-대한 구강 : 악안면방사선학회지 : Vol. 23, No. 2, 1993-

PRIMARY MALIGNANT MELANOMA OF THE NOSE AND PARANASAL SINUSES ; REPORT OF A CASE

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CONTENTS

Ⅰ. INTRODUCTION

Malignant melanoma is one of the most deadly of all human neoplasms. By definition, melanomas are malignant tumors originating from melanoblast, which produce melanin pigment or are capable of doing so[1,2]. Melanin is a characteristic feature of melanomas. The amount of melanin may vary considerably, both in the same tumor and between the primary and metastatic tumors. Certain parts of the tumor may contain more pigment than others, and in some forms of melanoma, pigment may be absent[2].

The etiology of malignant melanomas is yet an unsolved problem. A history of traumatic injury or irritation at the local site can be elicited in between 10 % and 60 % of all cases of melanoma[3].

The malignant melanoma may occur as a primary lesion not only on the skin and oral mucous membrane but also in the eye, vaginal mucosa, and upper respiratory tract[4,5]. Primary malignant melanoma of the nasal and paranasal sinuses is extremely rare. Nasal and paranasal melanomas represent 0.6 % to 0.7 % of melanomas in all sites, 2 % to 9 % of melanomas in the head and neck, and 3.6 % to 4 % of all nasal tumors[6].

The mucosal melanomas are often symptomatically silent until very large, leading to poor prognosis when discovered late[7-9].

Primary malignant melanoma of the nose and paranasal sinuses has been reported but rarely. The authors have experienced one case of malignant melanoma originating from the mucosa of the nose and paranasal sinuses. We reviewed the literatures related to the malignant melanoma and report this one case.

II. REPORT OF A CASE

1. Clinical findings

A 36-year-old female was admitted to our hospital with a 2-month history of right facial swelling, pain and pus discharge(Fig. 1). Clinically, there was a nasal stuffiness. Intraoral examination revealed the incisional wound and black discoloration of mucobuccal fold of the upper right molar area(Fig. 2), and the tenderness to palpation at the mass was elicited.

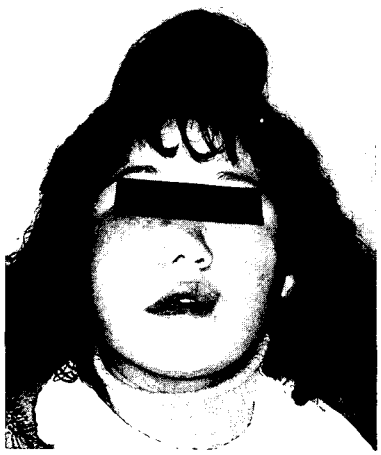

Fig. 1. Facial photograph shows marked right facial swelling.

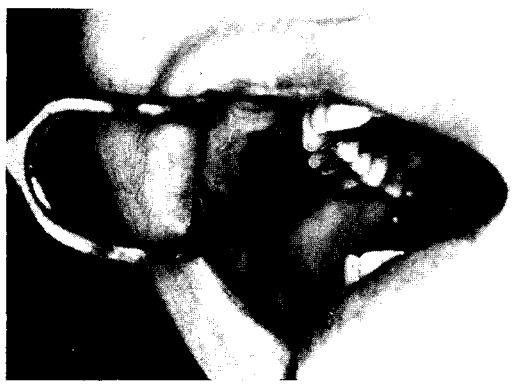

Fig. 2. Intraoral photograph shows incisional wound and black discoloration of mucobuccal fold of upper right molar area.

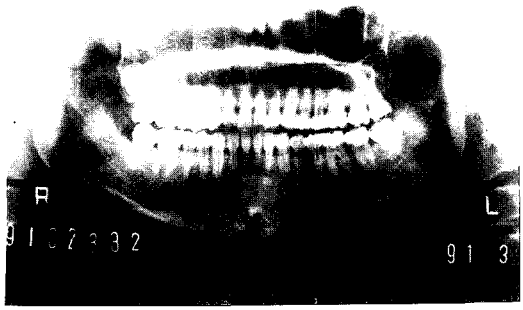

Fig 3. Pantomogram shows an aggressive bone destruction of floor of right maxillary sinus and lateral wall of nasal cavity.

Fig. 4. Mandible P-A view shows destruction of anterior wall of maxillary sinus.

Fig. 5. Waters' view shows sinus opacification of right maxillary sinus and nasal cavity, and destruciton of floor of right orbit.

2. Radiographic findings

The pantomogram revealed an aggressive bone destruction of floor of the right maxillary sinus and lateral wall of the nasal cavity(Fig. 3). The mandible P-A and Waters' views revealed sinus opacification of the right maxillary sinus and nasal cavity. It is noted that mandible P-A view revealed destruction of anterior wall of the maxillary sinus and Waters' view revealed destruciton of floor of the right orbit(Fig. 4,5). The periapical and occlusal views demonstrated infiltrative bony destruction in the upper right canine area extending to the first molar area, and indistinct floor of the right maxillary sinus. But there were no definite root resorption and tooth displacement (Fig. 6,7).

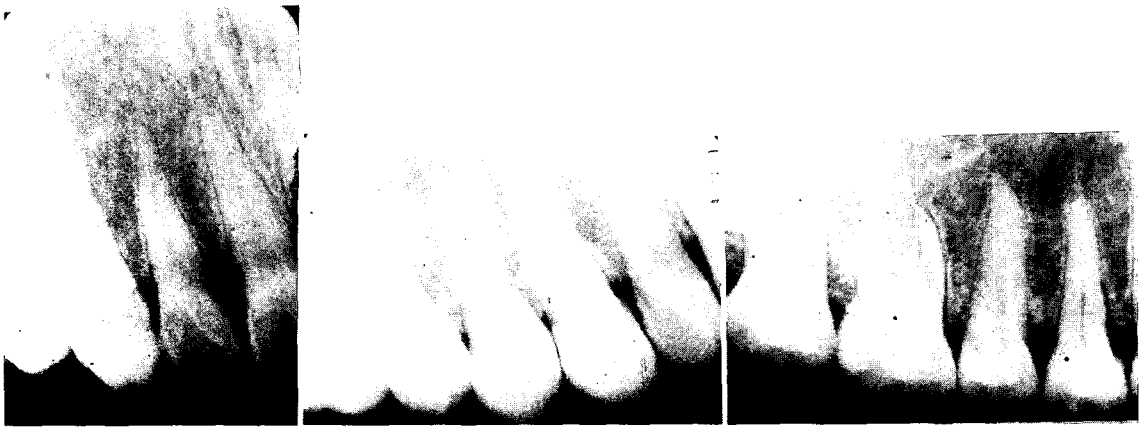

Fig. 6. Periapical views show infiltrative bony destruction in upper right canine area extending to first molar area.

Fig. 7. Occlusal view shows indistinct floor of right maxillary sinus.

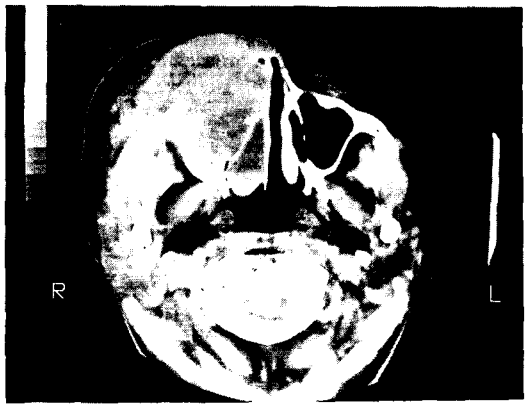

Fig. 8. Axial view shows large inhomogeneous mass in right cheek, maxillary sinus and nasal cavity, and destruction of medial and anterior walls of right maxillary sinus.

The facial CT scan showed that the large inhomogeneous mass was noted in the right cheek, maxillary sinus, and nasal cavity, that medial and anterior walls of the right maxillary sinus were destroyed, and that medial border of mass was attached to the nasal septum(Fig. 8). Also it showed that the right orbital floor was destroyed, that small portion of mass was extended to the right orbit, and that the right globe was displaced superiorly but preserved(Fig. 9).

3. Histopathologic findings

Incisional biopsy was performed. The specimen was obtained from the oral mucosa. In hematoxylin-eosin stain, the section disclosed diffuse and infiltrating neoplastic cells, showing large, hyperchromatic, and bizarre nuclei and atypical mitoses(Fig. 10). Soft tissue by Fontana

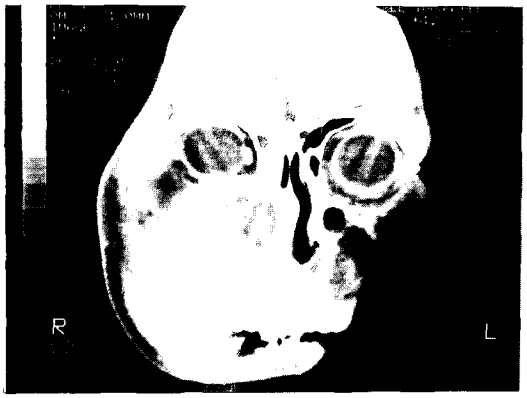

Fig. 9. Coronal view shows destruction of right orbital floor, small portion of mass extended to the orbit, and displacement of right globe superiorly.

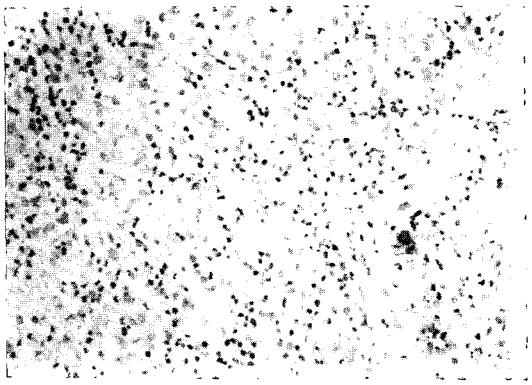

Fig. 10. Photomicrograph shows diffuse and infiltrating neoplastic cells (H & E stain, X400).

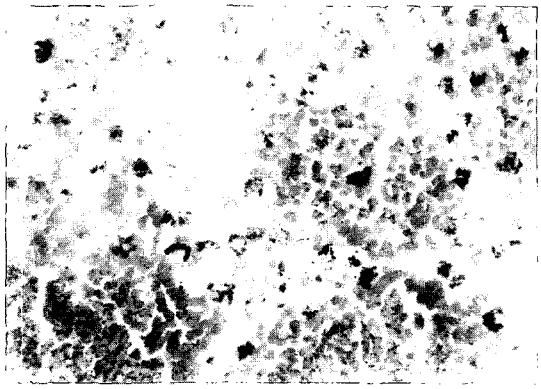

Fig. 11. Photomicrograph shows melanin pigments (Fontana-Masson stain, X400).

-Masson stain showed that melanin pigments was seen frequently(Fig. 11). Diagnosis was malignant melanoma.

4. Treatment

After the debulking surgery of engrowing mass to oral cavity, the patient was transferred to Internal Medicine because of inoperability. She is under the radiation therapy and chemotherapy.

Ⅲ. DISCUSSION

Malignant melanoma involving the nasal and paranasal sinus mucosa is a rare disease that is difficult to treat and generally has a poor prognosis[10]. Ravid and Esteves[8] found only 117 cases on melanomas of the nose and paranasal sinuses in their review of the world literatures. Allen and

Spitz[4], in a study of 934 cases of malignant melanoma, found 8 cases(0.85%) in which the tumor was located in the nasal cavity and 2 cases(0.2%) in the accessory sinuses. Moore and Martin[11], in a further review of 1,546 cases of melanomas, found 9 cases(0.5%) which originated in the nasal fossa. The exact site of origin in this case may be difficult to determine, it is merely stated that the tumor filled or located within the nasal cavity and paranasal sinuses. Since many patients have tumor in the nasal cavity and paranasal sinuses at presentation[12]. However, the majority of lesions originated in the nasal cavity and less than 25% began in the maxillary sinus[10].

The highest incidence of malignant melanoma of the nose and paranasal sinuses is between the ages of 50 to 70 years. The average age is from 56 to 61 years, with a range of 17 to 84 years. The tumor is rare before puberty and the majority of cases occur after the age of 30 years[6,12,13]. There is not a significant sexual difference in incidence[6,11].

The most common presenting signs and symptoms are epistaxis, nasal obstruction, or both[7,11,15]. Gallagher's series, 86 % of 39 patients had these initial complaints. Less symptoms are those of nasal discharge, swelling and deformity of the nose, and headache[8]. The duration of symptoms before medical attention is sought average four to nine months, with a range of one day to two years[12,15].

On physical or gross examination, the size of the tumor varies within a rather wide range. In some cases the tumor was described as of the size of a pea, while in others as a large mass which had extended to the adjacent structures and destroyed the natural landmarks and osseous barriers. Two thirds of nasal melanomas have a dark color, from black to brown to blue-gray, and the remainder are pink or white. The consistency may be firm, friable or cystic, and mass may be sessile, polypoid, or large obstructing lesion. They usually bleed easily on manipulation[8,15]. This case was a 36-year-old female with a chief complaint of the right facial swelling, pain and pus discharge on the ulcerative surface of mucobuccal fold of the upper right molar area. Pain is actually a rare symptom, but when present it is usually due to pressure, blockage of sinus, or an accompanying inflammation[8]. Clinically, there were nasal stuffiness and inflammation in this case. Intraoral examination revealed the incisional wound and black discoloration of mucobuccal fold of the upper right molar area. Incision was done at local clinic diagnosed as inflammation. Melanin pigment is a peculiar feature of melanomas, but the absence of melanin pigment is not a reliable basis for excluding the diagnosis of mucosal malignant melanoma, for the pigment is absent in about half of the case. This pigmentation may vary during the course of the disease and has no influence on course or prognosis[2,12].

It is generally accepted that the histopathology of melanotic lesions of mucous membrane origin is no different than that of melanotic lesions occurring in the skin. The mordern concepts of junctional change occurring in the stratified squamous epithelium of the skin apply to that in the squamous epithelium of pseudostratified epithelium of the nasal cavity and paranasal sinuses[8,11]. Malignant melanomas may be difficult to diagnosis. Histologically, their microscopic appearance is quite variable, and three general cell types are described; small polygonal, large polygonal, and spindle-shaped. The nuclei are often large and vesicular, with prominent nucleoli. Multinucleated tumor giant cells may be present[2,8,13]. They have a higher incidence of pleomorphism and mitotic figures than their cutaneous counterparts[11]. Malignant melanomas have melanin pigment, but it may be quite sparse. From 60% to 69% of lesions are said to contain easily visible pigment in

hematoxylin-eosin stained material, while the remainder either require prolonged search of multiple sections or special stains to demonstrate pigment[6,15]. The confirmation of melanoma should be achieved by the following methods, as advocated by Gallagher[13], Lund[17], Holdcraft[6], and others: (1) The Fontana-Masson stain for melanin should be positive. (2) The pigment should be bleached by potassium permanganate oxalate. (3) A stain for hemosiderin should be negative. Electron microscopy may aid in the diagnosis by identification of intracytoplasmic melanosomes [6,15,16]. We have found melanin pigments in the Fontana-Masson stain in this case.

In the differential diagnosis of primary malignant melanomas of the nose and paranasal sinuses, special care must be exercised to be certain that the tumor in question is not a metastatic melanoma or an undifferentiated carcinoma[5,7,8]. Malignant melanoma of the nose and paranasal sinuses is usually primary. Those affecting the nose secondarily, are known, but are extremely rare. The presence of junctional changes within the mucosal epithelium should be the most valid proof that the growth is primary, but this is usually quite difficult to demonstrate[2,5,9].

Widespread metastases are common, first to regional lymph nodes and then to distant sites such as the liver, lungs, and skin[1,12]. Apparent lymph node metastasis was not noted in this case. Allen and Spitz[4] found that some malignant melanomas remain localized and never metastasize.

The prognosis in all mucosal melanoma is grave. Allen and Spitz[4] pointed out four reasons for the desperate outlook of the melanomas: (1) Delay in treatment was most important. Patients were frequently referred after repeated biopsies had been formed with a wide range of diagnoses. (2) These lesions are not so exposed as are the lesions of the skin, thus allowing time for deeper penetration and metastasis. (3) The lesions are generally more liable to infection and ulceration than those of the skin. (4) Surgical removal from the sinuses or from mucosal lined cavities is technically more difficult than from the skin.

Although melanoma has not been thought of as being a radioresponsive lesion, reports have recently surfaced in the literature supporting the use of radiation as a therapeutic modality[1,7]. Chemotherapy is at present unpromising, and surgery is the treatment of choice[10,12]. Electro-desiccation[18] and cryosurgery[19] have also been reported.

Ⅳ. SUMMARY

In the pigmented tumors, the diagnosis of malignant melanoma is not alway easy. Primary mucosal melanoma arising in the nose and paranasal sinuses is a rare disease with a generally poor prognosis. Melanoma in these area is frequently silent at the onset and produces insufficient symptoms to force the patient to the physician in the early stages of the disease. The importance of early recognition of melanoma by the physician is emphasized. We have an experience a case of primary malignant melanoma of the nose and paranasal sinuses in a 36-year-old female and present this case with a brief review of literatures.

REFERENCES

1. Schoolman, J.G. and Anderson, H.W.: Malignant melanoma of the nose and sinuses. Ann. Otol. Rhinol. Laryngol., 59:124-140, 1950.

2. Crone, R.J.: Malignant amelanotic melanoma of the nasal septum and maxillary sinus. Larynogoscope, 76:1826-1888, 1966.
3. Conley, J.J. and Pack, G.T.: Melanoma of the head and neck. Surg. Gynecol. and Obstet., 116;15-28, 1963.
4. Allen, A.C. and Spitz, S.: Malignant melanoma; clinicopathological analysis of the criteria for diagnosis and prognosis. Cancer, 6:1-45, 1953.
5. Batsakis, J.G.: Tumor of the head and neck. 2nd ed., Williams and Wilkins, 1979, pp. 440-442
6. Holdcraft, J. and Gallagher, J.C.: Malignant melanomas of the nasal and paranasal sinus mucosa. Ann. Otol. Rhinol. Laryngol., 78:5-20, 1969.
7. Bizon, M.J.G. and Newman, R.K.: Metastatic melanoma to the ethmoid sinus. Arch. Otol. Head Neck Surg., 112:664-667, 1986.
8. Ravid, J.M. and Esteves, J.A.: Malignant melanoma of the nose and paranasal sinuses and juvenile melanoma of the nose. Arch. Otolaryngol., 72:431-444. 1960.
9. Jackson, D. and Simpson, H.E.: Primary malignant melanoma of the oral cavity. Oral Surg., 39:553-559, 1975.
10. Trapp, T.K., Fu, Y.S., and Calcaterra, T.C.: Melanoma of the nasal and paranasal sinus mucosa. Arch. Otol. Head Neck Surg., 113:1086-1089, 1987.
11. Moore, E.S. and Martin, H.: Melanoma of the upper respiratory tract and oral cavity. Cancer, 8:1176, 1955.
12. Carter, T.R.: Clinical and pathologic diagnosis. Arch. Otol. Head Neck Surg., 112:450-453, 1986.
13. Gallagher, J.C.: Upper respiratory melanoma pathology and growth rate. Ann. Otol. Rhinol. Laryngol., 79:551-556, 1970.
14. Eneroth, C.M. and Lundbery, C.: Mucosal malignant melanoma of the head and neck. Acta Otolaryngol., 80:452-458, 1975.
15. Mesara, B.W. and Burton, W.D.: Primary malignant melanoma of the upper respiratory tract. Cancer, 21:217-225, 1968.
16. Harrison, D.F.N.: Malignant melanomata arising in the nasal mucous membrane. J. Laryngol., 90:993-1005, 1976.
17. Lund, V.: Malignant melanoma of the nasal cavity and paranasal sinuses. Ann. Otol. Rhinol. Laryngol., 78:5-20, 1969.
18. Conley, J. and Pack, G.T.: Melanoma of the mucous membranes of the head and neck. Arch. Otol. Head Neck Surg., 99:315-319, 1986.
19. Barton, R.T.: Mucosal melanoma of the head and neck. Laryngoscope, 85:93-99, 1975.

- 국문초록 -

비강과 부비동에 발생된 원발성 악성 흑색종의 1예

경희대학교 치과대학 구강악안면방사선학교실

권기정 · 이주현 · 황의환 · 이상래

악성 흑색종은 멜라닌모세포로부터 유래되는 종양으로서, 이의 병인에 대하여서는 현재까지 확실히 구명된 바 없으나, 외상이나 만성 자극 등에 의하여 발생되는 것으로 알려져 있다. 이는 전체 악성 종양의 1.2%를 차지하는 발생빈도가 매우 낮은 질환으로서, 대부분 피부에서 발생되고, 점막에서의 발생은 매우 드물다. 특히 비강과 부비동에서 원발되는 경우는 전체 악성 흑색종의 0.6% - 0.7%에 불과하며, 예후가 극히 불량하다. 비강과 부비동의 원발성 악성 흑색종은 50세 - 70세 사이에서 호발되고, 사춘기 이전에는 거의 발생되지 않으며, 남녀 성차는 없는 것으로 알려져 있다. 또한 비폐색과 비출혈이 가장 흔한 증상이며, 간혹 비루, 안모종창, 두통등을 호소하기도 하나, 종물이 상당한 크기로 성장되기 전에는 특이한 임상증상을 보이지 않으므로 이의 조기발견 및 적절한 치료가 요구된다.

저자들은 최근 안모종창과 동통을 주소로 내원한 36세 여자 환자의 비강과 부비동에 원발된 악성 흑색종 1예를 경험하였기에 문헌고찰과 함께 이를 보고하는 바이다.