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Adenoid Cystic Carcinoma of Sphenoid Sinus: Case Report of a Rare Entity

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Abstract

Adenoid Cystic Carcinoma (ACC) of sinonasal tract is a very rare, slow growing malignant tumour comprising less than 1% of head and neck cancers. It's presentation with non-specific symptoms at an advanced stage of tumour and proximity to important structures such as brain and cranial nerves, limits the management options intended for curative approach. A combination of surgery followed by adjuvant radiotherapy is the current choice of treatment. We describe a case of Sinonasal ACC originating from left Sphenoid sinus presenting with an isolated symptom of epistaxis in a 71-year-old female who was managed with the combination approach.

Keywords: ***Sinonasal tumour; Adenoid cystic carcinoma; Perineural invasion; Rare sinonasal malignant neoplasm; Epistaxis***

1. Introduction

Adenoid Cystic Carcinoma (ACC) is a rare malignant neoplasm accounting for 1% cases of Head and Neck cancers [1]. Primarily a tumour of minor salivary glands, very rarely it is reported as a primary tumour in sinonasal tract where it shows more aggressive pattern of growth as compared to its salivary gland counterparts [1]. Its tendency for slow growth and indolent course usually leads to a delayed diagnosis. At the same time, its locally aggressive nature along with proximity to vital structures such as brain and a high propensity for perineural invasion limits the extent of complete surgical management and hence leads to a higher incidence of local recurrence and delayed distant metastasis [2-4]. Surgery, along with adjuvant radiotherapy, remains the mainstay of treatment depending upon the stage of tumour [3,4]. Here, we present a rare case of ACC arising from the sphenoid sinus in a 70-year-old female with isolated symptom of epistaxis resulting in a delayed diagnosis.

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Computed Tomography (CT) scan of nose and paranasal sinuses showed a mass along the floor of left sphenoidal sinus. The case was managed successfully with surgical excision and adjuvant radiation therapy.

2. Case Report

A 70-year-old female patient, known case of Type 2 Diabetes Mellitus, reported to hospital after an episode of epistaxis from left nasal cavity. Epistaxis was insidious in onset and profuse (5 ml - 10 ml) in nature and was managed conservatively. She had two similar episodes of profuse epistaxis in the following 2 months. Upon detailed interview, a history of intermittent nasal obstruction was elicited. No significant findings were noted on local examination of bilateral nostrils. Local examination of ear cavity showed bilaterally intact and mobile tympanic membrane. Throat examination displayed no significant anomaly. There were no associated complaints of pain/ loss of smell/ headache/ toothache/ rhinitis/ fever/ vertigo/ double vision.

In view of recurrent epistaxis in a short span of time, she was advised CT scan of Nose and Paranasal sinuses which revealed an enhancing lobulated soft tissue mass along the floor of left sphenoid sinus measuring 2.3 cm × 1.28 cm × 1.9 cm with focal areas of erosion of sinus floor (FIG. 1a, b). Magnetic Resonance Imaging (MRI) was suggestive of enhancing lobulated soft tissue along the floor of left sphenoid sinus; focal areas of erosion was seen in left sphenoid sinus floor. On further clinical evaluation, patient was found to be hypertensive and was started on treatment for same. Thereafter, an endoscopic examination and biopsy was undertaken during which fragments of soft, friable reddish coloured tissue were removed. Histopathologic examination of those tissue bits revealed a moderately differentiated ACC with perineural invasion comprising mainly of cribriform and tubular pattern (FIG. 2 a, b, c, d). Subsequently, the patient was taken up for Endoscopic Sinus Surgery.

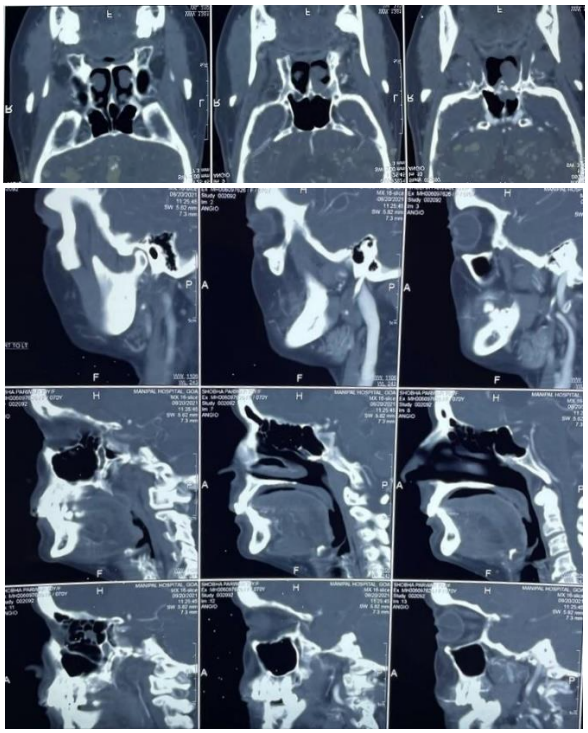

FIG. 1. **(a), (b): CT scan images of the Patient showing enhancing lobulated soft tissue mass along the floor of left sphenoid sinus measuring 2.3 cm × 1.28 cm × 1.9 cm with focal areas of erosion of sinus floor.**

At the tertiary care centre, the patient underwent removal of tumour along with adequate clear margins. Eroded floor of left sphenoid sinus was drilled out. Histopathologic examination of the excised tumour confirmed the previous biopsy diagnosis of an Infiltrative Adenoid Cystic Carcinoma. In due course, patient received four rounds of radiotherapy. After one and a half year of surgery, patient remains in remission.

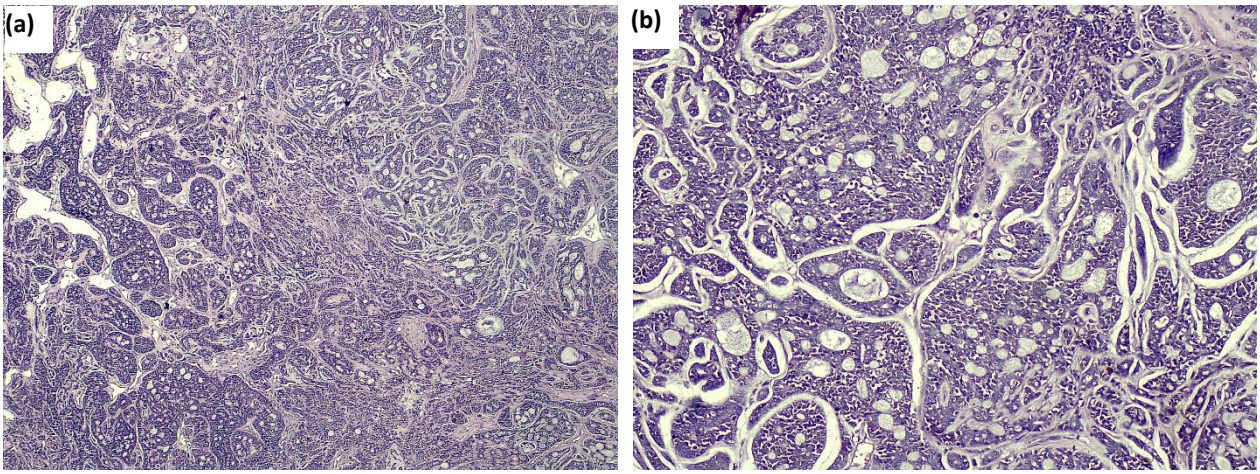

FIG. 2. **(a), (b): Histopathology images shows a tumour composed of basaloid cells arranged in cribriform and tubular pattern in a hyalinised stroma. (H&E stain x10, x40).**

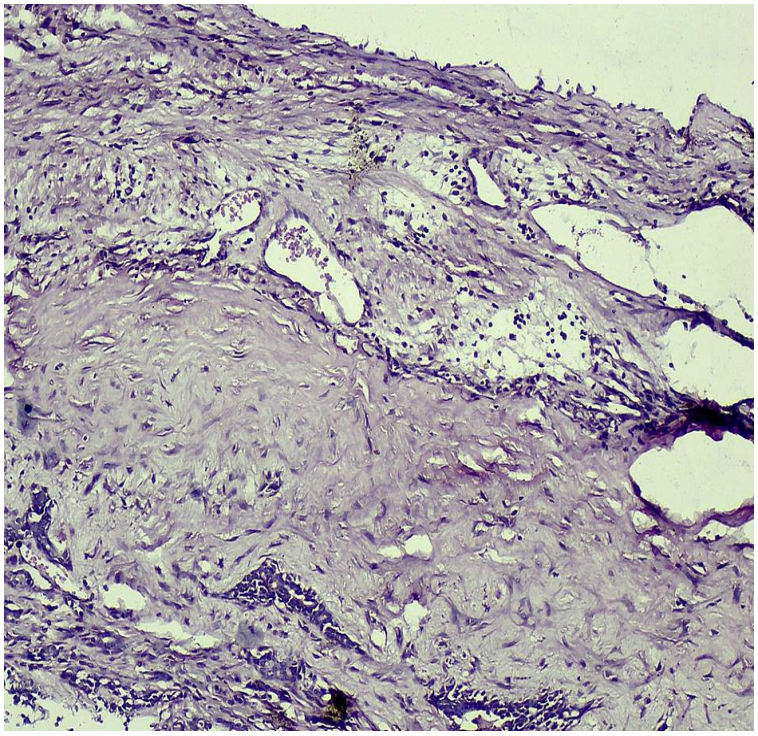

FIG. 2. **(c): Histopathology images showing tumour cells infiltrating around the nerve- Perineural invasion (PNI) (H&E stain, x40).**

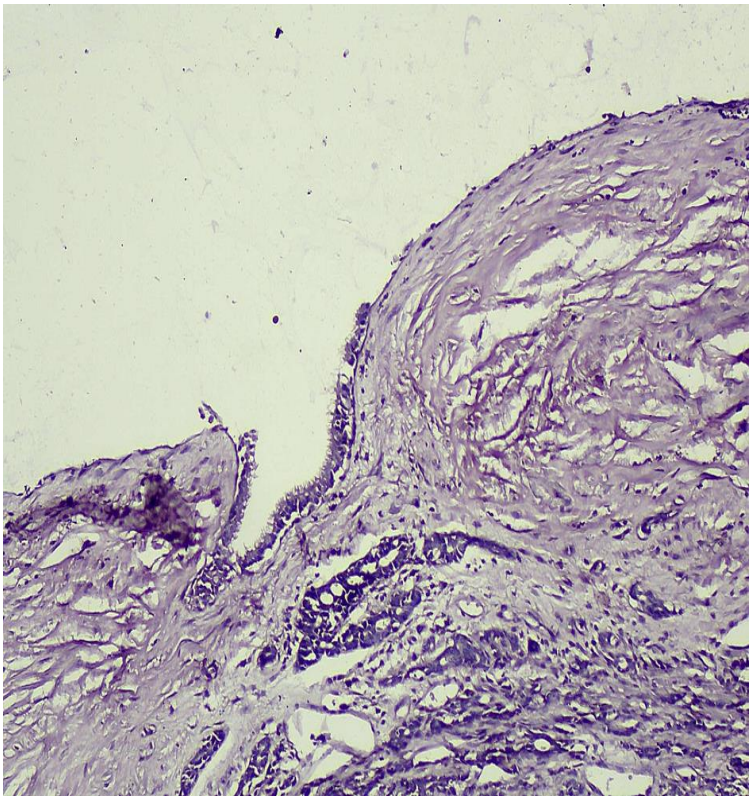

FIG. 2. **(d): Section showing ciliated columnar epithelium of the sinonasal tract in vicinity of the tumour.**

3. Discussion

ACC is a rare malignant epithelial tumour which is predominantly seen in the minor salivary glands. However, seromucinous glands underlying the respiratory epithelium give origin to a rarer form of ACC of sinonasal tract which accounts for less than 1% of Head and Neck malignancies [1,4]. Maxillary sinus is the most common site for Sinonasal ACC accounting for 46%-63% while sphenoid is seen in 3%-5% cases only [1,3]. It is usually seen in fifth to seventh decade of life without much gender predilection [1,5]. Due to its insidious onset and indolent course, most of the patients present at an advanced stage of disease with an average lag period of two years [2]. Additionally, due to close proximity to various important structures and a high tendency of perineural invasion, ACC of sinonasal tract shows a more aggressive nature than ACC originating in the salivary glands [1,4].

Clinically, ACC usually presents with non-specific symptoms such as nasal blockage, epistaxis, non-specific pain or local spread symptoms which can be auditory, visual and nerve infiltration related symptoms. This results in a lack of suspicion, thus causing a delay in investigations and hence diagnosis, as was observed with our patient [2]. Although CT scan and MRI are the mainstay of initial diagnosis, it is the histopathologic examination which is confirmatory [5]. Histologically, three main patterns are described namely tubular, cribriform and solid which are similar to the ACC seen in salivary glands [1,4,5].

On microscopy, the differential diagnosis includes other benign and malignant neoplasms of Head and Neck such as Pleomorphic Adenoma, Basal Cell Adenoma, Basaloid squamous Cell Carcinoma, Polymorphous Adenocarcinoma and

Mucoepidermoid Carcinoma. Immunohistochemistry can help to differentiate among these. Markers such as Pan CK, EMA, SMA, p63, S100 are positive in basal cells, CK7, CD117/C-kit, are positive in ductal cells while CK20 and Vimentin are negative in Sinonasal ACC [1,2,5,6]. Perineural invasion, which is characteristic of this tumour, doesn't seem to affect the overall prognosis though some authors report otherwise [3,4,6]. The treatment protocol depends upon the TNM stage, Site and Grade of tumour along with other factors such as functional status and cosmetic acceptability of the patient [7]. Surgical approach is tailored to patient needs as extensive spread and involvement of vital structures limits complete surgical excision in majority of the cases.

Hence, multimodality and multidisciplinary approach is the current paradigm [7,8]. Curative surgery combined with adjuvant radiotherapy is the preferred approach nowadays. It helps to reduce pre-op tumour burden thereby helping in achieving clear surgical margin. Adjuvant radiotherapy also reduces the incidence of recurrence contributing to better 5 yrs survival in patients [6,7]. Chemotherapy has no role in management of Sinonasal ACC because of low mitotic index of the tumour [5]. A few genetic mutations in relation to ACC have been described in recent times such as chromosomal translocation t(6;9) resulting in fusion of MYB-NFIB gene and high EGFR expression by tumour cells [3,9]. Thus far no targeted therapies based on these markers are available for clinical use. Overall, ACC of Sinonasal tract is generally associated with a poorer prognosis as compared to its salivary glands counterparts especially when there is a recurrence at the primary site [1,5,6].

4. Conclusion

Sphenoid Sinus ACC is a very rare malignant neoplasm notorious for its aggressive local behaviour, relentless progression with high chances of recurrence and late distant metastasis. Its indolent growth pattern, non-specific symptoms at presentation and a lack of suspicion due to rarity of tumour, all combined, results in delayed diagnosis which limits the management protocols available. Hence knowledge of the entity leading to a high index of suspicion can help in early diagnosis. In present times, surgical resection along with aggressive radiation therapy holds the key for better prognosis while the search for better targeted therapy continues.

5. Authors Contribution

Shergill KK conceptualised and prepared the manuscript, Pillai HJ managed the case, Raj N did scan reporting, Vatsgotra V did the final proof reading and editing.

6. Conflicts of Interest

None

7. Ethics Committee Approval

Due Approval for the study taken from the Hospital Ethics committee.

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