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Unexpected nasal mass histology- Case report of a rare tumour

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Abstract: Intestinal type adenocarcinoma (ITAC) is a rare malignancy of nose and PNS. We report a case of 40 year old man with intestinal type adenocarcinoma of nose and PNS. Presentation was unilateral nasal mass with nasal blockage. Initially was treated in community with topical steroid spray as rhino sinusitis and later on referred to ENT department. Treatment was given in form of major craniofacial surgery once diagnosis was confirmed. We discuss the presentation, investigation, differential diagnosis and management aspects. Case report also high lights the importance of early referral of patients with unilateral nasal mass to ENT department.
Keywords: Adenocarcinoma nasopharynx, Intestinal type tumour, nasopharyngeal carcinoma

INTRODUCTION:

Intestinal type nasopharyngeal carcinoma (ITAC) of nasal cavity and paranasal sinuses is a rare tumour of nasopharynx. Occupational exposure of wood dust is a known risk factor but sporadic cases are seen as well. Intestinal type nasopharyngeal carcinoma shows strong male predominance (M: F ratio 4:1) [2]. Mean age group affected is around 60 years [2].

Clinical features include unilateral nasal blockage, nasal discharge, and epistaxis. In advanced cases orbital involvement can cause visual symptoms and exophthalmos. Nerve palsies may also be apparent in advance cases. Diagnosis is based on histology. Radiological imaging helps in determining extent of tumour. Colonoscopy can be used to differentiate between primary and secondary disease. Surgery is considered as the primary modality of treatment. Prognosis depends on histologic subtype.

CASE REPORT:

A 40 year old man presented to ENT outpatient department with 9 months history of unilateral nasal mass associated with nasal blockage. Patient was given topical steroid nasal sprays in primary care with no improvement in symptoms and subsequently referred to ENT department. Other associated symptoms included nose bleeds and sensation of blocked ears. Patient has a background of smoking about 15 cigarettes per day, on multiple anti-epileptic drugs for epilepsy. No history of exposure to wood dust in this patient.

On examination patient was found to have large right side necrotic nasal mass (Figure 1) with associated bleeding on contact. Pus was found in both nasal cavities. A small biopsy sample was taken in OPD and CT scan was requested. Ear examination was normal on both sides. Initial biopsy results were inconclusive due to presence of only superficial tissue's scan showed soft tissue nasal mass with involvement of nasal sinuses (figure 3A, 3B). Ethmoid sinuses were involved bilaterally while frontal, sphenoid and maxillary sinuses were mainly involved on right side. The mass appeared to be expansible with pressure effects on surrounding tissue.

Fig-1: Endoscopic view of right nasal mass

Fig-2: Endoscopic view of right nostril after tumour debulking

Fig- 3A: Coronal CT scan nose and PNS showing right nasal mass extending into paranasal sinuses

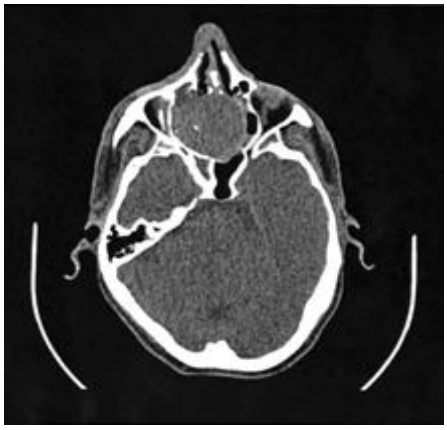

Fig- 3B: Axial CT scan nose and PNS showing right nasal mass extending into paranasal sinuses

Fig-4: Coronal CT scan chest, abdomen and pelvis showing thickening of sigmoid colon

Endoscopic biopsy and debulking of lesion (Figure 2) was done. Intraoperatively a large fibrotic and necrotic mass was found arising from pedicle on right middle turbinate. A clean and smooth edge posterior septal perforation was also seen. Biopsy results showed moderately differentiated intestinal type nasopharyngeal adenocarcinoma. Case was discussed in MDT and MRI of brain and sinus showed possible residual disease and hence decision was made to proceed with surgery. Subsequent imaging of chest, abdomen and pelvis showed thickening of sigmoid colon (Figure 4) but colonoscopy did not show any pathology of colon. Decision was made to proceed with surgery followed by radiotherapy. Endoscopic assisted major craniofacial resection was done, which involved bilateral ethmoidectomy, bilateral sphenoidectomy, right maxillary disease clearance, right medial orbital wall excised, partial posterior nasal septectomy and exploration of right pterygoplaatine fossa. There was no major complications seen in post-operative period apart from expected facial oedema, eye lid oedema restricting vision and nose bleeds. Oedema and nose bleeds eventually improved in a week's time. Patient is awaiting radiotherapy.

DISCUSSION:

Intestinal type nasopharyngeal carcinoma (ITAC) of nasal cavity and paranasal sinuses is a rare disease and it represents less than 4% of all malignancies of this region [1].These tumours resemble in the appearance of intestinal adenomas and carcinomas, or exceptionally the normal glandular epithelium of the intestinal mucosa [6]. Ethmoid sinus is the most common location (40%) followed by nasal cavity (25%) and maxillary antrum (20%) [3]. Wood dust exposure generally shows predilection for ethmoid involvement while sporadic cases arise in maxillary sinus [3]. Incidence is 1000 times higher than normal population for people working in wood dust industry [3]. About 40 years of exposure on average is reported [1]. Usual presentation is unilateral nasal mass. Imaging studies includes CT and/or MRI scans. Involvement of colon can be assessed by colonoscopy. Histologic diagnosis is essential. In general any unilateral nasal lesion needs urgent evaluation in order to exclude any malignancy. Delayed diagnosis can increase morbidity and mortality.

According to Barnesadeno carcinomas of nasopharynx can be classified as: colonic, papillary, solid, mucinous and mixed[3]. Alternatively, the pattern mentioned by Kleinsasser and Schroeder comprises four types: papillary tubular cylinder cell of various grades, alveolar goblet type, signet-ring type, and transitional type [4]. Common histologic types include papillary and colonic [3, 6]. Papillary type is characterised mainly by papillary growth pattern with some tubular elements vice versa colonic type is characterised predominantly

by tubuloglandular architecture [2]. Positivity for immunohistochemistry markers CD20 (73%), CDX2, villin, MUC2 aids diagnosis [2, 5]. This Immunophenotypic is quite different from respiratory mucosa, which helps in differentiating [1]. There may be Chromogranin cells present in cluster or scattered [2].

Differential diagnosis includes low grade sinonasal adenocarcinoma [2, 6]. Low grade sinonasal adenocarcinoma are usually not associated with wood dust exposure may have papillary/tubular architecture; more glandular, less papillary, few columnar / goblet cells[2,6]. Metastatic disease from colonic adenocarcinoma is rare, but can be differentiated on the basis of immunohistochemistry features i-e CEA+, CK7-, Chromogranin, clinical features and colonoscopy are helpful [2, 6]. Sometimes Papillary sinusitis with abundant mucinous material can mimic as well but has short and blunt papillae with thick and hyalinized basement membrane; ciliated surface cells; prominent eosinophils and no significant cytological atypia [2, 6]. History of wood exposure is generally missing in papillary sinusitis [2, 6].

Cytogenetic studies show frequent association with mutations of P53 and K-RAS genes [2]. Tumours associated with occupational exposure to wood dust show p53, p14 [ARF] p16 [INK4a] gene deregulation [2].

Treatment is based on adequate surgical removal and postoperative radiotherapy for advanced cases [2]. Early diagnosis is essential in order to catch disease process early, which may be help full in reducing extent of surgical resection. These are aggressive neoplasms, showing both local recurrence and metastases [5]. The five year survival for sinonasal high grade intestinal type adenocarcinoma is less than 20% [7]. Barnes noted that patients with papillary tumours had a slightly better prognosis [3].

CONCLUSION:

Intestinal type adenocarcinoma of nasopharynx is a rare entity. Exposure to wood dust is a known risk factor. Early diagnosis can affect morbidity in long term. Diagnosis is based on histology. Radiological Imaging studies helps in determining extent of disease and to exclude distant primary/metastasis. Surgical excision is the main modality of treatment. Post-operative radiotherapy is given for advance cases. Prognosis is generally poor.

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