Case Report

Adenoid Cystic Carcinoma Nose Masquerading as Basal Cell Adenoma

Authors:
Anjali Singh, Senior Resident,
Saurabh Varshney, Professor & Head,
Amit Kumar Tyagi, Assistant Professor,
Amit Kumar, Assistant Professor,
Tushar Kalonia, Senior Resident,
Departments of Otorhinolaryngology (ENT) and Pathology, All India Institute of Medical Sciences, Rishikesh 249203, Uttarakhand.

Address for Correspondence
Dr. Saurabh Varshney,
Professor & Head,
Departments of Otorhinolaryngology (ENT) & Head Neck Surgery,
All India Institute of Medical Sciences,
Rishikesh - 249203, Uttarakhand.
E-mail: drsaurabh68@gmail.com.

Citation

Submitted: May 14, 2020; Accepted: Aug 12, 2020; Published: Sep 20, 2020

Abstract: Basal cell adenoma is a benign basaloid cell tumor involving salivary glands. It requires biopsy and histopathology to distinguish from pleomorphic adenoma, adenoid cystic carcinoma and basal cell adenocarcinoma. Present manuscript reports a case of an adult female with nasal mass, which was clinically and cytologically mimicking benign lesion and adenoid cystic carcinoma was confirmed only on surgical biopsy. The possible clinical dilemma between Basal cell adenoma and Adenoid cystic carcinoma is discussed in this case report.

Key Words: Adenoid cystic carcinoma, Basal cell adenoma, Basal cell adenocarcinoma

Introduction:
Salivary gland tumors are rare tumors and include 3% of all head and neck tumors. Adenoid cystic carcinoma is a malignant tumor, commonly involving minor salivary glands of oral cavity. It resembles basal cell adenoma on cytology due to similar myoepithelial differentiation. Biopsy is most accurate for diagnosis (1). Definitive treatment modality is surgical excision. Radiotherapy has limited role, it can be used for positive margins after surgery and as an adjuvant therapy or for palliation for advance stage tumor (2).

Case Report
A 38 years female presented with swelling over nose for 1 year, which was gradually progressive and not associated with pain, epistaxis, nasal blockage, anosmia or hyposmia (Fig 1). On examination, a 2x2 cm firm swelling was noted over dorsum of nose, extending approximately 1 cm inferior to nasion and 1 cm above the tip of nose, it was round in shape, had smooth surface, with well-defined margins, non-fulcruant, non-compressible, non-transilluminant, non-tender and overlying skin was normal in texture, colour and was pinchable. Olfaction was normal.
On diagnostic nasal endoscopy, a pinkish mass was noted in roof of right nasal cavity, which was not bleeding on touch (Fig 2). There were no palpable cervical lymph nodes. Rest of ENT and Head and neck examination was normal. Contrast enhanced MRI showed a 22X15 mm well-defined round to lobulated heterogeneously enhancing mass lesion which was hypo to isointense on T1W and hyperintense on T2W, involving nasion, bilateral ala, nasal septum of upper nose and extending posteriorly into bilateral ethmoid sinuses (Fig 3). X ray chest was normal.

Fine needle aspiration cytology showed possibility of a) basal cell adenoma, b) pleomorphic adenoma or c) adenoid cystic carcinoma (Fig 8). Considering it to be a benign lesion, patient was counselled and planned for surgical excision. All blood investigations and pre-anesthetic clearance were done and patient was planned for surgical excision.

Under General anesthesia, bilateral lateral rhinotomy with extended Lynch Howarth incision was placed and skin flap was raised (Fig 4). Tumor was infiltrating inferior part of bilateral nasal bone, adjacent part of upper lateral cartilage, nasal septum and right ethmoid sinus. It was excised with adequate margins (Fig 5). Tumor infiltrating right ethmoid sinus was removed and sinus was curetted, small portion (1X1 mm) of dura was found exposed in the same region but no breech or CSF leak was noted. Reconstruction was done with rib cartilage, which was partially split to recreate the dorsum of nose and skin flap was sutured back (Fig 6).

Figure 2: Direct nasal endoscopy showing pinkish mass involving roof of right nasal cavity

Figure 3: CEMRI (sagittal view) showing a well-defined heterogeneously enhancing mass lesion hypo to isointense on TW1 (A, top), hyperintense on T2W (B, below), involving nasion, bilateral ala and nasal septum of upper nose, extending posteriorly into bilateral ethmoid sinuses

Figure 4: Intra-operative pictures (A, left) showing well defined, smooth tumor, free from overlying skin; (B, middle) Bilateral lateral rhinotomy with extended Lynch Howarth incision; (C, right) showing post-excision defect

Figure 5: Excised specimen with tumor involving dorsum of nose, upper lateral cartilage and upper part of nasal septum with surrounding margins.

Figure 6 (A, top) and (B): 6th Rib cartilage harvested to recreate the nasal dorsum and ala.

Figure 7 (A, top) and (B): showing clinical picture and direct nasal endoscopy on post-op day 21 with accepted graft and no residual disease.
Pleomorphic adenoma is a benign neoplasm composed of basaloid cells, organized with pseudo cysts and acellular basement-like material. It presents as a slow growing, mobile, painless, firm mass. It constitutes a mixture of ductal and myoepithelial elements (6).

Adenoid cystic carcinoma is a malignant tumor, commonly involving minor salivary glands of oral cavity. It involves sinonasal tract less commonly and rarely involves parotid gland. Adenoid cystic carcinoma constitutes 3-5% of all head and neck malignancies (7). Adenoid cystic carcinoma of sinonasal tract represents 10 to 25% of all head and neck adenoid cystic carcinomas and commonly involves elderly population (6th decade) (8). It is associated with occupational exposure to heavy metals like arsenic, nickel or exposure to wood dust. It arises from respiratory epithelium, i.e., pseudostratified columnar epithelium. Its origin is commonly from maxillary sinus followed by nasal cavity and has peculiar presentation with nasal mass, epistaxis or facial pain (9). It is a slow growing tumor, generally painful and do not bleed on touch. It spreads via perineural and hematogenous route. Local recurrence is quite common due to perineural spread along neural canal and haversian canal of bone. Distant metastasis occurs via hematogenous spread. Commonest site for distant metastasis are lungs, followed by bone, brain and liver. Recurrence and distant metastasis are associated with poor prognosis and poor quality of life, but it does not affect the longevity of life. Perineural spread and ethmoid or sphenoid sinus as site of origin has been found as poor prognostic factor in some studies (7). Lymph node metastasis is rare and associated with poor survival (1).

It is poorly encapsulated and infiltrative lesion. Histologically, it contains small deep basophilic nuclei with characteristic gland-like structure. It is classified into 4 histological types cribriform, basaloid or solid, cylindromatous and tubular. Among these, cribriform type is the most common growth pattern, while basaloid type has highest recurrence rate and is associated with poorer prognosis. Survival is associated with histological type and grade of tumor (10).

Management includes surgical excision with adequate margins, followed by radiotherapy. Complete surgical clearance with clear margins is treatment modality and
associated with best survival (2). Radiotherapy has no definitive role but can be used as adjuvant treatment for advanced stage, positive margins after surgical excision or for palliation. Chemotherapy can be considered for recurrence or distant metastasis (4). Challenges associated with treatment of adenoid cystic carcinoma of sinonasal tract include presentation at late stage, proximity with vital structures like orbit, dura, brain and cranial nerves, more chances of local recurrence due to bone destruction and perineural and perivascular spread. Extensive bony invasion can be present in absence of any significant radiological findings. Long-term follow-up is needed due to high recurrence rate and risk of distant metastasis (9). These tumours are related with a high risk of recurrences and significant morbidities from surgical management and adjuvant radiotherapy. Despite the aggressive management for these tumours, many patients succumb to distant metastasis, making overall prognosis of these tumours poor. (12) Approximately 10% of tumors that arise in the sinonasal tract originate in the ethmoid and/or frontal sinuses and are likely to involve the anterior cranial base. Most of the tumors involving the anterior cranial fossa arise from the nose, sinuses and orbits. Conservative surgery in these areas is fraught with local recurrence at the skull base. An anterior craniofacial resection should be performed in cases of nose and paranasal sinus tumors involving the cribriform plate with or without invasion of anterior cranial fossa (13).

Basal cell adenocarcinoma is a low-grade malignancy. It constitutes less than 2% of all salivary gland tumors. It commonly involves parotid gland (more than 90% cases) while minor salivary glands are rarely involved. Exact histogenesis is still not known. Most is the not known to be de-novo lesion but some studies believe it to be malignant transformation of basal cell adenoma. It rarely shows nodal or distant metastasis but has high chances of recurrence (25 to 30%). It generally presents as a slow-growing, asymptomatic swelling, which shows similar macroscopic and cytologic features as that of basal cell adenoma. It is necessary to distinguish true invasion or infiltration into adjacent structures from multinodularity of tumor and multifocal origin from adjacent salivary gland nodules. It shows positive staining for CK7 while negative staining for CK20 and moderate to strong positivity for S100 for myoepithelial cells. Surgical excision with adequate margins is the treatment of choice while radiotherapy can be an option for basal cell adenocarcinoma of minor salivary gland due to high risk of neurovascular invasion or diffusely infiltrative lesion. It should be differentiated from Adenoid cystic carcinoma for prognostic values and potential difference in treatment. It lacks cribriform pattern and pseudocysts of amorphous, basophilic material, high mitotic index and necrosis, which are features of adenoid cystic carcinoma (11).

**Conclusion**

Adenoid cystic carcinoma (ACC), a rare tumor of epithelial cell origin, commonly arises from the major salivary glands. Uncommonly it may be found outside the salivary glands and it's especially rare in the nasal cavity. (14) In nose adenoid cystic carcinoma commonly arises from minor salivary glands and it may mimic basal cell adenoma or basal cell adenocarcinoma. It is difficult to differentiate them cytologically. Histopathology is most accurate to confirm the diagnosis. Radiotherapy do not have definite role but can be used as an adjuvant therapy for advanced stage, positive margins after surgical excision or for palliation. Hence even a case of seemingly benign lesion clinically and a diagnosis of basal cell adenoma on cytology, possibility of Adenoid cystic carcinoma should be kept in differential diagnosis while managing such cases.

**References**