis B surface antigen, and Hepatitis C virus was negative.

Right nasal mass excision via lateral rhinotomy approach with functional endoscopic sinus surgery was performed (Figure 3). Histopathological examination of the excised specimen confirmed adenoid cystic carcinoma, staged as pT3NxMx (Figure 4).

Figure 3. Post op day 11

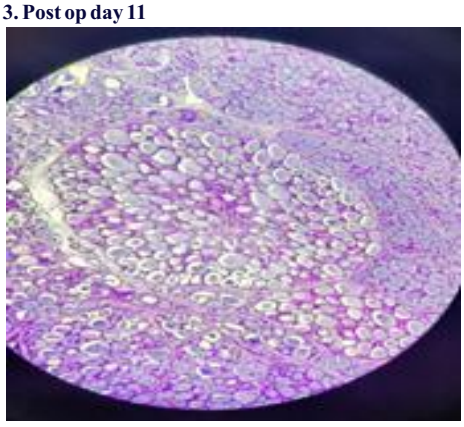

Figure 4. Histopathology picture of the tumour

Adjuvant radiotherapy (60 Gy in 30 fractions) was administered over 6 weeks without concurrent chemotherapy, delivered via linear accelerator using intensity-modulated/volumetric modulated arc radiotherapy technique.

Post-operative CECT of the paranasal sinuses on day 27 showed thinning with left lateral displacement of the nasal septum, thinning of the right hard palate with erosion of the right maxillary alveolus extending to the roots of the right upper incisors and canine, erosion of the uncinate process with blockage of the right osteomeatal complex, and erosion of the right nasal bone with overlying soft tissue thickening at the right nasal and premaxillary region .

DISCUSSION

Adenoid cystic carcinoma originates chiefly in the major and minor salivary glands, though the ceruminous and lacrimal glands can also be affected; the sinonasal tract remains a distinctly unusual primary site (Bavikar et al., 2024; Gurung et al., 2022). It occurs in both sexes, most often during the fifth through seventh decades, and several case series have noted a female preponderance (Bavikar et al., 2024; Apoorva et al., 2022).

The tumour typically pursues a slow, indolent course, although extensive local invasion can occur; intracranial or intradural extension, by contrast, is exceedingly uncommon (Rassam & Sood, 2022). Because its early growth produces vague symptoms — nasal obstruction, discomfort, and epistaxis — the disease is often mistaken for sinusitis or allergic rhinitis, and diagnosis is frequently delayed as a result (Bavikar et al., 2024; Apoorva et al., 2022; Gurung et al., 2022). With continued growth, the tumour may erode into surrounding bone and soft tissue, including the orbit, pterygomaxillary fossa, meninges, and skull base, producing more alarming features such as ophthalmalgia, otalgia, proptosis, headache, and seizures (Bavikar et al., 2024; Lahjaouj et al., 2021). Even though its clinical course is generally indolent, the tumour is well known for local recurrence and distant spread, sometimes emerging years after what appeared to be complete surgical clearance (Bavikar et al., 2024; Apoorva et al., 2022).

Three histological subtypes are recognised — tubular, cribriform, and solid — with the solid pattern carrying the poorest prognosis of the three (Apoorva et al., 2022). Perineural invasion occurs frequently and is considered characteristic of the disease (Apoorva et al., 2022; Gurung et al., 2022). Immunohistochemical staining can aid diagnosis, as tumour cells commonly express CD117, reflecting their ductal derivation (Apoorva et al., 2022).

Surgery remains the mainstay of treatment, with adjuvant radiotherapy added where indicated. Where intradural invasion is present, however, the associated neurological risk usually rules out resection, and radiotherapy-based treatment is favoured instead. Pooled data place the 5-year survival for sinonasal ACC at roughly 62% (Rassam & Sood, 2022). A more recent comparative study found that surgery followed by postoperative radiotherapy outperformed definitive radiotherapy given with or without concurrent chemotherapy, yielding 5-year local failure-free survival of 61.8% versus 37.8%, and overall survival of 93.2% versus 73.4% (Lee et al., 2024).

This patient's presentation departs from the typical epidemiological picture of sinonasal ACC in two respects. She was 28 years old — well below the fifth-to-seventh-decade window in which the disease usually presents — and her symptom history spanned an unusually long interval before diagnosis, in keeping with the slow, non-specific evolution described above.

CONCLUSION

Sinonasal adenoid cystic carcinoma is rare but should not be dismissed, and it warrants clear distinction from more common benign nasal masses, particularly when symptoms are prolonged and non-specific. This case illustrates that the disease can present atypically, reinforcing the need for sustained clinical vigilance. Complete surgical excision followed by adjuvant radiotherapy remains the treatment of choice and was applied successfully in this patient.

Statements And Declarations

Author Contributions

All authors contributed to the conception and preparation of the case report. Material preparation, data collection, clinical management, and manuscript preparation were performed by the authors. The first draft of the manuscript was prepared by Dr. Pratyusha Rath, and all authors critically revised the manuscript. All authors read and approved the final version of the manuscript.

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Ethics Approval: Ethics approval was not required for this case report according to the policies of Government Medical College, Chhatrapati Sambhajinagar, Maharashtra, India.

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

Data Availability: Data supporting the findings of this case report are available from the corresponding author upon reasonable request.

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