



ADENOID CYSTIC CARCINOMA NASAL CAVITY- A RARE CASE REPORT UDAYA CHANDRIKA G

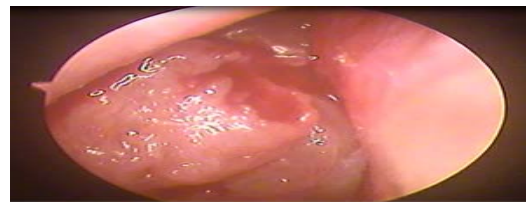
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Abstract : ABSTRACT INTRODUCTION Adenoid cystic carcinoma is a rare malignant tumor of nasal cavity constituting about 3-4 percent of all sinonasal malignancies. These tumors present with nasal obstruction and epistaxis. They have an indolent course but have high tendency for local recurrence along with perineural and haematogenous spread. Histologically they present with tubular, cribriform and solid patterns, of which solid form is associated with much aggressiveness. Lung metastasis is more common with these tumors. Local recurrence is common in adenoid cystic carcinomas of nasal cavity. CASE REPORT A 28 year old female presented with nasal obstruction , headache for the past one year with occasional nasal bleed . Anterior rhinoscopy revealed a fleshy, reddish , lobular mass occupying right nasal cavity till the anterior end of inferior turbinate. Computed Tomography of Paranasal sinuses was done, which showed homogenous soft tissue mass in the inferior turbinate of right nasal cavity. Clinical diagnosis of benign tumor of the nasal cavity, Papilloma, was made . Endoscopic assisted removal was done and suprisingly histopathologically Adenoid cystic carcinoma with cribriform pattern was reported. MRI was done to rule out perineural spread. Medial maxillectomy was done through lateral rhinotomy approach followed by post operative radiotherapy. CONCLUSION Adenoid cystic carcinoma constitute 4 percent of all sinonasal malignancies This case is presented for its rarity. This is the first reported case in our institution in the past 10 years based on a recent clinicopathological study conducted on Head and Neck Malignancies.

Keyword : sinonasal malignancy, indolent course, recurrence, lung metastasis.

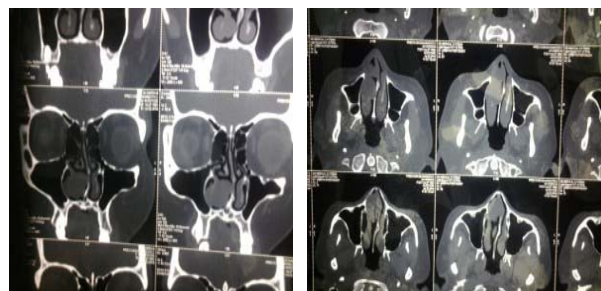
CASE REPORT:

A 28 year old female patient had nasal obstruction with headache for the past one year with occasional nasal bleed. No exposure to chemicals or wood dust. No eye or dental symptoms.

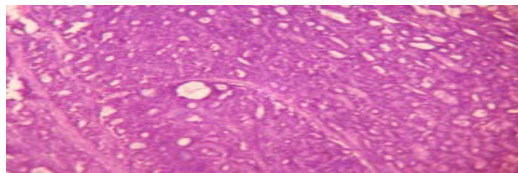


PREOPERATIVE PICTURE

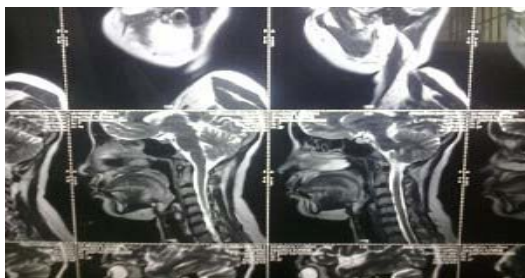
Anterior rhinoscopy showed an reddish, fleshy, lobular mass seen obliterating the right nasal cavity abutting on to the septum. On Probe test, the mass was found to be attached to the lateral wall of the right nasal cavity (anterior end of inferior turbinate). right nasal vestibule was free all around. The mass did not bleed on touch, but it was painful to touch. Gingivolabial sulcus was not obliterated. Posterior rhinoscopy was normal. No abnormality was detected in throat examination. Cervical lymphnodes were not enlarged or palpable on both sides. No restriction of eye movements and vision was normal. Systemic examination did not reveal any abnormality. CT PNS was done: well defined homogenously enhancing soft tissue mass measuring 2.7x 2.3mm in the anterior end of inferior turbinate of right nasal cavity. Nasal septum, other areas of lateral wall of nose, maxillary sinus appear normal.



A Diagnosis of Benign Tumor of Nasal cavity – Papilloma was made following which Endoscopic assisted removal of the nasal mass was done. Surprisingly the Histopathological report revealed Adenoid Cystic Carcinoma - cribriform pattern.



MRI was done after endoscopic assisted removal of the mass, since Adenoid cystic carcinomas have greater propensity for perineural spread. There was no perineural spread made out.



Medial Maxillectomy through lateral rhinotomy was performed : Incision started just above the level of medial canthus, passes along lateral border of nose to the upper edge of alar margin, preserving the lower alar rim. Upper lateral cartilage freed from nasal bones at the pyriform opening and soft tissue flap elevated from frontal wall of maxilla and nasal bones. Orbital periosteum elevated , lacrimal sac exposed by nibbling away the anterior lacrimal crest. osteotomies first made through anterior wall of maxilla, just medial to inferior orbital foramen, another into lower border of lateral nasal wall in the inferior meatus, floor of orbit, finally through frontal process of maxilla and nasal bone down to pyriform aperture, whole block freed from posterior attachment just in front of sphenopalatine foramen.



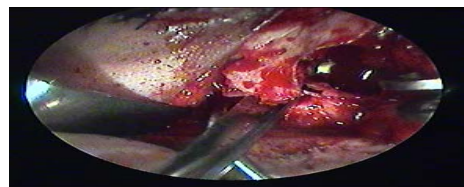
Incision made from medial canthus through lateral border of nose



Soft tissue retracted away



Osteotomies done



Medial maxillectomy done

Specimen sent for HPE which showed Marginal Clearance .Patient was referred to radio- oncology where she was given 60 Gy of irradiation .

Discussion:

Adenoid cystic carcinoma (ACC) was first described by three Frenchmen (Robin, Lorain, and Laboulbène) in two articles published in 1853 and 1854. It was they who described the cylindrical appearance of this tumor. Many articles since then have remarked that Billroth, in 1859, first described Adenoid cystic carcinoma under the name cylindroma, but it was he who described that the tumor had a "great tendency to recur."

Clinical behaviour :Tumor growth is slow, but its clinical course is relentless and progressive. Operative intervention is usually feasible, but multiple local recurrences are the rule. Metastatic spread to regional lymph nodes is uncommon, but distant spread to the lungs and bones is frequent.5-year survival rates are optimistically high, but 10- to 20 year survival rates are dismally low. It has been reported that Adenoid cystic carcinoma of the nasal cavity and paranasal sinuses has a worse prognosis than in any other area of the head and neck region

Histopathology :Adenoid cystic carcinoma can be categorized into three growth patterns: cribriform, tubular, and solid. In most studies, a solid growth pattern is associated with a worse prognosis, caused by advanced stage and development of distant metastases . A unique feature of Adenoid cystic carcinoma is the propensity for perineural invasion, even with early-stage. The cribriform variant demonstrated a significantly worse prognosis in terms of local recurrence rate. The choice of therapy is affected by site, stage, histologic grade, and biologic behavior of the Adenoid cystic carcinoma. Most tumors are treated surgically, with the possible addition of adjunctive radiotherapy. Pathologic findings correlated with local recurrence rates, and positive resection margins were significantly associated with an increased risk of local recurrence. In patients who receive postoperative radiation therapy, an improved outcome is observed with radical surgery compared with biopsy alone. A good response is usually seen initially with photon irradiation therapy alone and most tumors will recur locally with time. Radiation, usually in doses of 60 Gy or more, may be of benefit when there is minimal residual microscopic disease. It is likely that there is frequently unrecognized perineural invasion in specimens with "negative" margins that would have benefited from the addition of radiation therapy. Chemotherapy currently is seeking a role in the management of advanced and metastatic tumors. Adenoid cystic carcinoma arising from sites in proximity to the cranial base (nasopharynx, nasal cavity, and maxilla) have a significantly increased risk of local recurrence. This is related to the difficulty of securing clear resection margins at the cranial base because of technical difficulties associated with the surgery, intracranial extension of the tumor along nerves, and restrictions on the limits of resection caused by the proximity of critical neural and vascular structures. The use of the gamma knife has been

recommended for use in the treatment of recurrent tumors involving the skull base, using a median radiosurgery dose of 15 Gy. Incidence of other sites being involved by distant metastasis is likely to be more common, because once lung metastases are detected, no further metastatic investigations are performed. Median survival times after appearance of distant metastases among patients with isolated lung metastases and those with bone metastases with or without lung involvement were 54 and 21 months respectively. Despite local aggressive therapy, the majority of patients (60%) will develop recurrent disease. Approximately 50% of recurrences are clinically evident within 2 years after surgery and radiotherapy. The routine use of radiologic examinations (especially MRI) during the postoperative period may identify changes indicative of recurrent disease months to years before it is clinically evident.

Conclusion: Adenoid cystic carcinoma being a rare malignant tumor in nasal cavity is unique in clinical behaviour, histological presentation, with characteristic perineural spread and chances of recurrence and metastases. Treatment consists of surgery along with radiotherapy. This case is presented for its rarity being first reported one in our institution in the past 10 years.

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