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Case Report

A dormant yet belligerent tumour, the sinonasal adenoid cystic carcinoma- A case report

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ABSTRACT

Introduction: Adenoid cystic carcinoma, also known as cribriform tumour or Cylindroma owing to the cellular arrangement or type of tissue that it originates from, is rare and shows perineural invasiveness as a pathognomic feature making treatment challenging. The tumour is known to arise in the salivary glands and seldom seen in the nasal cavity or paranasal sinuses. We here present a rare case of ACC occurring beyond its usual premise.

Case Report: A male patient aged 32 presented with unilateral nasal obstruction and epistaxis and on examination a growth in the right nasal cavity was noted to peculiarly arise from beneath the mucosa of the nasal floor, extending posteriorly up to the choana, not involving the turbinates or septum. DNE with biopsy was done and CECT PNS was done for further evaluation and the mass was diagnosed to be stage III sinonasal adenoid cystic carcinoma.

Discussion: This tumour originates from minor salivary glands and its presentation in the nose and paranasal sinuses has been reported to be very sparse. This is perhaps a reason for it to be missed out in daily practice due to clinical features being similar to several inflammatory and neoplastic diseases of the nose and paranasal sinuses. Histopathological cribriform and tubular subtypes are less aggressive than solid form and that determine the prognosis. It is a disease of 6th and 7th decade, slow growing and locally invasive very unusual to find it in a young patient such as ours.

Conclusion: Though uncommon the ACC mustn't go undiagnosed and must be differentiated from commoner nasal masses. Otorhinolaryngologists need to work in collaboration with pathologists and oncologists to accurately diagnose and treat this belligerent tumour in a multifaceted approach. Complete surgical excision with post-operative radiotherapy is the most accepted treatment plan.

1. Introduction

Head and neck cancers are several in number, among them the malignancies of the nose and paranasal sinuses contribute to about less than 3%.[1] The commonest malignancies of the nasal cavity and paranasal sinuses are squamous cell carcinomas, whereas esthesioneurobalstomas, adenocarcinomas and adenoid cystic carcinomas are uncommon. The factors affecting the prognosis of the ACC are their locoregional spread, perineural invasion and high rate of recurrence. Usually, these tumours affect women more than men and also occur after the age of 50.[2] Their management includes detailed CT scans along with now a days PET to look for spread and extent, followed by total resection for all tumours except those of T4 and then if needed adjuvant radiotherapy.[3] It is also said that the histological variant of the tumour determines its prognosis. Adenoid cystic carcinomas are

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known by many other terms such as “cribriform tumour”, “Cylindroma” or “Adenocystic carcinomas” owing to their cellular arrangements or type of tissue they originate from. We present a case of this rare entity and highlight the many unique features of this tumour.

2. Case Report

A 32 yr. male patient presented to us with the primary complaints of right sided nasal obstruction for 4 months, insidious in onset, gradually progressive, aggravated by cold climate and not relived on medications. It was associated with epistaxis around 4 episodes, sudden in onset and stopped spontaneously. He also gave history of generalized weakness and his appetite was normal and there was no significant weight loss. He had been smoking cigarettes daily and also consuming alcohol three to four times a week for 15 years.

Nose examination showed normal external appearance, but on anterior rhinoscopy a red sessile growth found along the floor of the right nasal extending anteriorly from the anterior end of inferior turbinate and posteriorly up to the choana. It was insensitive to touch, bleeding present on touch, the septum was deviated to the left. There was no paranasal sinus tenderness, the hard palate was normal. Contrast enhanced CT of paranasal sinuses was done (Figure 1), which revealed an enhancing soft tissue hypodense polypoidal lesion occupying the inferior aspect of the right nasal cavity causing. narrowing of the right osteomeatal complex with mass effect, there was no bony erosion or involvement was noted bilateral maxillary polyposis and mild left DNS, but there was extension into the hard palate bone which was not clinically appreciable.(Figure 2) Diagnostic nasal endoscopy done showed the reddish sessile mass extended along the floor of right nasal cavity and was probable all around except inferiorly and laterally as if extending from the mucosa of floor and lateral wall but the middle meatus was free and patent. The choanae were free of disease and nasopharynx was normal. The images of the DNE are as shown below. Multiple punch biopsies were taken from the lesion which on histopathological examination showed, features suggestive of adenoid cystic carcinoma. Under higher magnification, the typical neoplastic cells arranged in cribriform pattern composed of ductal and myoepithelial cells are seen, which is pathognomic of adenoid cystic carcinoma. (Figure 3)

Based on the TNM staging for sinonasal tumours this case was staged as Stage III- T3N0M0. The patient was planned to undergo total surgical resection with adjuvant radiation therapy keeping in mind the stage, high rate of recurrence and risk of perineural invasion.

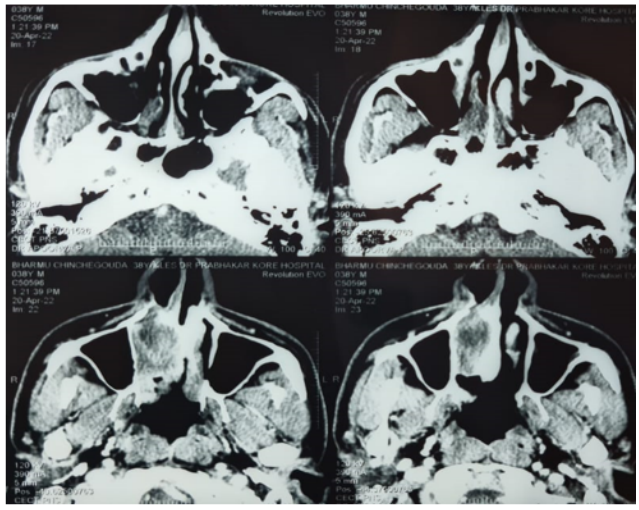

Fig. 1: CECT-PNS showing enhancing soft tissue hypodense polypoidal lesion occupying the inferior aspect of the right nasal cavity with narrowing of right osteomeatal complex

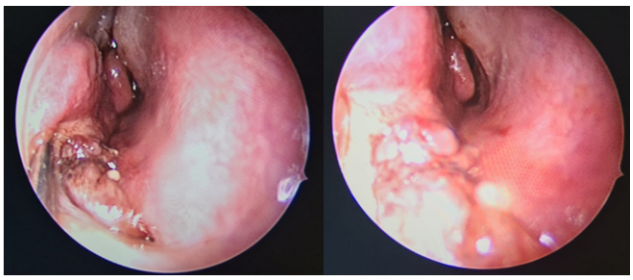

Fig. 2: Nasal Endoscopy- Reddish sessile mass extending along the floor of right nasal cavity

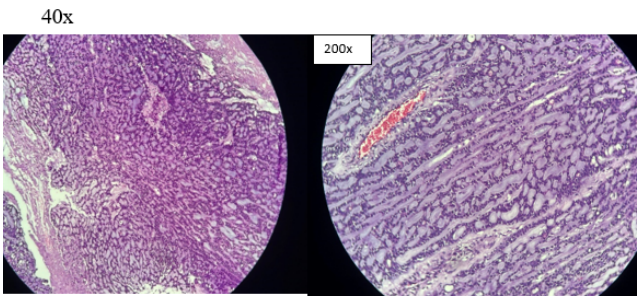

Fig. 3: Histopathology- Typical neoplastic cells arranged in cribriform pattern composed of ductal and myoepithelial cells are seen

3. Discussion

Sinonasal tumours are a small fraction of head and neck tumours. They arise from the various components of the sinonasal lining ranging from the mucosa, the Schneiderian mucosa, minor salivary glands and accordingly manifest, but many of them mimic common inflammatory or benign conditions. It is mainly seen to occur in the 5^{th} and 6^{th} decade of life and its occurrence in the 3^{rd} decade as in our case hasn't been reported as far as our knowledge. Clinically these patients present with swelling over the face, nasal obstruction, nasal discharge and epistaxis, they may have facial numbness, facial paralysis, dysphagia, hard palate erosion, swelling, headache and blurring of vision can also occur which was unlike what we saw in our case. There have been several risk factors associated with development of this tumour, ranging from smoking, inhalation of petrochemical fumes, due to wood dust or coal dust and alcohol but we see here that our patient had two of these factors probably contributing to the development of the disease.[4] The most worrisome features of ACC are its high rate of local recurrence even after as long as 10 years and its tendency of perineural invasion, making its treatment more challenging.

Histological features of ACC include a hyaline globule similar to that of a basement membrane and presence of finger like projections seen in-between cell clusters, these cells have minimal cytoplasm and their nucleus is round or oval shaped hyperchromatic.[5] The well differentiated ACC can be histologically divided as cribriform, tubular and solid based on their patterns, in these the tumour cells which are of a similar morphology form islands and they have punched out spaces, and produce a typical pattern which appears like "Swiss -cheese".[4] This is important because studies have suggested that the tubular and cribriform as seen in our case have a better prognosis as compared to the solid pattern.[6] On immune histochemistry they are seen to be positive for CD 117 the c -kit, this shows that the tumour cells have originated from ductal cells. small cell tumour, olfactory neuroblastoma, basaloid squamous cell carcinomas, all closely resemble ACC and must be ruled out with the help of IHC in cases in which the FNAC or biopsy is not conclusive, due to poorer prognosis. Recent studies in past few years have used MYB immunohistochemistry which can be used even for cytology specimens to diagnose ACC.

It becomes challenging to distinguish ACC from other basaloid carcinomas and even adenomas, hence along side histology this too must go hand in hand to ascertain a diagnosis.[7]

ACC is radio sensitive and hence commonly subjected to radiotherapy in cases of recurrence after resection. Based on the site and the involvement recent studies say we must determine pre or post operative radiotherapy, also now newer modalities such as carbon -ion radiation therapy are said to be more effective rather than older ones such as fast neutron which was previously one of the best options.[8] Morbidity following resection is high due to the tumour involving the facial bones, sinuses and hard palate, thereby affecting speech, swallowing and gross disfigurement. Chemotherapy is of only palliative value as suggested by several series.[9] These factors must be taken into account while planning treatment of a case of ACC.

In the past, the challenge was mainly the correct diagnosis after which the focus was shifted to surgery and now radiation but the most appalling thing is that ACC is known to show recurrence even after 10 years, reports showing that at 10 years and 15 years the life expectancy falls to less than 40% and thus adequate follow up is not just 10 or 15 years but lifelong, which must be considered by the clinician and also explained to the patient and attenders in the prognosis.[10]

4. Conclusion

Adenoid cystic carcinoma is a rare entity with multiple closely resembling tumours. Its high rate of recurrence and tendancy of perineural invasion make its management challenging. Rhinologists must rule out this entity as a differential while coming across sinonasal masses in daily practice even in the 3^{rd} to 4^{th} decade of life as seen in this case. The clinical diagnosis is confirmed by cytology or histopathology and the final treatment is total surgical excision with adjuvant radiation post-surgery as it increases post-operative life expectancy which is of tremendous value especially in young patients such as ours. The graveness of this rare disease and its sometimes-subtle presentation makes an expeditious diagnosis more crucial.

5. Conflict of Interests

The authors declare that they have no conflict of interests.

6. Ethical Approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

7. Consent for Participation

Informed consent was obtained from the patient and relatives of the patient.

8. Consent of Publication

Written and informed consent for publication was obtained from the patient and his relatives as per the guidelines of the institution and the journal prerequisites.

References

1. Ellington CL, Goodman M, Kono SA, Grist W, Wadsworth T, Chen AY, et al. Adenoid cystic carcinoma of the head and neck: incidence and survival trends based on 1973-2007 surveillance, epidemiology, and end results data. *Cancer*. 2012;118(18):4444–51.
2. Sharma D, Sharma N, Sharma V. Sinonasal cancers: diagnosis and management. London, UK: IntechOpen; 2019.
3. Qasim H, Vivek VK, Svider PF, Radvansky BM, Boghani Z, Liu JK, et al. Sinonasal adenoid cystic carcinoma: systematic review of survival and treatment strategies. *Otolaryngol Head Neck Surg*. 2013;148(1):29–39.
4. Amit M, Binenbaum Y, Sharma K, Ramer N, Rame I, Agbetoba A, et al. Adenoid cystic carcinoma of the nasal cavity and paranasal sinuses: a meta-analysis. *J Neurol Surg B Skull Base*. 2013;74(3):118–25.
5. Thompson LD, Penner C, Ho NJ, Foss RD, Miettinen M, Wieneke JA. Sinonasal tract and nasopharyngeal adenoid cystic carcinoma: a clinicopathologic and immunophenotypic study of 86 cases. *Head Neck Pathol*. 2014;8(1):88–109.
6. Nagao T, Gaffey TA, Serizawa H, Sugano I, Ishida Y, Yamazaki K, et al. Dedifferentiated adenoid cystic carcinoma: a clinicopathologic study of 6 cases. *Mod Pathol*. 2003;16(12):1265–72.
7. Sun T, Akalin A, Dresser K, Fischer AH, Zuo T. The Utility of MYB Immunohistochemistry (IHC) in Fine Needle Aspiration (FNA) Diagnosis of Adenoid Cystic Carcinoma (AdCC). *Head Neck Pathol*. 2021;15(2):389–94.
8. Cantù G. Adenoid cystic carcinoma. An indolent but aggressive tumour. Part B: treatment and prognosis. *Acta Otorhinolaryngol Ital*. 2021;41(4):296–307.
9. Alabi BS, Afolabi OA, Omokanye HK, Dunmade AD, Ayodele SO. Clinical presentation and outcome of sinonasal tumors in a Nigerian Tertiary Hospital–6-year review. *Niger Med J*. 2017;58(3):92–5.
10. Nicolai P, Bradley PJ. Anterior Skull Base Tumors. *Adv Otorhinolaryngol Basel Karger*. 2020;84:197–209.

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