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Case Report

Adenoid Cystic Carcinoma of Inferior Turbinate

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Abstract

Adenoid cystic carcinoma (ACC) is extremely rare malignant tumor that occurs in nasal cavity and paranasal sinuses. Diagnosis and treatment of ACC is challenging due to its behavior of slow growth, perineural invasion and high tendency of local recurrence, nature of presenting in a vague manner, so its often misdiagnosed and mistreated, in order to diagnose early it requires a high index of suspicion and close follow-up.

It comprises about 3-4% of all the upper aerodigestive tract malignancies, but of all malignant paranasal sinus tumors, 5–15% are adenoid cystic carcinomas [1].

Keywords: Adenoid Cystic Carcinoma (ACC); Nasal Endoscopy; Chemotherapy

Case Report

A 37 -year-old female had come to our OP with mucoid nasal discharge occasionally blood stained and nose block for 3 months, but did not have Facial pain, facial numbness, headache, or altered sense of smell.

Nasal endoscopy revealed a well-defined, reddish-pink fleshy smooth, non-ulcerative mass arising was seen arising from the posterior part of right inferior turbinate (Figure 1). Osteomeatal units were normal on both sides. It was non tender, it did not bleed on touch. Eye examination and neck examination were non-significant. Computed tomography scan of PNS revealed mass in right posterior end of inferior turbinate (Figure 1-3). No cervical lymphadenopathy was present. there was no metastasis in Chest X-ray, bone scan, and abdominal echo was also normal. after getting informed written consent, pre op investigations, anaethestic fitness was obtained, patient was taken up for surgery. Complete endoscopic resection of mass was done which was around 2*3 cm in size confined to only posterior end of inferior turbinate. It was sent for histopathology.

Histologically, There was focal surface ulceration with subepithelial stroma showing an infiltrating neoplasm composed of tumor cells arranged in solid nests, tubules and in focal cribriform pattern. The microcystic spaces of cribriform pattern were filled with hyaline mucoid material Moderate nuclear pleomorphism was noted focally in the solid nests with central come do type necrosis.

There was no evidence of squamous differentiation. The tumor infiltrates focally into the native seromucinous glands. There was no evidence of perineural or lymphovascular invasion noted. Immunohistochemistry was done, positive for P63, SMA, CD117, CD43, calponin, cytokeratin 7 all in basal layers. Ki-67 proliferation index -25%. All consistent with adenoid cystic carcinoma. The

Figure 1

Figure 2

Figure 3

patient recovered fully (Figure 5). There was no recurrence and distal metastasis during the course of follow up.

Figure 4: Pre op.

Figure 5: Post operative.

Discussion

Adenoid cystic carcinoma in the nasal cavity and paranasal sinuses origin, they have poor prognosis than in any other region in head and neck [1,2]. It can occur in any age but has peak incidence in the fourth to sixth decades, more common in females. Usually symptoms are nonspecific, such as epistaxis, nasal obstruction, and symptoms depends on which structure is involved. Nasal bleeding and obstruction were the only presenting symptoms in our patient. A biopsy for definite diagnosis is needed.

The slow-growing adenoid cystic carcinoma can spread within the hollow nasal and sinus cavities before becoming symptomatic. Since the nose and sinuses are surrounded by vital structures, such as the dura, brain, orbit, carotid arteries, and cranial nerves it may often result in an inadequate or high morbidity surgical resection [3]. Pathognomonic feature of adenoid cystic carcinoma is perineural invasion and is one of the determining prognostic factor for local recurrence.

Neck node metastases are extremely rare with adenoid cystic carcinoma. lungs are the most common site of metastasis and the less common sites include the bone, liver, brain, and kidney. Adenoid cystic carcinoma has the tendency to spread via perineurally and hematogenously. Three histologic patterns have been identified: tubular, cribriform, solid, most common type is Cribriform type [4]. Assessment of the histologic grade is of significance in predicting the outcome of tumor recurrence and survival. In one series of studies, 5-year recurrence rates are 99-100% for solid, 85-90% for cribriform and 60% for tubular growth patterns, respectively [5]. When there is presence of greater than 30-35%% solid growth pattern, then it has been reported to have a significantly poorer 5-year survival (5%) when compared to tumors with a predominantly cribriform (26%) or tubular (39%) growth pattern [4]. A combination of surgical resection and postoperative radiotherapy is the treatment modality for sinonasal adenoid cystic carcinoma[6] compared to either surgery or radiotherapy alone. but sometimes despite aggressive clearance, high incidence of positive margins was seen due to the complex anatomy of the nose and paranasal sinuses. So in such cases adjuvant radiotherapy is necessary [7]. Chemotherapy is not ineffective in the treatment of adenoid cystic carcinoma [8]. Long-term follow-up is required and important as high incidence of recurrence and distal metastasis is seen [9,10]. Factors that determine prognosis are perineural invasion, site of tumor, age, staging, histological type, positive surgical margins, and nodal involvement [7]. It has been documented that ACC in nose and maxillary sinus have poor prognosis [1]. For low-grade tumors, five year survival rates is 80-85%.

Immunohistochemical staining for biomarkers can augment information regarding prognosis. IHC markers such as S100, CD117, alpha smooth muscle actin, are positive for ACC while vimentin is negative. It is seen that if there is overexpression of c-KIT mutation in tubular and solid then there is poor prognosis [11].

Conclusion

here in our case, since patient presented early and the tumor was present in a relatively easily accessible area. Surgically clear margins were achieved without disturbing cosmesis or compromising oncologically surgically. Even though adenoid cystic tumors in the lateral nasal wall especially the inferior turbinates is rare, ACC should be borne in mind while having the differential diagnosis of the nasal tumors. Since it is prone for local recurrence and distant metastasis it is recommended to have regular follow ups for a period of time. Complete surgical excision is the aim of our treatment with clear negative margins and in case of close or positive margins adjuvant radiotherapy is needed.

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