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A Case Report - Malignant Melanoma of Nose Mucosa and Paranasal Sinuses

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ABSTRACT

Introduction: Primary malignant melanoma of nasal mucosa and paranasal sinuses is a rare tumor of uncertain aetiology, unpredictable biological behaviour and poor prognosis. Unlike skin melanomas, there are no risk factors and the disease frequently manifests in older patients, with non-specific symptoms of nasal obstruction, rhinorrhea and epistaxis. Unfortunately, this disease is diagnosed usually in advanced stages which makes it inoperable.

Case Report: Report: A 68 year-old woman presented with right nasal obstruction and epistaxis for 2 months. On clinical examination a pink polypoidal soft, non tender mass filled right nasal cavity which did not bleed to touch. Punch biopsy reported malignant melanoma with invasion of the nasal cavity structures and extend into anterior part of nasopharynx. The patient was submited for surgery and do lateral rhinotomy and pt follow up first time at 15 post op days.second follow-up at1.5 month, third follow-up at 3month , fourth follow up at 6month that time patient symptomps free and there after patient not coming for follow up.

Conclusion: Malignant melanomas of nasal mucosa and pns in general, diagnosed in advanced stages. Their histological characteristics in the mucosa difficult the surgical treatment, that is one of the most efficient options, because they are resistant to chemo and radiotherapy. The early diagnosis, then, is the best option for this tumor nowadays.

KEYWORDS Malignant melanomas , nasal mucosa, Paranasal sinuses, lateral rhinotomy , chemotherapy

INTRODUCTION

- Melanoma is a malignant, potentially fatal neoplasia, that usually arises from the skin, and more rarely from the mucosas. Over 90% of the melanomas are located on the skin , its mucosal appearances are less frequent and represent less then 5% of all the tumors of this nature. The latter has special characteristics and biological behavior which is very different then its skin counterpart, in spite of the fact that there are no morphological differences between them .
- The incidence of head and neck melanomas vary from 0.4 to 4%, and the tumors usually affect individuals between 50 and 70 years of age; there is a subtle preference for males, although neither gender nor age influence prognosis . The most common sites of mucosal melanomas in the upper airways are the oral cavity, the nasal cavity and the paranasal sinuses, in this order of frequence .
- The primary mucosal melanomas of the nose and paranasal sinuses represent 1% of all melanomas. Most of the cases affect the nasal septum, the inferior conchae and the middle conchae, in this order. The clinical manifestations are inespecific, corresponding to the same found in other tumors, that being: nasal obstruction, epistaxis and nasal volume enlargement .
- For diagnostic purposes, we should consider that the large metastatic head and neck melanoma (primary from the skin) is very rare . Only from 0.6 to 9.3% of patients with skin melanoma have metastasis to the mucosa of the upper aero-digestive tract, and the most frequent metastasis sites are the tongue base and the nasal cavity. These are lesions that appear usually from 2 to 7 years after the initial skin lesion and, when they appear in the head and neck mucosa, usually the patient already has disseminated metastasis .
- Broad surgery together with radiotherapy offer the best prognosis . Immunotherapy has not been useful in the treatment of melanomas of the mucosas. The most frequent treatment failure has been local recurrence of about 40% in the nasal cavity, 32% in the pharynx and 25% in the oral cavity. Survival rates are extremely low, for only 5 to 15% of the patients have a 5 year survival. The high rate of local recurrence, in spite of an apparently properly carried out initial surgery, mandates a more radical resection in initial and localized cases .

CASE REPORT

- A 68 year old female housewife born and living in Gangotrinagarjaipur came to the otolaryngology ward of SMS MEDICAL COLLEGE AND HOSPITAL JAIPUR complaining of rt nasal bleeding since 2 month and nasal obstruction since 2 month .Initialy bleeding was episodic and minor which was gradually inrease in episode and amount and it is treated by nasal packing and hemocoagulase and other medicine.she said nasal obstruction start before 2 month but progressivly increased and completely obstructed last 1 month
- She had no pain,pt take antihypertensive drug since last 2 month and BP had under controle 120/80 mmhg
- Physical examination revealed normal symmetry and no proptosis ,vision are normal
- The Otolaryngology examination showed-
- Mouth inspection:Modrate quantity of yellowish secretion present without detectable macroscopic alteration
- Anterior rhinoscopy: pinkish polypoidallike mass ,elastic painless filling up nostril(Rt) mild deviation of nasal septum present to the left side,modrate quantity of yellowish secretion present in both nostril,fetid nasal odour.
- Otoscopy:otoscopy of both ear are normal.
- Nasal endoscopy:0 degree telescope pass through left nasal cavity sero-purulent discharge present and right nasal cavity fill-up with mass.
- Biopsy taken from nasal mass and send for histopathology. The biopsy report areundifferetiatedmalignent round cell tumour.Histogenesisare uncertain and marker study are required for exact histogenesis.
- The marker study :The tumour cell express HMB 45,S-100 protein Melan A&EMA (focal) and immunogenative for cytokeratin,synaptophysin and chromogranin A_MALIGNANT MELANOMA
- CECT SCAN PNS:-The ct revealed enlargement of rt.infiror turbinate due to heterogenously enhancing lesion.
- itocculededrt.OMC leading to rt. Maxillary sinusitis and the evidence of sinusitis occures in right half of frontal sinus ,right ethmoid air cells.
- Size of leasion approximate 35x19x22mm.postriorly mass

extending into anterior part of nasopharynx.

- MRI PNS:-Mass lesion (55x41mm)seen in right nasal cavity protruding into the nasopharynx crossing midline reaching left postrior concha -?sqamous cell carcinoma ?melanoma
- We are plan for surgery and done lateral rhinotomy all the mass are excised which was extend into nasopharynx and send for hpx and hpx report show malignant melanoma and pt is follow first time after 15 days at this time pt is normal and she had no complain.

DISCUSSION

The case reported shows initial inespecific symptoms for this disease and the difficulties in obtaining an early diagnosis that would allow better cure possibilities.These are tumors of uncertain etiology and they have no established risk factors or prophylactic measures . These are lesions which are hardly diagnosed in their initial stage because of their intracavitary location, and this allows for obvious and unnoticeable expansion for a considerable period of time. The initial nasal obstruction is subjective and the assessment by the general practitioner is difficult, often times based exclusively on the simple facial X-ray, that only depicts a blurring in the nasal sinuses. This leads the physician to treat the patient as if he/she were having sinusitis, as we have seen also in other nasal diseases such as nasal polyps.

The progress in specialized radiology, most specific with the CT-Scan and MRI and the nasal endoscopy methods have greatly enhanced nasal disorder diagnosis. Notwithstanding, rare entities such as the nasal mucosa melanoma are usually diagnosed already in advanced stages because they grow inside a cavity or because they were insistently treated based on exams such as the facial X-Ray, considered to be too simple in the investigation of nasal obstruction and epistaxis.

It is necesdsary to consider that mucosal melanomas have a distinct behavior when compared to its skin counterpart; those are considered more malignant and of worse prognosis. Radical surgery with broad safety margins makes up the treatment pillar, and it should be indicated whenever the tumors may be resected, together with radio and chemotherapy that, in most cases are only palliative.

Immunotherapy has not proven efficient in this tumor type. Despite all efforts, the attempts to control this neoplasm have not proven successful. Despite promising genetic research progress, the rareness, high malignancy trait and difficulty in diagnosis put the nasal mucosa melanoma at the top of disease severities list; therapeutic frustrations generate generalized pessimism when such disease is diagnosed.

RAVID and ESTEEVES[1] (1960) reported that LUCKE, in 1869, performed surgery to remove a melanocytic sarcoma in the nasal mucosa of a 52 year old man. In the American literature, the first case of nasal melanoma was described by LINCOLN in 1885.

Few years ago, a new therapy was described as yielding benefits in the treatment of nasal mucosa melanomas. SEO et al[9] (1997) reported 3 cases of this neoplasm in which hormonal chemotherapy led to a favorable clinical outcome. They used Tamoxifen, an anti-estrogen chemotherapeutic agent that competes for the estrogen receptor. Although its action mechanism in mucosal melanomas has not yet been ascertainned, the authors believe that this may prove to be one future option in the therapeutic approach to these lesions.

STAMMBERGER et al[10] (1999) evaluated the possibilities and limitations of the endoscopic nasal surgery in the treatment of malignant lesions. In the case of nose and nasal sinuses melanomas, 5 patients underwent endonasal surgery and 2 died between 5 and 14 months after surgery (patients in advanced stage and remote metastasis - T4), and the most survival was of 34 months in one patient with local recurrence.

In a similar way, LUND et al[11] (1999) made a retrospective analysis of 58 individuals diagnosed with nasal mucosa melanoma, followed at the Institute of Laryngology and Otology (London, England) from 1963 to 1996. The patients received surgery alone, surgery with radiotherapy, with or without chemotherapy, radiotherapy or chemotherapy alone. The authors did not see any improvement as far as survival rates are concerned, regardless of the therapy method employed, being single or combined.

In a retrospective study, RICHTIG et al[12] (2002) reviewed data from 9 nasal mucosa primary melanoma patients, followed in Osterreich (Germany), from 1985 to 2000, and concluded that this is a highly malignant tumor of unknown cure. The authors stressed that even with surgical treatment and radiotherapy, most of the patients would still present with local recurrence and remote metastasis.

PRASAD et al[14] (2003) assessed 95 patients with a diagnosis of primary mucosal melanoma; of those, 36 were primary of the oral mucosa (stratified epithelium) and 59 of the nasal mucosa (pseudostratified epithelium). They showed that the latter presented a more aggressive histological pattern than the former, being characterized by a larger vascular proliferation and tissue invasion. However, in both cases, the average patient survival was similar (2.8 years for nasal melanoma and 3 years for the oral melanoma).

CONCLUSIONS

Primary malignant nasal mucosa melanomas are very rare tumors and, due to their specific location, are of hard diagnosis in its initial stages. Of uncertain etiology and risk factors, these are highly recurrent tumors, both locally as well as remotely, and it spreads in the mucosa compromising total and effective surgical resection. Thus, it bears bad prognosis, and cure possibilities are based on the surgical treatment of early diagnosed cases.

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