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CASE REPORT

# Dedifferentiated Variant of Adenoid Cystic Carcinoma: A Clinicopathologic Report of Two Cases

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## ABSTRACT

Dedifferentiated form of adenoid cystic carcinomas is the rare variant and is recently recognized entity with only few cases reported in the literature. We examined two cases of adenoid cystic carcinoma arising from nose with the analysis of there clinicopathologic profiles, their management, follow-up and our experience with this uncommon disease.


**Keywords:** Adenoid cystic carcinoma, Dedifferentiation, Nose.

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## INTRODUCTION

'Dedifferentiation' has become a well-known phenomenon in bone and soft-tissue tumor pathology,[1] after the first report of a 'dedifferentiated' salivary gland acinic cell carcinoma (AcCC) in 1988,[2] several investigators described this phenomenon not only in AcCC, but also in other salivary carcinomas, such as adenoid cystic carcinoma (AdCC).[3] 'Dedifferentiation' is defined as the abrupt transformation of a well-differentiated tumor into high-grade (HG) morphology that lacks the original distinct histologic characteristics. In the literature 'dedifferentiated' salivary gland carcinomas have sometimes been confused with hybrid carcinomas, which are composed of two distinct tumors entities,[4] but they should be distinguished from each other from a clinicopathological point of view.

The incidence of malignant tumors from the nasal septum is quiet rare ranging from 2.7 to 8.4% of nasal and paranasal malignant tumors.[5] Here, reported are the unusual cases of the dedifferentiated type of AdCC of the nasal septum including there clinical features, pathologic features, management and there follow-up.

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## CASE REPORTS

### Case 1

A 69-year-old patient came with chief complaints of nasal obstruction with growth in the nose for past 9 months with occasional blood stained nasal discharge. On anterior rhinoscopy, there was a lobulated, nonulcerative mass arising from the cartilaginous nasal septum extending onto the columella caudally and the ala laterally, measuring about 3 × 1.5 cm. Punch biopsy was taken which showed AdCC of nasal septum. Contrast-enhanced computed tomography (CECT) scan peripheral nervous system (PNS) was done showing that the tumor was arising from cartilaginous septum extending to columella and nasal tip anteriorly. Bony septum and paranasal sinuses were clear. Ultrasonography (USG) neck showed no neck nodes. X-ray chest was normal. Patient was then prepared for surgery and the tumor was excised with wide excision followed by second stage reconstruction using illiac bone graft, buccal mucosal graft and forehead flap.

However, patient reported with a complaint of a mass in the neck region on a follow-up visit after 2 months. On investigating, it was diagnosed as the dedifferentiated AdCC. Patient then underwent radical neck dissection with postoperative radiotherapy.

### Case 2

A 58-year-old patient came with chief complaints of growth in the nose from past 7 months with nasal obstruction. On anterior rhinoscopy, there was mass arising from the cartilaginous nasal septum, measuring about 1 × 1.5 cm. Punch biopsy was taken which showed dedifferentiated variant of AdCC of nasal septum (Fig. 1). Contrast-enhanced computed tomography scan PNS was done showing that the tumor was arising from cartilaginous septum. Columella, bony septum and paranasal sinuses were clear. Ultrasonography neck showed no neck nodes. X-ray chest was normal. Patient was then prepared for surgery and the tumor was excised with wide excision followed by second stage reconstruction using illiac bone graft, buccal mucosal graft and forehead flap followed by radiotherapy.

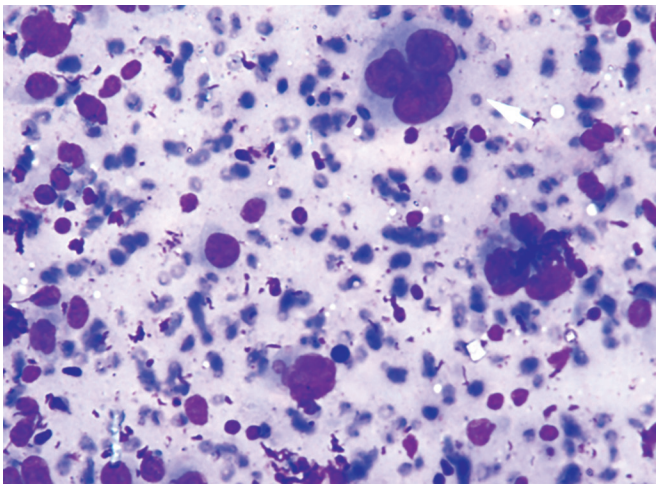

**Fig. 1:** Histopathological image

## DISCUSSION

Adenoid cystic carcinomas are common malignancies arising in major and minor salivary glands (most common in oral cavity), including the seromucinous glands of the upper respiratory tract, rarely seen within the nasal cavity where the lateral nasal wall is the most common site and the lacrimal glands. The peak incidence is found to be in the fourth to sixth decades and slight predominance in females. Adenoid cystic carcinomas are characterized clinically by an indolent course, a relatively high rate of local recurrence and late onset of distant metastases.[6]

Three histological patterns of growth have been described: tubulo ductal, cribriform and solid. Cribriform is the most common type and it also has the best outcome. Solid variant is the least common type and also has the worst prognosis.[7] Patients with nasal cavity disease has better prognosis as compared to other sites in paranasal sinuses.[8] Prognostic factors are size of tumor, margins of resection, lymph node metastasis and perineural invasion.

Since the first documentation of 'dedifferentiated' AdCC presented by Cheuk et al, a total of approximately 40 cases with 'dedifferentiated' or high-grade transformation (HGT) AdCC have been recognized in the literature to date.[3]

'Dedifferentiation' is defined as the abrupt transformation of a low-grade, well-differentiated tumor into a tumor of high-grade morphology. It is a well-recognized phenomenon in bone and soft tissue tumor pathology.[1] Dedifferentiated tumors generally are associated with a poor prognosis. Although the concept of dedifferentiation appears to be established for salivary gland neoplasms,[9] this phenomenon is uncommon in AdCC.

High-grade transformation tumors are composed of conventional carcinomas juxtapozed with areas of HG morphology, usually either poorly differentiated adenocarcinoma or 'undifferentiated' carcinoma, in which the original line of differentiation is no longer evident. The HG component is generally composed of solid nests, sometimes occurring in cribriform pattern of anaplastic cells with large vesicular pleomorphic nuclei, prominent nucleoli and abundant cytoplasm. Frequent mitoses and extensive necrosis is evident. The Ki-67 labeling index is consistently higher in the HG component. p53 abnormalities have been demonstrated in the transformed component in a few examples, but the frequency varies by the histologic type. HER-2/neu over expression and/or gene amplification is considerably exceptional.

Dedifferentiated AdCC contains two histologic components: an area of conventional AdCC of any grade and an area of high-grade undifferentiated carcinoma or poorly differentiated adenocarcinoma. The high-grade areas do not display any histologic features of AdCC and most importantly display loss of ductal myoepithelial differentiation with increased mitotic activity, comedonecrosis, micropapillary and squamoid growth patterns.

The data indicate that dedifferentiated AdCC is a highly aggressive tumor with a strong tendency to recur and metastasize, similar to that observed in AdCC with a predominantly solid growth pattern.[10]

The development and progression of malignant tumors is regulated by the expression and genetic and/or epigenetic alterations of various oncogenes and tumor suppressor genes. Although the data are limited, involvement of one or several genes has been documented in the HGT process.[11]

While the presence of a transitional zone between conventional and HG carcinoma components suggests an identical origin and is considered to indicate progression of malignancy, it remains unsettled whether the process of HGT represents a failure of differentiation in stem cells or whether differentiated neoplastic cells undergo 'dedifferentiation'.[11]

Before the diagnosis of dedifferentiated AdCC is made, other types of salivary gland neoplasms with morphologically high-grade features that mimic low-grade AdCC should be excluded. These include the solid type of AdCC,[10] collision tumors, and hybrid carcinomas.[12] Even in the solid type, the tumor cells of AdCC exhibit the original line of differentiation.

Among various treatment modalities, combined primary excision with postoperative radiotherapy appears to achieve more satisfactory local control when compared to either surgery or radiotherapy alone.[13]

## CONCLUSION

Adenoid cystic carcinomas of the nasal cavity are uncommon and description of dedifferentiation form is rare,

despite this the weightage to the diagnosis and management of these tumors should be considered in the differential diagnosis of aggressive nasal cavity tumors. Close follow-up and surveillance of the individual is essential to know about the recurrence and metastasis of the disease and to assess the success of the management of this disease.

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