Case Report

Adenoid-Cystic Carcinoma of Nasal Septum and Pterygo-Palatine Fossa

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ABSTRACT

Introduction

Adenoid cystic carcinoma (ACC) is an uncommon malignant tumour arising from salivary glands. It is more common in minor salivary glands, palate being the commonest site. Local recurrence, distant metastasis and perineural invasion are the common clinico-pathological features of this tumour. ACC of nasal septum and pterygo-palatine fossa is extremely rare entity with a very few reported cases in the available literature.

Case Report

We report a case of ACC of nasal septum and pterygo-palatine fossa with its surgical management through endoscopic approach and follow up in the background of available literature. The article is aimed to highlight the clinical features, diagnosis, endoscopic approach for surgical management and long term post operative follow up of this rare entity.

Discussion

Adenoid cystic carcinoma (ACC) itself is rare in nasal cavity; where lateral nasal wall is the commonest site of occurrence. ACC arising in the pterygo-palatine fossa is further rare entity; only 3 or 4 cases have been reported till date. ACC arising in these two sites simultaneously is a unique occurrence.

Keywords

Adenoid Cystic Carcinoma; Nasal Septum; Pterygo-palatine Fossa; Endoscopic Resection

Adenoid cystic carcinoma (ACC) is a malignant neoplasm of major and minor salivary glands. ACC corresponds to 1% of the Head and Neck region malignant tumors and contributes 4-10% of all salivary tumours.[1] ACC of nasal septum is exceedingly rare and ACC arising in the pterygo-palatine fossa (PPF) is also very rare; only 4 cases have been reported till date.[2] ACC arising in the septum as well as pterygo-palatine fossa is a unique clinical presentation. Tumour location and perineural spread pattern should be considered in the planning of surgical resection and further treatment. Post operative radiation is now generally recommended and long term follow up is usually necessary keeping in mind that incidence of local recurrence and distant metastasis rate are high.[3]

Case Report

A 63 years old male patient, presented to ENT OPD of a tertiary care hospital with complaints of nasal blockage, more towards left side, for last 1 year along with recurrent epistaxis, mostly from left nostril, for the same duration. He suffered at least two episodes of moderate to severe bouts of spontaneous bleeding per nose, for that, he had to seek medical advice. He consulted many local doctors and took medicines to stop bleeding, nose drops, anti-allergic medications; but his nasal blockage gradually progressed and nose bleeding did not stop. On anterior rhinoscopy, there was mild deviation of nasal septum towards right with thickened, pale mucosa on the right side. On the left side, a swelling or tumour like mass seen arising from the nasal septum and occupying the

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valve area of the left side of nose. The mucosa overlying the mass appeared pale, thickened and there was mild bleeding on touch. The mass was sessile, non-mobile, attached to the septum and free from other sites. No evidence of active bleeding per nose or muco-pus in the nostrils was noticed at the time of examination except while touched. Posterior rhinoscopy examination was unremarkable. Patient had a dimness of vision on left eye for last 5 to 6 years, which according to the ophthalmologist, was due to presbyopia. There was no proptosis and eye-ball movement was not restricted on either side. Other cranial nerve examination was within normal limits and clinically no neck nodes were palpable. A plain and contrast-enhanced C.T. scan of para nasal sinuses was done, which showed an irregular enhancing soft tissue mass measuring 12x11 mm at left pterygo-palatine fossa with erosion of left pterygoid plates, left sphenoid wing, floor of sphenoid sinus with extension to left parasellar region. The mass was protruding into left sphenoid sinus, middle meatus and left orbital apex. (Figure 1)

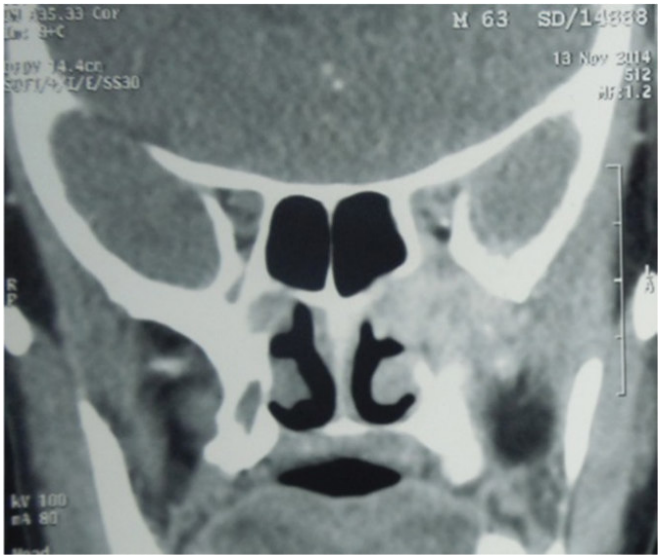

Fig. 1. Enhancing soft tissue mass in left pterygo-palatine fossa

There was deviated nasal septum to right side with septal mucosal thickening and a soft tissue mass arising from the septal mucosa seen on the left side. (Figure 2)

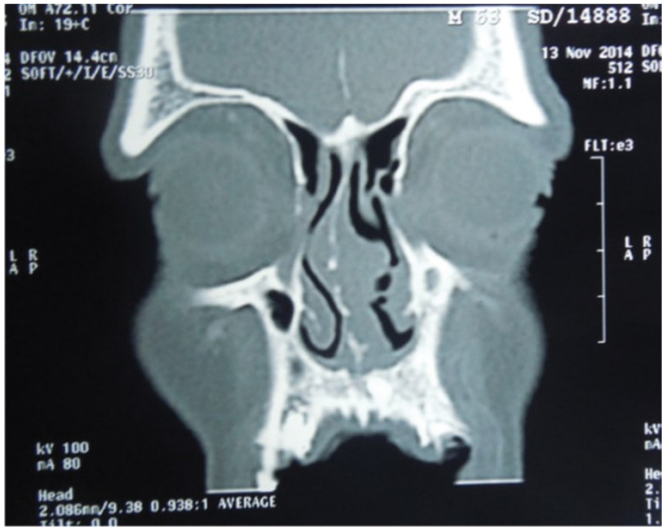

Fig. 2. Enhancing soft tissue mass in left side of nasal septum

Diagnostic nasal endoscopy corroborated the radiological findings. An excision biopsy of the both the masses at left pterygo-palatine fossa and septum were planned after proper anaesthetic check up and making the patient fit for general anesthesia. Patient was admitted and operated under general anesthesia. Diagnostic nasal endoscopy was done to plan the operative procedure. First, the tumour arising from the nasal septum excised with wide local margin dissecting sub muco-perichondrially. Bleeding was controlled with bipolar diathermy. Then, the medial wall of the left maxillary sinus and the middle turbinate were removed to gain access to the posterior wall of the sinus. The orbital process of the palatine bone was removed and the sphenopalatine foramen was enlarged. The posterior wall of the maxillary sinus was then removed up to the vertical process of the palatine bone medially and up to the angle between the lateral and posterior wall of the sinus laterally to expose the pterygo-palatine fossa. The tumor was then scooped out from the fossa and the bleeding was controlled with endoscopic bipolar cautery. The nasal cavity was packed with roller gauge pack which was removed after 48 hours. Patient was discharged from the hospital with a course of oral antibiotics, anti-histaminics, local decongestant drops, and analgesics. A post-operative nasal endoscopy was performed after a fortnight to check mucosal healing and to clear crust and debris. No bleeding points or left out tumor were found.

Histopathological examination (HPE) report came as ACC of nasal septum and pterygo-palatine fossa (Figures 3 & 4) and the resected margins of the septal tumor was free of malignancy three dimensionally, but comment on margin for other tumor was not available as it was scooped out.

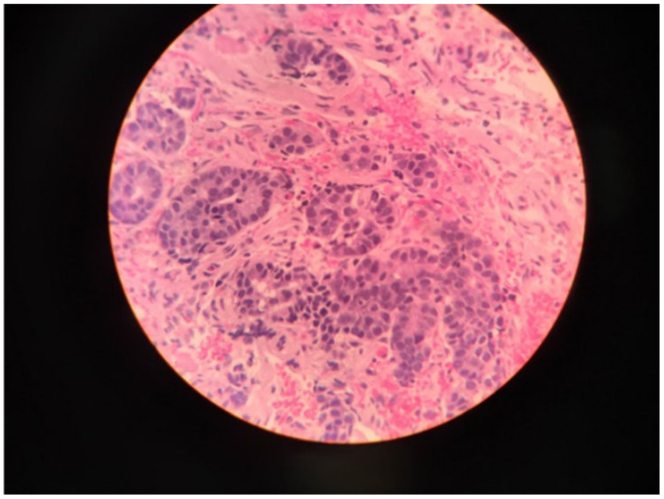

Fig. 3. HPE picture of mass in left Pterygopalatine fossa (H-E stain wit 10 X magnification)

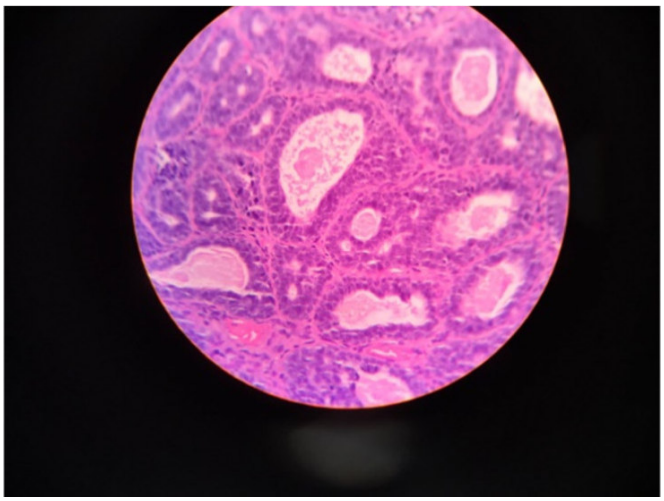

Fig. 4. HPE picture of mass in Nasal Septum (H-E stain wit 40 X magnification)

With this HPE report patient was referred to medical oncologists for opinion regarding further management and they advised for a course of adjuvant radiotherapy for this patient. Accordingly the patient underwent a 28 days external beam radiation therapy which he managed to complete over a time span of 6 weeks (5 days a week).

Thereafter patient was followed up for a period of one year, at the three months interval. Diagnostic nasal endoscopy was done at each visit to check local recurrence and complication, if any. No evidence of any recurrence of tumour was found till the end of one year follow up. Patient suffered some radiation related complications like dry mouth, otitis media with effusion, mucositis and dermatitis; which were managed accordingly.

Discussion

Incidence of malignant tumours from the nasal septum is quite rare ranging from 2.7% to 8.4% of nasal and paranasal malignant tumours.[1] Adenoid cystic carcinoma (ACC) is rare in nasal cavity; where lateral nasal wall is the commonest site of occurrence.[1,4] ACC arising in the pterygo-palatine fossa is also a rare entity; only 3 or 4 cases have been reported till date.[2] ACC arising in these two sites (Double Primary Cancer) at the same time is a unique case. ACC of nasal cavity or septum most commonly presents with mass or epistaxis and that of pterygo-palatine fossa (PPF) usually presents with epistaxis, facial pain and the symptoms attributed to the spread of tumour from PPF; visual disorders being the commonest.[2,5,6] Biological behavior of ACC includes slow growth rate, perineural spread and high tendency of local recurrence.[1] Three architectural growth patterns have been described, namely: cribriform, tubular and solid (anaplastic). Cribriform pattern is also called cylindroma; presents with typical "Honeycomb" or "Swiss-cheese" pattern. Tubular pattern has a very aggressive course while the solid variety has the worst prognosis.[7] Though a slow growing tumour (peak incidence in 4^{th}-6^{th} decades), it has a high incidence of metastasis; early perineural and late hematogenous spread. Most common site of distant metastasis is lung, usually multiple and being associated with tumours with a solid growth pattern.[1,5]

Combined wide local excision with post-operative radiotherapy is the treatment of choice; which gives more satisfactory local control when compared to either surgery

or radiotherapy alone.[1,3] Endoscopic trans-nasal excision of tumors of septum, nasal cavity and PPF is now widely practiced[8,9] which may be combined with navigation, where facilities available. The medial wall of the maxillary sinus and the middle turbinate are removed to gain access to the posterior wall of the maxillary sinus. The orbital process of the palatine bone is removed and the sphenopalatine foramen is enlarged. The posterior wall of the maxillary sinus is then removed up to the vertical process of the palatine bone medially and up to the angle between the lateral and posterior wall of the maxillary sinus laterally to expose the PPF. Following resection of the tumour, all the symptoms attributed to the tumour in PPF and extension wherefrom; can be resolved including visual deficit, if any. Post-surgical radiotherapy amounts not less than 60 Gy (200 cGy/ fraction – 30 fractions in 6 weeks)[7] which enhances the local and regional control of the tumour. The role of chemotherapy in the management of ACC is still controversial.[7] But as the ACC in sinunasal compartments are often not completely resectable, adjuvant therapy with neutron irradiation, chemotherapy with taxols and carboplatins and targeted therapy are suggested by some authors.[10] Long term follow up is needed in these cases owe to increased chances of local recurrence and distant metastasis. Early detection of the tumour and treatment guides prognosis due to less likely to have chances of advanced neural involvement and metastasis to regional or distant sites.

Conclusion

Adenoid cystic carcinoma may be considered in the differential diagnosis of tumours of nasal septum and pterygo-palatine fossa; common presenting symptom being the epistaxis. Diagnostic nasal endoscopy and C.T.scan of paranasal sinuses guides the definitive treatment. Endoscopy guided excision and post-operative radiotherapy is the treatment of choice. Long term follow up is necessary, keeping in mind its perineural invasion, local recurrence and distant metastasis.

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