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Review Article

Adenoid cystic carcinoma of nasal septum: A review of literature

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ABSTRACT

Adenoid cystic carcinoma (ACC) is a common salivary gland tumour which constitutes about 10% of all salivary gland malignancies and 1% of all head & neck malignancies.¹ It is a slow growing tumour with propensity for perineural invasion.^{2,3} It occurs most commonly in minor salivary glands, most commonly in oral cavity. It is the third most common malignancy of paranasal sinuses with most common site being maxillary sinus followed by nasal cavity, nasopharynx and ethmoid sinus.⁴ It is most commonly seen in 4th and 5th decade with female predominance.⁵ ACC arising from nasal septum is very rare with only 10 cases published in literature. In this paper we present a case of nasal septal adenoid cystic carcinoma with review of the literature.

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1. Case Report

A 43-year-old male patient came to ENT OPD with complaints of left nasal obstruction and bleeding from nose since 6 months. Nasal obstruction was insidious in onset and gradually progressive and commencing to complete left nasal obstruction since 2 months. Bleeding from nose was scanty in amount with 1-2 episodes per day. There were no complaints of recurrent cold, excessive sneezing, headache, facial fullness. Patient was a chronic smoker with 20 pack years, non alcoholic and vegetarian. On anterior rhinoscopy, a mass was seen in left nasal cavity which was sensitive to touch. Probe couldnot be passed on medial side. On nasal endoscopy, mass of size 3*2 cm was seen arising from nasal septum from mid of cartilaginous septum to mid of bony septum (Figure 1). On noncontrast computed tomography of paranasal sinuses soft tissue attenuation lesion in noted obstructing left nasal cavity and lesion in contact with

partially visualised left inferior turbinate. Bony left inferior turbinate is not visualised, lesion is causing remodelling of medial wall of left maxillary sinus (Figure 2). On contrast enhanced MRI of nose and paranasal sinuses, there was well defined soft tissue lesion showing moderate heterogenous enhancement in left nasal cavity involving nasal septum, causing obstruction of left nasal cavity, abutting left inferior turbinate with obstruction of left maxillary sinus causing sinusitis, suggestive of neoplastic lesion (Figure 3).

Endoscopic nasal biopsy was taken under local anaesthesia which on histopathological examination showed luminal and adluminal proliferation of cells with scant cytoplasm forming pseudoglandular spaces with basement membrane material and mucin suggestive of adenoid cystic carcinoma. On immunohistochemistry, tumour cells were positive for CK-7, S-100 and CD-117 and negative for CK-20 and CEA.

Surgical excision was planned. By lateral rhinotomy incision, nasal septum was removed after taking 1 cm

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margins. Post operative radiotherapy was given to patient in 33 cycles over 6.5 weeks with 2Gy per cycle. After 2 years of follow up, there was no recurrence.

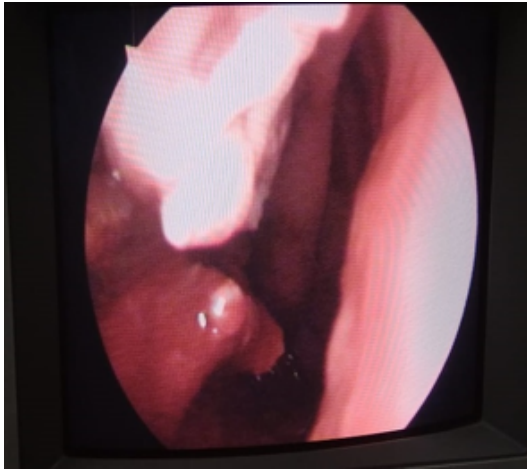


Fig. 1:

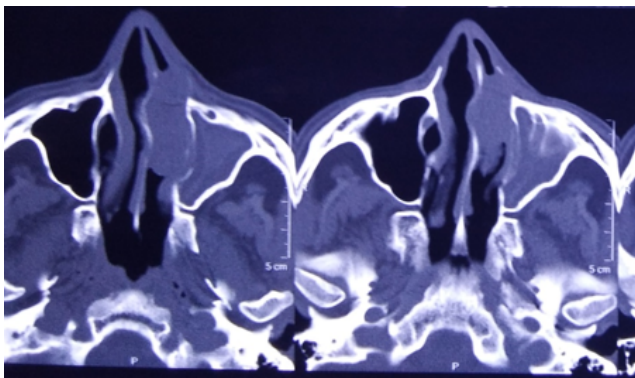


Fig. 2:

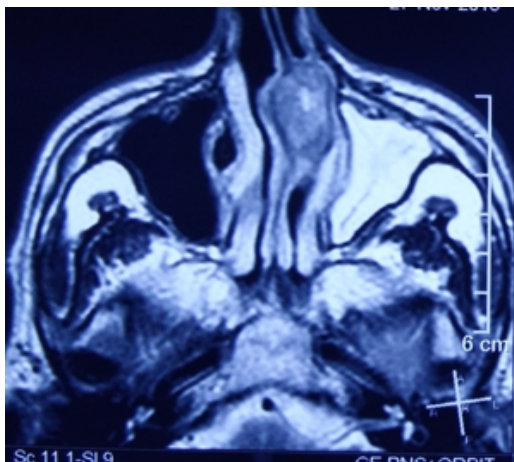


Fig. 3:

2. Discussion

Nasal septal malignancies constitutes 2.4%-8.7% of all sinonasal malignancies. Adenoid cystic carcinoma is a common sinonasal malignancy but is very rare in nasal septum.⁶ It presents with non specific symptoms such as nasal obstruction and epistaxis. It is a slow growing tumour with invasion of perineurallymphatics. It is known to produce blood borne metastasis most commonly to lung. Metastasis to regional lymphatics is very rare.² 43 cases of septal tumours were described by young, out of which only 1 was ACC.⁷ Another study by Beatte et. Al. stated only one case of setal ACC out of total 85 cases.⁸

It is histologically of 3 types cribriform, tubular and solid. Cribriform is the most common type with best prognosis while solid is least common with worst prognosis. There is high incidence of recurrence and distant metastasis with solid type.⁹ Solid type ACC is further subdivided into 3 types by szantoe. al as grade 1 with 0 % solid component, garde 2 with less than 30% solid component and garde 3 with solid component more than 30%. 5 year survival rate was 95%, 65% & 14% respectively and 10 year survival rate as 76%,26% an d 5% respectively.¹⁰ On immunohistochemistry these stains positive for smooth muscle actin, S100,vimentin, smooth muscle myosin heavy chain, CD117 and myeloblastosis oncogene.¹¹

Its treatment is generally combined surgery and radiotherapy.¹² Surgical approach depend on the tumour size and location. Small and localised tumours can be excised via endoscopic approach while larger tumours requires lateral rhinotomy approach. Mid facial degloving and lateral rhinotomy with sublabialincisions may be required for lower septal and posterior septal tumours.¹³ Post op radiotherapy is given with a total dose of 66-70 Gy in 2Gy per fraction with five fractions per week. In a study by Wiseman et. al , it was seen that local recurrence rate was lower in group of surgery with post op radiotherapy than surgery alone.¹² In a study by Horiuchi et. al, he stated that minimum 50 Gy radiation must be given post operatively to effectively reduce local recurrence.¹⁴ Chemotherapy is ineffective in its treatment. Radiotherapy doesn't have any effect on cure but it reduces its recurrence by treating residual microscopic disease.¹⁵ Long term follow up is required for its slow growth and high local recurrence and distant metastais.

3. Review of Literature

On reviewing literature on ACC of nasal septum, we could find only 10 cases out of which 4 were females and 6 were males. Average age of presentation was 53.7 years which is 56.5 years in males and 49.5 years in females. Thus, female have an early age of presentation of this tumour. In reported cases from india, out of four, three are males showing male predominance with average age of presentation 56.75 years

Table 1:

Author	Age	Country	Symptoms			Treatment	Recurrence
			Obstruction	bleeding	others		
Tai et. Al6 ,2007	56yr\male	Taiwan	Yes	Yes		Surgery by lateral rhinotomy Radiotherapy	1.5 yrs follow up, no recurrence
Akiyama et. A115 ,2013	42yr\female	Japan	Yes	No	Dysosmia	Anterior canialfossa surgery *ensoscopic nasal surgery*transpalatal approach radiotherapy	9 months follow up, no recurrence
Lit Yee et. A116, 2018	54yr\male	Malaysia	Yes	Yes		Endoscopic excision with radiotherpay	15 months follow up , lung metastasis were found
Handa et. A117, 1992	50yr\female	Japan	Yes	No	Nasal pain	Lateral rhinotomy No radiotherapy	2 yrs follow up, no recurrence
Schneiderman et. A118 ,2002	66yr\male	USA	Yes	No		Intranasal resection No radiotherapy	9 months
Sivaji et. A119 ,2003	64yr\female	India	Yes	Yes		Lateral rhinotomy with radiotherapy	Presented in very few days of follow up after treatment
Konadir A &Hassoumi K20, 2018	42yr\male	Morocco	Yes	Yes		Endoscopic resection with post op radiotherapy	1yr and 9 months, no recurrence
Beladavar B, Batra R5 ,2018	60 yr\male	India	Yes	Yes		No treatment explained	
Priya SR, Chaukar D, D'Cruz A,2016	50yr\male	India	Yes	Yes		Excision by lateral rhinotomy	15 months, no recurrence
Priya SR, Chaukar D, D'Cruz A,2016	53yr\male	India	Yes	Yes		Excision was done at some other center 3 years ago ,technique not explained	3 yrs of follow up, no recurrence

and of males 53.3 years and only female was of 64 years of age. Treatment was given in 9 patients. Maximum duration of follow up was of 3 years by Priya SR and minimum duration of follow up was just after the treatment with an average of 16.33 months. Recurrence was seen in only one report after a follow up of 15 months to lung.

4. Conclusion

ACC of nasal septum is a very rare malignancy. ACC of nasal septum has early presentation because of early nasal obstruction and early nasal bleeding, in comparison to ACC of maxillary sinus which appears usually in t3/t4 stage after bony erosions and rhinological, ophthalmic and palatal symptoms.

5. Source of Funding

None.

6. Conflict of Interest

None.

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