

Mucosal malignant melanoma of sinonasal : is it necessary to treat the neck?

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Abstract

Primary mucosal melanoma of the paranasal sinuses is a rare tumor of the head and neck. Malignant melanoma had two type; mucosal and cutaneous malignant melanoma. Lymph node of paranasal is drainage to submandibular and Jugulo-omohyoid lymph nodes. This case report is represent to remind us about the important of neck management for malignant melanoma sinonasal with N0. We did the tumour extirpation with nasoendoscopic approach and supra omohyoid selective neck dissection.

Key word: *malignant melanoma sinonasal, lymph node metastasis, neck dissection*

Abstrak

Melanoma mukosa primer dari sinus paranasal merupakan tumor langka kepala dan leher. Melanoma maligna mempunyai dua tipe; melanoma maligna tipe mukosa dan tipe kutan. Drainase kelenjar limfe dari sinus paranasal mengalir ke kelenjar limfe submandibular dan jugulo-omohioid. Makalah ini dibuat untuk mengingatkan kembali mengenai pentingnya diseksi leher pada melanoma maligna sinonasal dengan N0. Kami lakukan ekstirpasi tumor dengan pendekatan nasoendoskopi dan diseksi leher selektif supra omohyoid.

Kata kunci: melanoma maligna sinonasal, metastasis kelenjar getah bening, diseksi leher.

Introduction

Primary mucosal melanoma of the paranasal sinuses is a rare tumor of the head and neck which can be a devastating disease with poor outcome. Because most of the series extend retrospectively several decades, we sought to determine

prognostic factors and outcomes with recent treatment modalities. Melanomas of the sinonasal tract are infrequent and account for less than 1% of all melanomas and up to 4% of all sinonasal malignancies. The rarely of this tumor is because of its origin from melanocytes that have

migrated as neuroectodermal derivatives in the ectodermal mucosa. Melanomas originating from the respiratory mucosa and those originating from the squamous mucosa have different clinical and histopathological features, but share a similar prognosis. The most common sites for the development of mucosal melanoma are the nasal cavity and paranasal sinuses.^{1,2,3}

Tash and Keskhin⁹ found that face was the most common anatomic site of cutaneous (50,53%) which was followed by scalp (26,28%) and ear region (18,19%). The most common histological subtypes of sinonasal mucosal melanoma include spindle cell, epitheloid and pleomorphic.^{9,13}

The aggressiveness of mucosal melanoma may be explained by its late presentation and delayed diagnosis, the vascularity of the mucous membranes, which promotes hematogenous metastases, or by cellular and molecular differences that have been shown to exist between cutaneous and mucosal melanoma. The regional metastasis of mucosal malignant melanoma of sinonasal is according to sinonasal lymph node drainage to the neck. Therefore is important for ENT

doctor do neck dissection for mucosal malignant melanoma of sinonasal treatment.⁴

Table 1. Comparison of cutaneous and mucosal melanoma.⁵

	Cutaneous melanoma	Mucosal melanoma
Tissue of origin	Skin	Mucosal surfaces
Mean age at presentation	55years	67years
Staging	American Joint comitee on Cancer staging applicable	American Joint comitee on Cancer staging applicable
Presentation	Less than one third present with advanced disease	>50% present with advanced disease (metastasis)
Amelanotic appearance	1,8%-8,1%	20%-25%
Risk factors	Sun exposure	Unknown
Race	White 94%; Black 0,8%	White 85%
Regional lymph node metastasis	5%	32% (head and neck)

Epidemiology and presentation

Primary mucosal malignant melanoma of the nasal cavity, paranasal sinuses and nasopharynx is rare, accounting for between 0,3% and 2% of all malignant melanomas and about 4% of head and neck melanomas. Overall melanomas account for 3,5%-7% of all sinonasal neoplasms. Melanoma is malignancy of ectodermal origin arising from

melanocytes which are found in respiratory mucosa. There are clinical and histopathological differences between melanomas originating from the sinonasal respiratory mucosa and those that originate from the oral squamous mucosa. Sinonasal melanomas have been described to have a lower rate of regional lymph node metastasis and the presentation of the tumour are commonly present ulceration, necrosis, perineural invasion, polypoid morphology, and a pseudopapillary growth pattern. Despite these well-documented differences, the prognosis is comparable to melanomas originating from squamous oral cavity mucosa. The most frequent sites of noncutaneous melanoma are the eye (5,3%), melanoma of unknown origin (2,2%) and then mucosal melanoma.^{4,5,6}

Ciptomangunkusumo hospital in 2013 had 4 case of sinonasal malignant melanoma and 2 case in 2014. Microscopically, sinonasal mucosal melanoma (SNMM) is another “small blue cell tumor” with similarities to lymphoma, rhabdomyosarcoma, plasmacytoma, olfactory neuroblastoma and poorly differentiated carcinoma. The most

common histological subtypes of sinonasal melanoma include spindle cell, epitheloid and pleomorphic. Regan et al⁵ found that regional lymph node metastasis was 32%.^{4,5,6}

Table 2. Patient Demographics for Mucosal Melanoma of the Head and Neck.³

Characteristic	Number	Presentation
Sex		
Male	133	43.8
Female	171	56.3
Race		
White	276	90.8
Black American Indian/Alaska Native	13	4.3
Asian or Pacific Islander		
Age, yr	0	0.0
0–49		
50–64	15	4.9
65–74	24	7.9
75	61	20.1
	69	22.7
Site		
Nasal cavity		
Maxillary sinus	150	49.3
Ethmoid sinus	199	65.5
Accessory sinuses	46	15.1
	27	8.9
	32	10.5

Histological appearance

This is the histopathology figures of malignant melanoma.⁶

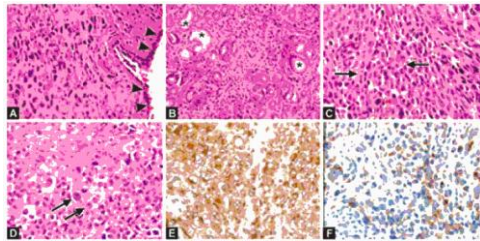


Figure 1. A to F: Mucosal malignant melanoma (A) partially denuded respiratory epithelium (arrowheads) with underlying malignant spindle cells exhibiting marked cytologic atypia (H and E stain: original magnification , 400x).(B) these cells infiltrate deep into the submucosa and surround the seromucous gland (asterisks) (H and E stain; original magnification 200x). (C) Scattered malignant cells demonstrate intranuclear inclusion (arrows) (H and E stain, original magnification 400x). (D) in some areas, the malignant cells appear epithelioid with prominent macronucleoli (arrows). (E) HMB 45. (F) MART 1 imm exhibit strong, diffuse, cytoplasmic positivity within malignant cells(original magnification, 400x).⁶

Staging

In contrast to cutaneous melanomas, there widely classification and staging system and evidence-based treatment concept specifically

focused on SNMM. Ballantyne⁷ describe a simplified staging system for head and neck mucosal melanomas in 1970, which continues to be the most widely utilized. This system classifies tumors in the following three stages: stage I for localized lesions; stage II for cervical lymph node metastasis and stage III for distant metastasis. The advantage of this system are its simplicity and the fact that it can be used for oral and mucosal primaries. Unfortunately, since the vast majority of patient present with local disease only (stage I), the prognostic value of this staging system is relatively limited and it appears that the current AJCC sinonasal staging system to be useful.^{6,7}

The 7th edition of the American Joint Committee on Cancer (AJCC) staging manual contains an entirely new chapter focusing on the staging of mucosal melanoma of the head and neck. The utility of this primarily arises from the difficulty of staging melanoma, essentially a cutaneous disease, using the classification schemes for mucosal head and neck cancer. The difficulty arises from the fact that the former is almost exclusively tumor thickness specific, and the latter is a combination of

tumor size and tumor site. What is less clear is whether the changes in the classification of head and neck mucosal melanoma allow for the useful delineation of survival by staging group. The American Joint Committee on Cancer (AJCC) in 2013 made staging of mucosal melanoma of head and neck in this table below.³

Table 3. NCNN staging of mucosal melanoma.⁸

American Joint Committee on Cancer (AJCC) TNM Staging System for Mucosal Melanoma of the Head and Neck (7th ed., 2010)		Anatomic Stage/Prognostic Groups			
Primary Tumor (T)		Stage III	T3	N0	M0
T3	Mucosal disease	Stage IVA	T4a	N0	M0
T4a	Moderately advanced disease Tumor involving deep soft tissue, cartilage, bone, or overlying skin		T3-T4a	N1	M0
T4b	Very advanced disease Tumor involving brain, dura, skull base, lower cranial nerves (IX, X, XI, XII), masticator space, carotid artery, prevertebral space, or mediastinal structures	Stage IVB	T4b	Any N	M0
		Stage IVC	Any T	Any N	M1
Regional Lymph Nodes (N)		Histologic Grade (G)			
NX	Regional lymph nodes cannot be assessed	GX	Grade cannot be assessed		
N0	No regional lymph node metastases	G1	Well differentiated		
N1	Regional lymph node metastases present	G2	Moderately differentiated		
		G3	Poorly differentiated		
		G4	Undifferentiated		
Distant Metastasis (M)					
M0	No distant metastasis				
M1	Distant metastasis				

Survival

The 5-year overall survival range from 20% to 35% among different series. Recently, Lund et al⁶ reported 28% and 20% 5-year and 10-year overall survival rate respectively, with a 21-month median survival. After a mean follow-up of almost 14 years, 35% of the patients in the study were alive or died without evidence of disease.⁶

Tash and keshkin⁹ found an

estimated 1- and 5-year overall survival rates in 41 Sinonasal Mucosal Melanoma (SNMM) patients were 81% and 58%, respectively. The 1- and 5-year survival rates were 84% and 64% for patients with sinonasal and 79% and 53% for oral cavity. Advanced disease (stage II and III) at presentation was the significant prognostic factor for outcome, age and gender of SNMM patients did not affect survival.⁹

Lester et al¹⁰ found that 40 patients of SNMM who were without evidence of disease at last follow-up had a mean follow-up of 13.9 years. These results yield a raw 5-year survival of 42.6% and a raw 10-year survival of 24.3%. This contrasts to a disease free 5-year survival of 31.3% and a disease-free 10-year survival of 22.6%. Cheng et al¹⁰ found the mean 5-year overall survival rate was only 15.65%.¹⁰

Lymph node

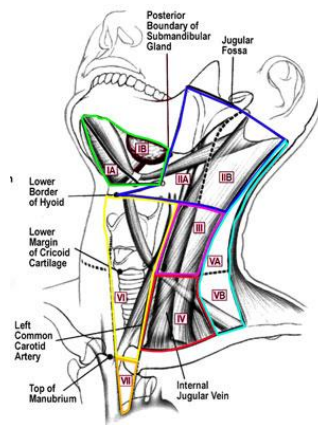


Figure 2: Level of the cervical lymph node.¹¹

The American Academy of Otolaryngology–Head and Neck Surgery has modified the system used to assign level by subdividing levels I, II and V into A and B designation for each levels. Level I divide into level IA and IB, the submental group of level IA include is the lymph nodes between the anterior belly of the digastric muscle and cephalad to the hyoid bone. The submandibular group of level IB includes lymph nodes in the triangular area bounded by the anterior and posterior bellies of the digastric muscle and the inferior border of the body of the mandible also the lymph nodes adjacent to the mandibular salivary gland and along the facial artery.

Level II is the midjugular group, which includes lymph nodes around the upper portion of the internal jugular vein and the upper part of the spinal accessory nerve, extending from the base of the skull up to the bifurcation of the carotid artery or the hyoid bone. The posterior limit for this level is the posterior border of the sternocleidomastoid muscle, and the anterior border is the lateral limit of the sternohyoid muscle. Lymph nodes anterior the spinal accessory nerve are designated level IIA, and those posterior to it are designated level IIB.¹¹

Level III is the midjugular group, which includes lymph nodes around the middle third of the internal jugular vein from the hyoid bone up to the inferior border of cricoid cartilage. The anterior and posterior borders are the same as those for level II. Level IV is the lower jugular group, which includes lymph nodes around the lower portion of the spinal accessory nerve and along the transverse cervical vessels. It is bounded by triangle formed by the clavicle, the posterior border of the sternocleidomastoid muscle, and the anterior border of the trapezius muscle. Level V is divided into two

levels by a plane at the level of the inferior border of the cricoid cartilage. Level VA is superior to this plane, and level VB is inferior to it. Level VI is the central compartment group, which includes lymph nodes in the prelaryngeal, pretracheal, (Delphian), paratracheal and tracheoesophageal groove. The boundaries are the hyoid bone to the suprasternal notch and between the medial borders of the carotid sheaths. Level VII is the superior mediastinal group, which includes lymph nodes in the anterosuperior mediastinum and tracheoesophageal grooves, extending from the suprasternal notch to the innominate artery.¹¹

Lymph node drainage of head and neck

The Lymph drainage of the head & neck tends to follow the general rule of Superficial lymphatic with the vein and deep lymphatic with the arteries. The Lymph nodes of the head & neck arranged in three chains; 1) Superficial circular chain: superficial nodes extend from the chin to occiput i.e. around the junction of the head & neck; 2) Vertical chain: “ deep cervical nodes” arranged along the internal jugular vein from digastric to the root of the neck; 3) Deep circular

chain: deep lymph nodes, surround the upper air and alimentary passages.¹¹

Superficial circular nodes are

- 1) Submental nodes is lymph node that lies in the superficial fascia of submental triangle and drain wedge of tissue corresponds to the premaxilla, it receives afferents from both sides of the face and the efferent lymph vessels will drain to the submandibular lymph nodes or pass directly to Jugulo-omohyoid lymph nodes;
- 2) Submandibular Lymph nodes is lymph node that lies beneath the deep fascia on the surface of the submandibular salivary gland, it receive afferents from the submental lymph nodes and wide area extend from the center of the forehead, nose, cheek, upper lip, the anterior 2/3 of the tongue, floor of the mouth the upper teeth and gums also, It drains also frontal, maxillary and ethmoidal air sinuses. The efferent drain into the Jugulo-omohyoid lymph nodes;
- 3) Buccal or mandibular node is small node lies isolated on the buccinator muscle & lower border of the mandible anterior to the masseter, it receives afferents from part of the cheek & lower eyelid. The efferent passes to antero- superior group of deep

cervical nodes; 4) Preauricular nodes (parotid) is lymph node that lies on & within the parotid gland. Some of these lymph nodes are located around the external jugular vein. It receives afferents from the temple, vertex, eyelids, orbit, & external acoustic meatus. The efferent passes to the deep cervical lymph nodes; 5) Occipital nodes are few nodes lie at the apex of posterior triangle & mastoid process. It receives afferents from the posterior part of the scalp & auricle. The efferent passes to the supraclavicular lymph nodes. Lymphatic drainage of the face are the central part i.e. the chin and tongue tip drain to submental lymph nodes and wedge of tissue above includes central forehead and frontal sinuses, anterior half of the nose & maxillary sinuses, side of the tongue and floor of the mouth drain into submandibular nodes. Lateral part of the forehead, temple, orbital contents & cheek drain to the preauricular group.¹¹

The deep circular lymph nodes are lymph node that surround the larynx, trachea & pharynx. It is a collection of scattered lymph nodes. Classified as: 1. Pretracheal nodes: drain part of lower larynx, trachea & thyroid isthmus; 2. Retropharyngeal nodes: drain the

soft palate, posterior part of hard palate, nose & pharynx; 3. Paratracheal. The vertical chain arranged around the whole length of internal jugular vein. The nodes are arranged in demonstrated groups, superior and inferior, anterior and posterior. From these groups two nodes are well known: 1) The Jugulo digastric that below the posterior belly of digastric, between the angle of the mandible and anterior border of Sternomastoid, it is a member of antero-superior group; 2) Jugulo omohyoid is lies above the inferior belly of Omohyoid, behind the jugular vein under cover of sternomastoid, it is a member of postero-inferior group.¹¹

Mucosal Melanoma regional lymph node metastasis

Regional lymph node metastasis was defined as histologically proven disease in the primary draining lymph node basin for each primary site. These sites included cervical nodes (sinonasal primary), pelvic nodes (vaginal and rectal primary) and inguinal nodes (vaginal and anal primary). In patients without regional lymph node metastasis at presentation, the time to development of regional lymph node involvement was recorded, in other research by Regan et al⁵ found that

regional lymph node metastasis of Head and Neck Mucosal Melanoma was 32%.⁵

The biological behavior of mucosal melanoma is not well understood and explanations for its more aggressive behavior have been proposed, including delays in clinical presentation and diagnosis, the rich lymphatic and vascular supply of the mucosal surfaces, and an inherent more aggressive biology.⁴

Distant Metastasis

O'regan et al⁵ research about distant metastasis of mucosal melanoma, they found that the commonest initial site of distant metastasis was the lungs (66%), followed by the liver (50%) and peritoneum (33%). Peritoneal metastasis was more common in patients with vaginal melanoma. Sinonasal melanoma metastasized to the liver in all cases and peritoneal disease was seen in 50% of patients. No CNS metastasis was seen in patients with sinonasal primaries. Metastasis to the musculoskeletal soft tissues (e.g. skin and muscle) was also infrequent (16%). Regional lymph node metastasis preceded distant metastatic disease in 33% of patients, with a median time interval from

lymph node involvement to distant metastasis of 13 months (range 4-36 months).⁵

Imaging Findings

CT appearance of sinonasal melanomas is nonspecific. Density values and enhancement pattern do not provide key information. Bone destruction is observed in most malignant tumors. The site of origin may offer more substantial clues for the differential diagnosis whenever the lesion arises from the anterior nasal septum or middle/inferior turbinate. MRI (Magnetic Resonance Imaging) findings largely depend upon the histological features of the lesion. Melanotic melanomas exhibit a peculiar pattern - consisting of hypo intense signal on T2 and spontaneous hyperintensity on T1 – as the result of paramagnetic properties of melanin. More in detail, paramagnetic effect could be due either to metal ions bound to melanin, or to free radicals formation. Conversely, amelanotic variant displays a less specific pattern: hyper intense on T2, hypo intense on plain T1. MRI findings of amelanotic melanoma are nonspecific as they are shared by a longer and wider list of different

lesions, including squamous cell carcinoma, adenocarcinoma, minor salivary glands neoplasms, olfactory neuroblastoma, and fibro-osseous lesions.¹²

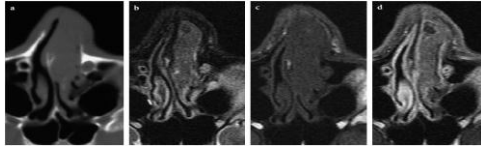


Figure 2. Melanoma of the left nasal cavity. Axial CT (a), TSE T2 (b), unenhanced (c), and enhanced (d) T1. The tumor presents as a polypoid mass, filling the nasal cavity, without specific signal features.¹²

Diagnosis

The diagnosis of mucosal melanoma is usually made on the basis of histology and immune histochemistry. Radiologic evaluation is important for the purposes of staging, operative planning and monitoring of patients with metastatic disease undergoing systemic treatment. Computed tomography (CT) and Positron Emission Tomography (PET)/CT are of relatively limited value in local disease evaluation but are primarily used to detect clinically unsuspected metastatic disease. The role of fluorodeoxyglucose (FDG)-PET/CT in cutaneous melanoma management is well established. Its role in mucosal melanoma is less well

investigated, but given the high metabolic activity of these tumors, it is likely to provide similar staging information but this requires validation in large-scale trials.⁴

The NCNN guidelines madeguidelines for diagnosis of mucosal malignant melanoma such as complete head and neck examination, mirror and fiberoptic examination as clinically indicated, verification of pathology using appropriate staining (HMB-45, S-100, Melan-A), CT-scan and/or MRI to determine anatomic extent of disease, particularly for sinus disease, chest imaging as indicated and consider PET-CT scan to rule out metastatic disease. Histologic diagnosis was confirmed in all cases by means of fine-needle aspirate, sentinel node biopsy, and/or surgical lymph node resection. In patients without regional lymph node metastasis at presentation, the time to development of regional lymph node involvement had to be recorded.⁸

Treatment

All patients were treated by partial or complete surgical excision; complete surgical removal of the tumor was not always possible as a result of the complex anatomy of the nasal cavity,

paranasal sinuses, and nasopharynx. NCNN 2013 made guidelines for mucosal melanoma of paranasal, stage III treated by wide surgical resection of primary tumour and strongly consider postoperative radiotherapy to primary site. T4a with No treated by wide surgical resection and postoperative radiotherapy to primary site. T3-T4a with N1 treated by wide surgical resection and neck dissection of positive neck also postoperative radiotherapy to primary site and neck. Stage IVB treated by primary radiotherapy or systemic therapy, the last is stage IV C treated by best supportive care or primary radiotherapy or systemic therapy.

Lester et al¹⁰ research to compare the succeed between surgery alone and with radiotherapy or chemotherapy, specific type of therapy did not seem to influence the overall patient outcome, as there was no statistically significant difference between patients managed by surgery alone, surgery with chemotherapy, surgery with radiation therapy, or surgery and combination therapy.^{8,10}

Complete tumor excision is widely accepted as the standard of care for

treatment of patients with mucosal melanomas of the sinonasal tract. The anatomical complexity of the sinonasal passages and proximity to vital structures makes a complete resection difficult in most cases. Even more, radical procedures do not appear to be justified when there is evidence suggesting that >50% of the patients who achieve local control with surgery will ultimately develop distant metastasis. In this context, the surgeon's experience and thorough knowledge of the anatomy are key to achieve a successful oncologic resection with a minimum cosmetic and functional impact.^{8,10}

Tash and Keshkin⁹ research found that all the patient except 2 patients underwent surgery with curative intent; the type of surgery and surgical approach were based on the location and extension of the tumor. Patients who underwent endoscopic resection had better survival. Surgical margins were described as microscopically positive or close (within 1 mm) in 20% of patients. Bachar et al⁹ has reported where margin status did not affect overall survival. In contrast, Penel et al⁹ has reported recently that margin status affects overall survival. The potential

benefits of negative margins in terms of local control and overall outcome may be concealed by the development of distant metastasis in a significant number of patients with a profound negative impact on survival.⁹

Neck Treatment

Moreno and Hanna¹³ found elective treatment of the neck is usually not performed, as the incidence of nodal disease at the time of presentation is relatively low, ranging from 6 to 25%. However, the incidence of nodal disease is higher in oral cavity than in sinonasal melanomas, both at presentation and during the course of the disease. Medina et al¹³ has recommended elective treatment of the neck in patients with melanomas of oral origin, although most authors would still recommend a conservative approach. In a recent report of 74 patients treated for Head and Neck Mucosal Melanoma (HNMM), Krenkli et al¹³ described a 77% regional recurrence rate for HNMM arising in the oral cavity, suggesting a potential advantage for prophylactic neck treatment in this subset of patients.¹³

Selective Neck dissections (SND)

The SND consist of the removal of

only the lymph node groups at the highest risk containing metastases according to the location of the primary tumor, preserving the spinal accessory nerve, the IJV, and the sternocleidomastoid muscle. There are four main types of SND such as; 1) SND of Level I to II (supraomohyoid neck dissection) commonly used in the treatment of patients with squamous cell carcinoma of oral cavity. The lymph node that removed are in the submental and submandibular triangle (level I), the upper jugular region (level II) and the midjugular region (level III). The posterior limit is cutaneous branches of the cervical plexus and the posterior border of the sternocleidomastoid muscle, the inferior limit is the omohyoid muscle as it crosses the IJV. 2) SND of level II (Lateral neck dissection). Commonly used in the treatment of patients with squamous cell carcinoma of the larynx, oropharynx and hypopharynx. Removed of the upper (level II), middle (level III) and lower (level IV) jugular lymph nodes. The superior limit is the digastric muscle and the mastoid tip. The inferior limit is clavicle. The posterior limit is cutaneous branches of the cervical plexus and the posterior border of the

sternocleidomastoid muscle. 3) SND of level VI (anterior or central compartment) is used for patient with cancer of the midline structures of the anterior inferior aspect of the neck and thoracic inlet such as the pyriform sinus and the cervical esophagus and trachea. Removal of the prelaryngeal, pretracheal and paratracheal bilateral lymph node. 4) SND for cutaneous malignancies of the head and neck. The extent of the regional node dissection in patient with cutaneous malignancies depends on location of primary lesion. Skin cancers originating from the posterior scalp and the upper-lateral aspect of the neck commonly used SND levels II-V, retroauricular, suboccipital (posterolateral neck dissection). The superior limit of this dissection is the posterior belly of the digastric muscle and the mastoid tip anterior laterally and the nuchal line/ridge posteriorly. The inferior limit is the clavicle. The anterior-medial is the lateral border of the sternohyoid muscle. The posterolateral limit of the dissection is marked by the anterior border of the trapezius muscle inferiorly and the posterior midline of the neck superiorly. The regional node dissection often performed for cutaneous malignancies originating

from the periauricular skin, anterior scalp and temporal region is a SND (parotid, facial and external jugular nodes, level II, III, VA).¹⁴

Radiation Therapy.

The available evidence from radiation postoperative studies suggests that the use of adjuvant radiation therapy improves locoregional control, although there is no evidence of benefit in overall survival. Some authors report the use of postoperative radiation more commonly in sinonasal versus oral cavity melanomas, probably because of the inherent difficulty of obtaining negative margins in the nasal cavity. To date, there is no consensus regarding the indications for postoperative radiation therapy, although most authors agree regarding its use in patients with positive and close margins, especially as these have been recently identified as negative prognostic factors.²

Mereno et al² study find the use of postoperative radiation improved locoregional control but only when a total dose greater than 54 Gy was used. It is difficult to determine whether the lack of improvement for the group that received a lower dose was determined by total

radiotherapy (RT) dose, hypofractionation, or a combination of both. In contrast with these findings, other authors have reported that hypofractionation might improve local control and overall survival in head and neck mucosal melanomas. The optimal RT dose and fractionation schedule for mucosal melanomas of the sinonasal tract remains undetermined.²

NCNN made guidelines for radiation therapy, radiotherapy for unresectable locally advanced melanoma treated by 66 Gy to 72 Gy and palliative radiotherapy dose and schedule may be considered. Post operative (after primary site resection) got radiotherapy to primary site +2-3cm margins or to anatomic compartment. Primary and neck melanoma (high-risk site) got 60-66Gy or 70Gy for gross disease, if the melanoma was low risk, undissected or uninvolved portions of neck got 50-6-Gy.⁸

Follow Up

NCNN made guidelines for follow up mucosal melanoma; head and neck examination every 1-3 months in first year, every 2-6 months in second year, every 4-8 months year 3-5 and every 12 months after year 5.

Post-treatment baseline imaging of primary (and neck if treated) recommended within 6 months of treatment.⁸

CASE

Patient male, 62 years old, with bleeding from right nasal cavity since 4 months prior to admission. He also complain about right nasal blockage, bloody discharge, post nasal drip, headache and smell disturbance on the right nose. Patient went to public hospital in Bogor and referred to ENT oncology division outpatient clinic of Ciptomangunkusumo Hospital. Physical examination within normal limit. Nasoendoscopy examination from the right nasal cavity showed a narrow nasal cavity, there was mass covered nasal cavity until 1/3 anterior nasal cavity, craggy and bleed easily, inferior turbinate, middle meatus and middle turbinate could not be evaluated. Nasoendoscopy of the left nasal cavity showed a wide nasal cavity, inferior turbinate was eutrophy, middle turbinate was eutrophy, the middle meatus was open, there was deviated septum at 1/3 middle nasal cavity and there were no mass at the nasopharynx.

CT-scan revealed a mass suggestive malignant that involve the inferior turbinate and spread to right maxillary sinus, there were also had veiled at bilateral ethmoid sinus suspicious sinusitis. Biopsy result was malignant melanoma nodular type. Patient then diagnosed with malignant melanoma T3N0M0. The patient had already been discussed in tumor meeting of ENT and plan for re-evaluation of biopsy result. The review of biopsy results was malignant melanoma nodular type and plan for mass extirpation with nasoendoscopy approach and selective neck dissection.

Patient was hospitalized one day before the surgery, Cefazolin 2grams as a prophylaxis antibiotic and methylprednisolone 250mg once daily also omeprazole 40mg once daily was given. Mass extirpation and concotomy with nasoendoscopic approach and selective radical neck dissection was performed under general anesthesia. Lidocaine and adrenaline tampon was applied in both nasal cavities. Evaluation of the right nasal cavity, showed that right nasal cavity full of mass until 1/3 anterior nasal cavity, craggy and bleed easily which attach to middle part of inferior

turbinate, 1/3 anterior until 1/3 middle of nasal septum and inferior part of middle turbinate also lateral nasal wall. Mass attachment at inferior turbinate and attachment at inferior part of middle turbinate was released with suction cauter and bipolar cauter. The sphenopalatine foramen was infiltration with lidocain and adrenaline continued with cauter the sphenopalatine artery. The left side of nasal septum was infiltration with lidocain and adrenaline continued with hemitransfiction incision and elevation of mucopericondrium until nasal base, lateral nasal wall and posterior part of posterior fontanel continued with incision of mucoperiosteal flap. The spongostan was applied at nasal base, septum and lateral nasal wall also at sphenopalatine foramen. The whole tumour mass was extirpated from the nose. Net cell was applied at right nasal cavity.

The operation was continued with neck dissection. Site marking was from region submental along the mandible until infra auricular area at the point below the mastoid process. The incision was done at the site marking area until platisma muscle was seen. Platisma was aside until

mylohyoid and sternocleidomastoid muscle was seen. Lymph node at level I,II and III was identified and separated from its surrounding structures. The Marginal Mandibularis and Accesorius nerve has been preserved after that all of the lymph node level I, II and III was excluded from the neck. The patient got ceftriaxone 1mg twice daily as antibiotic post operative, he also got ranitidine 50mg twice daily, tranexamic acid 500mg three times daily and ketorolac 30mg three times daily. Two day after the surgery the patient hemoglobin was 8,9 g/dL and has been PRC transfusion 250cc and the hemoglobin increase become 11,0 g/dL. One week after surgery the wound was healed and patient was discharge from Cipto Mangunkusumo hospital.

Follow up at outpatient clinic one week post surgery, from nasoendoscopy examination, there was no inferior and middle turbinate and there were crusting covered the middle meatus. The right colli region the wound was healed and there was no pus, all the stitches was removed. We gave Cefixime 200mg twice daily, paracetamol plus tramadol three times daily, nasal wash 30cc

twice daily and mometashone furoate nasal spray 2 puff twice daily was prescribed. One week after that the patient came for follow up, from nasoendoscopic examination there was still crusting covered the middle meatus and the uncinate process. there was serous post nasal drip, nasal wash was continued. Result from the histopathology the tumor was malignant melanoma nodular type and there was no locoregional lymph node metastasis. Follow up two months post operation there was recurrent mass at the left nasal cavity that craggy and bleed easily.

Discussion

The subject of this case was a male patient 62 years old with bleeding from right nose since 4 months prior to admission, right nasal blockage, bloody discharge, post nasal drip, headache and smell disturbance on the right nasal cavity. The nasoendoscopic examination from the right nasal cavity showed a narrow nasal cavity, there was mass covering nasal cavity until 1/3 anterior nasal cavity, craggy and bleeding easily, inferior turbinate, middle meatus and middle turbinate could not be evaluated. This patient had mucosal malignant melanoma at sinonasal that is the most common

site of MM at the head and neck. The patient's age was 62 years old that included in the third highest presentation group of age. This case showed the tumor was on the right nasal cavity originating from mucosal surface of the nasal cavity and was in accordance with mucosal malignant melanoma type.^{2,15}

The result from the histopathology was malignant melanoma nodular type and there was no locoregional lymph node metastasis and assessed with Melanoma malignant T3N0M0. Vijay et al said that mucosal melanoma had 3 subtypes including spindle cell, epitheloid and pleomorphic. This was not in accordance with the result of histopathology of this patient. The result was malignant melanoma nodular type. Malignant melanoma nodular type is in accordance with cutaneous malignant melanoma subtype.⁶

NCNN 2013⁸ made guidelines for mucosal melanoma of paranasal, stage III was treated by wide surgical resection of primary tumor and strongly considers postoperative radiotherapy to primary site.⁸

Mucosal malignant melanoma

patients was treated by partial or complete surgical excision; complete surgical removal of the tumor was not always possible as a result of the complex anatomy of the nasal cavity, paranasal sinuses, and nasopharynx so that we could not have free margin to eradicate complete removal.

This patient had selective neck dissection with Supraomohyoid neck dissection which was dissection of the neck that removed lymph node at level I, II and III in accordance with the flow of the lymph node from sinonasal area. The supraomohyoid neck dissection was performed for prevention to cervical lymph node metastasis. Elective treatment of the neck was usually not performed although from literature there were still debating about whether it was necessary for the neck dissection of mucosal malignant melanoma.¹¹

Before the neck dissection we did nasoendoscopic approach with the sentripetal dissection that dissection was find the outer border of the tumor, after found all the border and separated it from its surroundings structure, we elevated the tumor and release whole tumor from its surroundings structure. Intra

operative we found that the tumor origin form inferior turbinate and we did the concotomy. We did the nasoscopy. We did the nasoscopy because the advantages from nasoscopy approach was the margins were one of the strongest predictors of disease control, and thus this remained a critical factor in the assessment of evolving techniques. Advances in tumor imaging, surgical techniques, appropriate patient selection and the ability to apply combined endoscopic/craniofacial approaches to difficult cases. An endoscopic approach should be considered for tumors which occupies the central nasal cavity between the frontal and sphenoidal sinuses but do not extend to the lateral lamella of the pterygoid bone.^{5,16}

Tumor invasion of the nasal bones, anterior/posterior table of the frontal sinus or frank orbital invasion are considered contraindications. Posteriorly, it is important to assess invasion of the carotid & cavernous sinus. If tumor is noted tracking along nerves (most importantly trigeminal), these are also relative contraindications to an endoscopic approach. Malignant tumor types that have been resected with favourable results include adenocarcinoma,

adenoid cystic carcinoma, chordoma, malignant melanoma, olfactory neuroblastoma, osteosarcoma and squamous cell carcinoma. The most common serious complication of nasoscopy approach is CSF leak. Tash and Keshkin⁹ found patients who underwent endoscopic resection had better survival.^{5,9,15,16}

In this patient we did the CT-Scan because CT-Scan represented the best modality with which to assess for the presence of bony remodelling or bony invasion, for example of the sinus walls, orbital margins, and the floor of the anterior cranial fossa. We wanted to know the spreading of the tumor that occurred in the sinus wall and metastatic disease.^{5,16}

This patient didn't get the radiotherapy because according to Lester et al¹⁰ that the specific type of therapy did not seem to influence the overall patient outcome, as there was no statistically significant difference between patients managed by surgery alone, surgery with chemotherapy, surgery with radiation therapy, or surgery and combination therapy.

Two months follow up at this patient post operation we found that this

disease was recurrent (reoccurred) and plan for maxillectomy. We should evaluate the neck area whether there was lymph node enlargement during follow up and the Ct-Scan examination.^{5,8}

The conclusion of this case was that we had to do the neck surgery although we didn't find any regional lymph node metastasis.

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