EOSINOPHILIC MUCIN RHINOSINUSITIS
MASQUERADING AS MALIGNANCY

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Abstract

Introduction: Extensive inflammatory nasal masses with orbital and intracranial extension can mimic nasal malignancies and their management generally require extensive open surgical approaches. However with good imaging, preoperative knowledge of histology and use of endoscopic approaches with powered instruments, such extensive open surgeries can be avoided.

Case report: We are reporting a case of an extensive unilateral nasal mass with orbital and intracranial extension which on initial evaluation appeared to be a malignant lesion. It was managed with endoscopic sinus surgery. Final histopathology and microbiology were suggestive of an inflammatory lesion - eosinophilic mucin rhinosinusitis (EMRS).

Conclusion: EMRS is an inflammatory nasal mass and it can be extensive mimicking a malignancy. Conservative endoscopic approach is now the treatment of choice for most of these inflammatory nasal masses and even intracranial, extensive supraorbital and orbital extensions can be excised endoscopically.

Keywords: Rhinosinusitis, Nasal polyps, Mucin, Eosinophil, EMRS

Introduction

Extensive inflammatory nasal masses with orbital and intracranial extension masquerade as malignancies and have been traditionally excised by open surgical approaches with facial incisions. However nasal masses which are medial to midpupillary line can be excised endoscopically without using open surgical approach. Some cases of chronic rhinosinusitis with polyps present as unilateral nasal mass with bony destruction and proptosis. They masquerade as malignant lesions. However with good imaging for extent of lesion, preoperative knowledge of benign histology they can be managed well with conservative surgical approach of endoscopic sinus surgery instead of open surgery.

Similar outcomes with better patient satisfaction are achieved with endoscopic approach as an external scar is avoided and there is preservation of facial sensations as infraorbital nerve is preserved.

We report one such case of an extensive right nasal mass.

Case Report

A 22 year old male patient presented to our centre with complaints of rhinorrhoea and nasal obstruction right nasal cavity of three months and proptosis right eye of two months duration. (Fig 1).
Rhinorrhea was mucoid and nasal obstruction was gradually progressive. He developed proptosis of right eye which was also insidious in onset and gradually progressive and was affecting his gaze in superior direction. He did not complain of anosmia, hyposmia, epistaxis, facial pain/paresthesia, headache, epiphora, diplopia or diminished vision. He was immunocompetent with no history of any addiction to drugs/alcohol/tobacco. There were no comorbidities.

On examination he had no external nasal deformity. Diagnostic nasal endoscopic examination of right nasal cavity revealed polyps completely filling it, inflamed mucosa and septum pushed to left side. (Fig 2).

MRI of paranasal sinuses (PNS) revealed an extensive heterogeneous right nasal cavity lesion on T1 weighted images. The lesion involved right maxillary, frontal, ethmoid and sphenoid sinuses extending into spheno-ethmoidal recess. It had intracranial, supraorbital and orbital extension. (Fig 3 A,B) Right orbital contents were compressed however no infiltration of muscles or eyeball was seen and fat planes were preserved. Components of right frontal sinus were focally extending into anterior cranial fossa without any meningeal enhancement or involvement of neuroparenchyma. There was no evidence of cavernous sinus thrombosis. Due to the clinical picture and its extent on imaging it appeared to be a malignant lesion.
examination.
Non-contrast computed tomogram (NCCT) of PNS showed hyperdense contents involving right paranasal sinuses with lysis of intervening walls of PNS. (Fig 4A) Floor and medial wall of right orbit was eroded including medial wall of optic canal. Posterior table of frontal sinus on right side was eroded and lesion was extending intracranially.

Fig 4: NCCT paranasal sinuses of sinonasal lesion
4A – Preoperative axial NCCT showing intraorbital extension of sinonasal lesion
4B – Postoperative axial NCCT showing cleared sinuses and correction of proptosis

In view of a clinico-radiological impression of a malignancy and a negative biopsy, a repeat biopsy was planned under general anesthesia (GA) so that a representative sample could be obtained. Also a differential diagnosis of allergic fungal rhinosinusitis was entertained so the patient was counseled and consented for endoscopic sinus surgery too.

Patient was taken up for Endoscopic sinus surgery under GA, after the superficial polyps were removed and a deeper biopsy was taken. Insipissated secretions were found in the depth which are usually seen in allergic fungal rhinosinusitis (AFRS). Orbital decompression was also performed in the same sitting. Inspissated secretions and polyps seen in maxillary, frontal, ethmoids, sphenoid sinuses, supraorbital cells and near orbital apex were removed. Duramater of anterior cranial fossa was found exposed at the posterior table of frontal sinus which had been eroded by the disease. Frontal sinus floor was also eroded. Septum was pushed to the left side.

Disease was cleared completely which encompassed addressing anterior skull base by endoscopic skull base surgery. (Fig 5).

Fig 5: Excised nasal mass
Left nasal cavity was also inspected and its mucosa was normal. No polyps, mass or any mucopus was seen there. Histopathological examination was reported as inflammatory polyp. Microbiology did not show any fungal elements, however there was eosinophilic infiltrate and Charcot-Leyden crystals.

Post operatively, proptosis resolved completely and eye mobility achieved in all directions of gaze.

On follow up, nasal endoscopy revealed healed nasal mucosa with no recurrence of disease.

Repeat NCCT PNS after 03 months, showed post operative status with no evidence of recurrence. (Fig 4B)

Discussion
A number of sinonasal masses are encountered in clinical practice. Extensive nasal masses are many a time a diagnostic as well as therapeutic
challenge. In our case too, the lesion was extensive with intraorbital and intracranial extension. The lesion being unilateral and patient presenting with nasal obstruction and proptosis of short duration, malignancy was definitely high on our list of differential diagnoses. However it was not proved so on histopathology. A number of such similar cases have been reported in literature—(13). We were able to go ahead with complete surgery when we had taken up the patient for biopsy under GA because we had considered AFRS too in the differentials and had accordingly taken patient’s consent. So it highlights the importance of considering other differentials too even in a positively malignant looking lesion on imaging.

This case also highlights those extensive lesions which are compressing orbital contents eroding skull base, extending intracranially can be excised endoscopically without the need for open surgical approach. It has been reported in literature that lesions which are medial to mid-pupillary line can be excised endoscopically—(4, 5).

The other point for discussion in this case is the histology and microbiology. Intraoperatively it appeared to be a case of AFRS because of the allergic mucin and unilateral polyps. The excised specimen had been sent for histopathology as well as microbiology including bacterial stains and culture as well as fungal stains and culture. No fungal elements were demonstrated so AFRS remained unproven on histopathology or microbiology. However eosinophilic infiltrate and Charcot Leyden crystals were noted in the specimen. This brings us to another entity described in literature as eosinophilic Mucin rhino-sinusitis (EMRS).

Eosinophilic rhino-sinusitis is a group of clinical conditions in which AFRS and EMRS are its subcategories(6). EMRS is histologically similar to AFRS in clinical picture but it lacks the fungal elements on histology/microbiology. It has been thought that etiopathogenesis of EMRS is not a hypersensitivity to fungal elements but a systemic dysregulation associated with upper and lower airway eosinophilia(6). It is important to know both about AFRS and EMRS so as to identify common elements associated with both of them and to differentiate between the two entities. Aetiologically, presence of allergic mucin rich in eosinophils and non-invasive fungi, together with marked elevation of blood levels of specific IgE antibodies is diagnostic of AFRS. Ferguson et al describe EMRS as local immune reaction occurring secondary to a systemic disorder with normal IgE levels and absence of fungi. A study by Orlandi et al demonstrated that the genetic profiles of both AFRS and EMRS were similar, but differences do exist and further studies need to be done for conclusive results(7). A study done by Uri et al demonstrated more aggressive behaviour of AFRS in comparison to EMRS in terms of orbital complications (50% as compared to 8% respectively). AFRS is a more common diagnosis made by most of the ENT surgeons and clinical suspicion for EMRS is raised either with post-operative histopathological and culture reports or with history of multiple relapses or multiple surgical procedures(8).

Conclusion

A unilateral nasal mass with orbital and intracranial extension can have differential diagnosis from inflammatory, benign to malignant lesions, and warrants workup accordingly. Malignancy should be ruled out by deep biopsies which may require general anaesthesia. When a surgeon is approaching such a case for biopsy under general anaesthesia, it is good practice to take consent of the patient for a full-fledged endoscopic sinus surgery for removal of disease completely if intraoperative findings suggest a benign lesion. It also reiterated here that extensive lesions of nose which are medial to midpupillary line can be excised endoscopically as has been seen in this case.

Another important point in this case report is the difficulty to differentiate between AFRS and EMRS.
as both of them have the similar initial clinical presentation. AFRS and EMRS are used interchangeably many a times however the outcomes of surgery may differ in the two. There is increase in the chances of relapses and the requirement of surgical procedures in EMRS. More specifically, the clinical course of AFRS always carries a higher prevalence of orbital complications and associated morbidity. The role of fungus and the ability to confirm its presence are still controversial issues.

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**Compliance with Ethical Standards**

**Ethical Approval** - The research involved a human participant.

**Conflict of interest** - The authors have none to declare.

**Consent** – Written informed consent was obtained from the individual participating in the study

### References


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