

LYMPHOEPITHELIAL CARCINOMA OF PARANASAL SINUS: A RARE CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Background: Lymphoepithelial carcinoma is one the rare tumor of nose and paranasal sinus with only few cases reported so far. Epidemiology, etiopathogenesis, modes of presentation and the management of this condition still remain fields to be explored.

Case Presentation: A 55 years old lady presented with right sided nasal obstruction with nasal discharge for 2 years. Nasal endoscopy showed multiple pinkish, polypoidal mass almost filling whole of right nasal cavity. Imaging study showed soft tissue density with hyperdense area in right ethmoid and sphenoid sinuses. Biopsy of the lesion revealed Lymphoepithelial carcinoma. Patient was managed by endoscopic debulking followed by radiotherapy. There was no evidence of tumor on follow up evaluation, six months after radiotherapy.

Conclusion: Lymphoepithelial carcinoma can affect nose and paranasal sinuses. Although, a consensus on management of the disease is yet to be achieved, the best management so far is surgery followed by an adjuvant radiotherapy.

Key words: paranasal sinus neoplasm, sphenoid sinus, ethmoid sinus

INTRODUCTION

Lymphoepithelial carcinoma (LEC) of nose and

paranasal sinuses is a rare malignancy.⁽¹⁾ With very few cases reported so far, epidemiology, etiopathogenesis, modes of presentation and the management characterized by dense lymphocytic infiltration. Being radiosensitive tumor, radiotherapy (RT) is considered primary modality of the management.⁽²⁾ Here, we present a case of lymphoepithelial carcinoma of the ethmoid and sphenoid sinus which was managed with surgery and radiotherapy with favorable outcome.

CASE PRESENTATION

A 55 year old lady presented with the chief complaints of right sided nasal obstruction with nasal discharge for last 2 years. Nasal obstruction was gradual in onset and slowly progressive. It was associated with unilateral, right sided nasal discharge which was mucoid most of the times and sometimes associated with a blood tinge. She had no other symptoms such as headache, fever, midfacial discomfort, frank epistaxis, anosmia or hyposmia, altered sensation in the face or any change in the vision. There were no aural complaints such as earache or decreased hearing. She was a newly diagnosed hypertensive with no other significant past medical and treatment history. She was a smoker for 20 years with consumption of 8-10 cigarettes per day, which she left 8 years back. She is a farmer by occupation. General examination didn't yield any

abnormal findings. Anterior rhinoscopy and nasal endoscopy showed multiple pinkish, polypoidal mass almost filling whole of the right nasal cavity. On probing the mass was insensitive and bled on touch. Probe could not be passed on the superior and lateral aspect of the mass. Rest of the clinical examination findings were normal. Ophthalmological examination was also normal. As it was a unilateral pinkish nasal mass which bled on touch, inverted papilloma was considered first differential diagnosis. Contrast enhanced computed tomography (CECT) scan and biopsy was advised. The CECT scan showed soft tissue density with hyperdense area within, involving right ethmoid and sphenoid sinus. The lesion showed slight enhancement. No obvious bone erosion or destruction was seen (Figure 1).

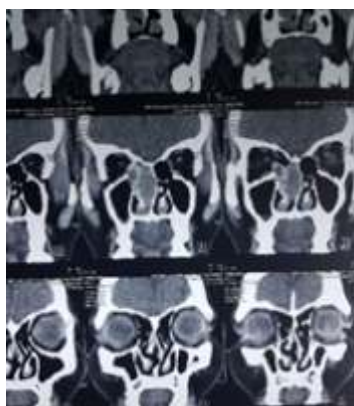
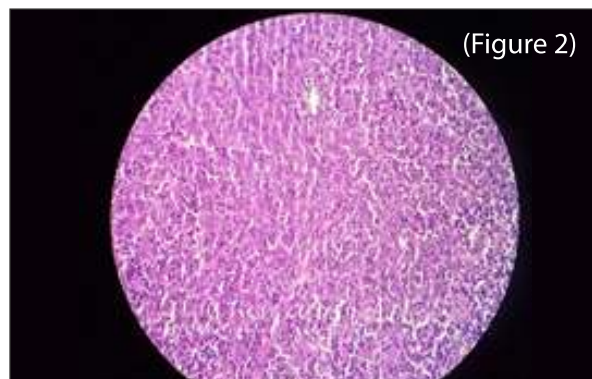


Figure 1 – Computed tomographic scan coronal view shows soft tissue density in right ethmoid sinus and right sphenoid sinus extending to right nasal cavity.

Biopsy of the nasal mass came out to be lymphoepithelial carcinoma. Histological features showed infiltration with malignant cells arranged in syncytial sheets with oval tumor cells showing ill-defined borders and large vesicular nuclei with single nucleolus. There was dense lymphocytic infiltration in the stroma intervening the tumor nests.

Figure 2- Malignant cells arranged in syncytial sheets with oval tumor cells showing ill defined cell borders and showing large large vesicular nuclei with prominent single nucleolus. Stroma

intervening the tumor nests show dense lymphocytes infiltrating these tumor nests.



(Figure 2)

Patient was planned for endoscopic debulking followed by radiotherapy. Per operative findings were pinkish, friable mass present in right sphenoethmoidal recess and adjacent part of the septum, sphenoid sinus and posterior ethmoids. All sinuses were opened along with debulking of all the disease. There was no underlying bone erosion in the skull base or any extension into the orbit. Right nasal packing was done by polyvinyl alcohol pack which was removed after 48 hours. Patient was discharged on oral antibiotics and followed up after 2 weeks with the histopathological report which came out to be lymphoepithelial carcinoma again. She was advised for radiotherapy. Adjuvant radiotherapy was given in dose of 2 Gray (Gy) five times a week for a total dose of 70 Gray. Six months after completion of radiotherapy she was evaluated again. Nasal endoscopy showed no presence of mass or any abnormal mucosal architecture except synechiae and minimal mucosal edema in the region of ethmoid air cells.

DISCUSSION

Carcinoma of nasal cavity and paranasal sinus accounts for 3% of head and neck malignancies. (2) Lymphoepithelial carcinoma is rare malignant tumor first described by Schminke and Regaud in 1921.(1) It is non keratinizing undifferentiated carcinoma commonly seen in head and neck region including nasopharynx, paranasal sinus,

oral cavity, larynx, and salivary glands. Besides this, it can also occur in many organs such as lung, stomach, skin, breast, bile ducts, and oesophagus.^(3,4) Involvement of nose is more common than the involvement of the paranasal sinuses although overall incidence of LEC in these regions is very rare.⁻⁽⁵⁾

Lymphoepithelial carcinoma is mostly reported from Southeast Asia where nasopharyngeal carcinoma is also common. Males are frequently affected than females with a ratio of 3:1. It affects adults in the fifth to seventh decade of life.⁻⁽⁶⁾ It shows strong association with Epstein Barr Virus (EBV).^{-(4,6,7)} LEC has to be differentiated from more aggressive sinonasal undifferentiated carcinoma which are EBV negative. Presentation of LEC affecting nose and paranasal sinuses may vary among individuals. It may be asymptomatic or may present with nasal obstruction, mass in the nasal cavity, epistaxis, lacrimal duct obstruction or neck nodes. Lesion in nasal cavity is diagnosed at early stage due to early symptoms however, frontal and sphenoid sinus tumors are diagnosed at late stage. In our case the disease was diagnosed at early stage. Intracranial extension of tumor may cause cranial nerve palsy or proptosis.

Confirmation of diagnosis always requires histopathological examination as there are no classical diagnostic features on radiological imaging. Histologically the tumor is characterized by presence of islands, sheets, trabeculae or singly arranged cells. There is intermingling of tumor cells with lymphocytes and plasma cells. There is no necrosis or keratinization, separating this entity from sinonasal undifferentiated carcinoma.^{-(6,8)} Immunostaining for pancytokeratin (AE1/AE3) and epithelial membrane antigen helps to epithelial nature of tumor.⁻⁽⁹⁾ Immunohistochemistry (IHC) or Epstein Barr encoded RNA In-situ hybridization can help in defining EBV positivity however, this offers no additional benefits on therapeutic strategy and overall prognosis remains same.⁻⁽⁷⁾ Being an undifferentiated carcinoma, this tumor must be differentiated from sinonasal undifferentiated

carcinoma, malignant melanoma and lymphoma by IHC.

Having a handful number of cases reported so far, there has not yet been a consensus regarding the treatment guidelines for LEC of nose and paranasal sinuses. Various treatment modalities have been mentioned in literature such as RT only, surgical resection followed by RT, and chemoradiotherapy.⁻⁽¹⁰⁾ Radiotherapy remains the mainstay of treatment as this tumor is highly radiosensitive.⁻⁽¹¹⁾ Early stage disease of nose and paranasal sinus (Stage I and II) can be managed by RT alone however, Stage III and IV requires combination of surgery and radiotherapy. The dose of irradiation ranges from 55.8 Gy to 77.2 Gy with mean dose of 68 Gy. Irradiation can be given 1.8 Gy once daily or 1.1 to 1.2 Gy twice a day.⁻⁽¹⁰⁾ In our case, adjuvant radiotherapy was given in dose of 2 Gray (Gy) five times a week for a total dose of 70 Gray.

Due to close proximity of orbit, skull base and central nervous system it is difficult to manage only with high dose of radiotherapy. Previously most of these lesions were treated with high dose of irradiation only resulting in high incidence of blindness and mortality.^{-(9,10)} Recently, management includes resection of tumor followed by postoperative irradiation. It has reduced the eye complication, improved local control and absolute survival. In another study, LEC of maxillary sinus was managed by radical excision followed by external beam RT (48Gy).⁽¹⁾ In our case also, endoscopic debulking of tumor was followed by RT. Combined modality including both chemo- and radiotherapy can be considered for primary tumor as well as those associated with systemic metastases.⁽¹⁾ Concurrent cisplatin during RT followed by adjuvant cisplatin and fluorouracil has been used for advanced cases.⁻⁽¹²⁾ Hajjiannou et al.⁽²⁾ reported a case of LEC of nasal cavity and ethmoid sinus successfully managed by external radiotherapy (66 Gy) and chemotherapy (5-fluorouracil and cisplatin). Advanced LEC of maxillary sinus have been managed by surgical excision followed by

chemotherapy (Docetaxel + Carboplatin) and Radiotherapy (6300 cGy) with 3 years disease free survival.⁽¹³⁾ Elective neck dissection not done for early stage disease however, advanced stage LEC requires neck dissection along with other treatment modality.(3)

Prognosis of this tumor depends entirely on staging. The cases with locally confined tumors fare well in contrast to those cases who have developed distant metastasis.⁽⁵⁾

CONCLUSION

Lymphoepithelial carcinoma can affect nose and paranasal sinuses. Although, a consensus on management of the disease is yet to be achieved, the best management so far is surgery followed by an adjuvant radiotherapy.

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