Case Report

Sinonasal Malign Melanoma: A Rare Case Report

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ABSTRACT

Sinonasal malignant melanoma is a rarely seen aggressive tumor. Its diagnosis is usually delayed since they have no specific clinical signs or risk factors, present a different histological appearance and mimic many lesions. Its 5-year survival rate is low because of delayed diagnosis and high metastatic potential and reported to be 25% in the studies. In the present report, we have presented the 64-year-old female patient diagnosed with sinonasal malignant melanoma.

Keywords: *Malign Melanoma, Nasal Cavity, Sinonasal.*

ÖZ

Sinonazal Malign Melanom: Nadir Bir Olgu Sunumu

Sinonazal malign melanom nadir görülen agresif bir tümördür. Spesifik klinik belirtilerin veya risk faktörlerinin olmaması, değişik histolojik görünümünün olması ve birçok lezyonu taklit etmesi nedeniyle tanı genellikle geç konmaktadır. Geç tanı konması ve metastaz potansiyelinin yüksek olmasına bağlı olarak 5 yıllık sağkalım düşüktür ve yayınlarda %25 kadar olduğu bildirilmiştir. Burada sinonasal malign melanom tanısı koyduğumuz 64 yaşındaki kadın hastayı sunuyoruz.

Anahtar Sözcükler: *Malign Melanom, Nazal Kavite, Sinonazal.*

Bu makale atıfta nasıl kullanılır: Gürsoy Kuzuluk D, Doran Y, Okuyucu Ş. Sinonazal Malign Melanom: Nadir Bir Olgu Sunumu. Fırat Tıp Dergisi 2023; 28(2): 160-163.
How to cite this article: Gursoy Kuzuluk D, Doran Y, Okuyucu S. Sinonasal Malign Melanoma: A Rare Case Report. Firat Med J 2023; 28(2): 160-163.

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Sinonasal malignant melanoma (SMM) is a rarely seen aggressive tumor and has been first defined by Lucke in 1869 (1, 2). These tumors that constitute 50.2% of all the melanomas are more frequently found in the male gender, Caucasian race and age range of 65-70 years (2). Its diagnosis is usually delayed since they have no specific clinical signs or risk factors, present a different histological appearance and mimic many lesions. Its 5-year survival rate is low because of delayed diagnosis and high metastatic potential and reported to be 25% in the studies (3). The exposure to sunlight and light phototype that are the risk factors for cutaneous melanomas are not considered as risk factors for SMMs (4, 5). Its etiopathogenesis is not yet clear. Mucosal melanoma is neuroectodermal tumor originating from the melanocytes. The intensity of melanocytes in the sinonasal mucosa is higher than other localizations and this can explain the relative higher frequency of primary mucosal melanoma in the sinonasal cavities (4, 6).

In the present report, we have presented the 64-year-old female patient diagnosed with sinonasal malignant melanoma.

CASE REPORT

The 64-year-old female patient applied to the polyclinic of Ear-Nose-Throat with the complaint of dyspnea. The patient had no feature in her medical history. The preoperative imaging results of the patient could not be obtained. The patient was operated with prediagnosis of paranasal squamous cell carcinoma. The material sent for pathological examination consisted of curetted gray-white colored irregular tissue fragments with a volume of 0.3cc. Microscopic examination revealed a tumoral lesion that covered the whole field beneath surface epithelium and developed as nidations with various sizes, solid fields and cell clusters in the tissue samples with largely ulcerated surfaces (Figure 1).

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Geliş Tarihi/Received: 06.04.2022 Kabul Tarihi/Accepted: 23.11.2022

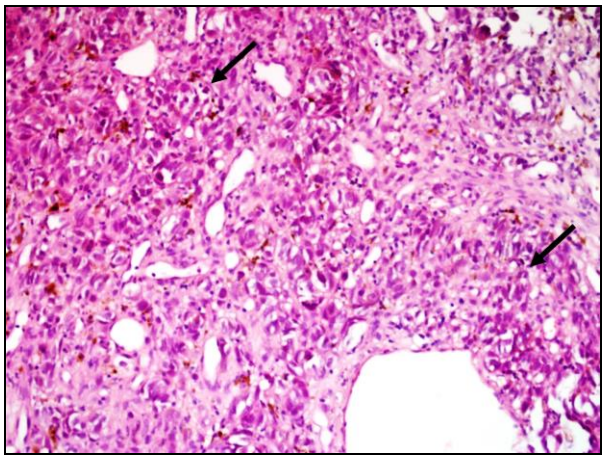

Figure1. *Tumor cells forming solid islands and nidations(H+E, x200).*

Melanin pigment was remarkable in the cytoplasm of some tumor cells (Figure 2).

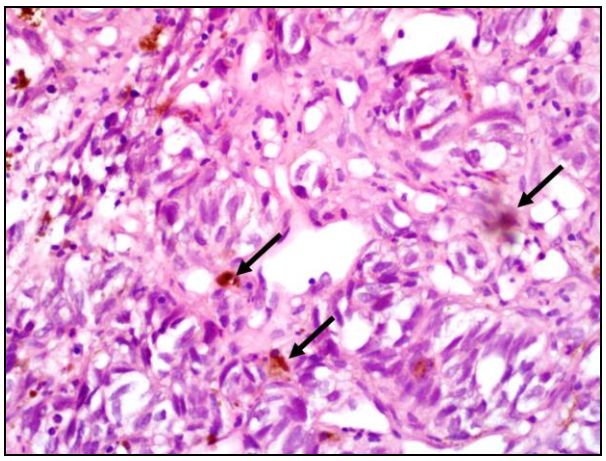

Figure2. *Melanin pigment in the cytoplasm (H+E, x400).*

Necrosis was present. The immunohistochemical examination demonstrated positive immunoreactivity with S100, HMB45 and MelanA in the tumor cells (Figure 3).

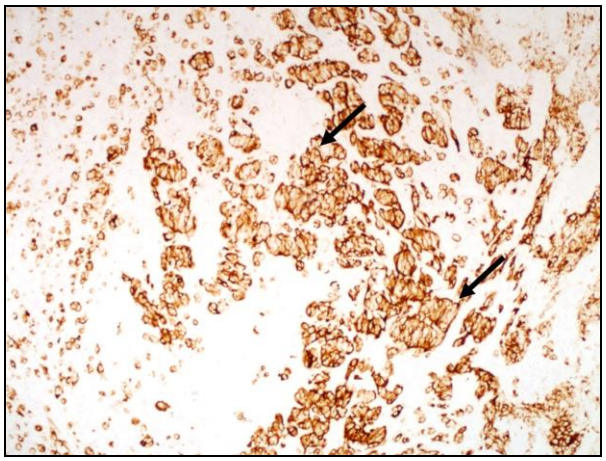

Figure3. *Positivity with HMB45 in the tumor cells (HMB45, x200).*

No immunoreactivity with pancytokeratin and LCA was identified. The case was diagnosed with malignant melanoma according to the morphological and immunohistochemical findings. No feature was detected in the skin examination of the patient. Hence, the patient was evaluated as primary SMM.

DISCUSSION

Primary SMMs constitute 0.5-2% and 4% of all malignant melanomas and malignant melanomas of the head-neck region, respectively (2, 7). They may be seen in all age groups, however they are arely found in the young subjects and its incidence increases after the age of 60 years (2, 8). Only 10-20% of the cases were below 50 years of age (9). SMMs are more frequently seen in the nasal cavity compared with paranasal sinuses (7, 10). Septum and lateral wall are the most common localization in the nasal cavity whereas maxillary and ethmoid sinuses are the most frequently involved sinuses (11). Even though, mucosal melanomas are supposed to develop from the melanocytes that originate from the neural crest in the embryonic period, however, etiopathogenesis is not clear (4, 12, 13). A specific risk factor has not been defined for melanoma, nevertheless, the presence of long-term melanosis has been demonstrated to be associated with the development of oral melanoma (14). The occupational exposure of formaldehyde may be considered as a potential risk factor for nasal sinus malignancies (2, 15). The absence of the symptoms at the beginning or the presence of non-specific symptoms such as nasal obstruction and epistaxis delays diagnosis (2). Rhinorrhea, frontal headache, proptosis, diplopia, hyposmia and epiphora are the late-stage symptoms due to the invasion of the neighboring structures (2, 16). The early diagnosis and treatment of SMMs is very important for a longer survival duration (7). Our case was also over 60 year of age consistently with the literature and no risk factor was present in the medical history.

They are macroscopically polypoid masses with single or multiple pedicles. They may be occasionally hemorrhagic or necrotic and be easily broken down. They are pigmented due to the presence of intracytoplasmic melanin pigment however 1/3 of the cases have no pigment or slightly pigmented (9).

The pathological diagnosis of SMMs may be difficult because of remarkable cytological and structural differences, mimicking other malignancies and inadequate sampling. Several cell morphologies such as epithelioid, spindle, pleomorphic, rhabdoid, plasmacytoid and small cell have been defined for SMMs. SMM is an excellent mimicker and may mimic many tumors that originate from the sinonasal region in the category of “small round blue cell tumor” (17, 18). NK/T cell lymphoma, plasmacytoma, squamous cell carcinoma, NUT carcinoma, sinonasal undifferentiated carcinoma, olfactory neuroblastoma, Ewing sarcoma, rhabdomyosarcoma, and neuroendocrine carcinoma are the tumors

that should be involved in the differential diagnosis of SMM. Since these tumors require different treatment modalities and present different prognoses, the differential diagnosis should be carried out between these tumors and SMM (17, 19). Occasionally, its diagnosis may be mixed with chronic sinusitis or a nasal polyp. SMM is a common presentation of a nasal polyp which that would be eliminated as a preliminary procedure with a final diagnosis established on detailed histopathological examination of SMM (9).

A metastasis from cutaneous malignant melanoma to the sinonasal tract should also be involved in the differential diagnosis, although quite rare. The primary SMM may manifest junctional activity and is not encountered in metastatic lesions, however, its absence does not preclude the diagnosis of primary SMM. Intramucosal spread of melanocytes in the adjacent epithelium (epidermal migration) histologically strengthens the possibility of a primary tumor (9, 20). Surface epithelium is usually preserved in the metastatic melanomas.

Even though, the presence of melanin pigment supports the diagnosis of SMM, many cases have very limited or no pigment. The positivity of melanocytic marker has not adequate sensitivity and specificity, however they are helpful diagnostic markers. Some of the SMMs may show aberrant differentiation both histomorphologically (e.g., cartilaginous or skeletal muscle differentiation) and immunohistochemically (e.g., staining for epithelial, mesenchymal, or neuroendocrine markers). As a consequence of these facts, these tumors can be delayed or misdiagnosed (17).

Skin melanoma may genetically exhibit the mutations in BRAF, c-kit and NRAS. However, BRAF and c-kit mutations are very rare in the mucosal melanomas whereas NRAS mutation is relatively more frequently seen (21).

SMM requires an aggresive and multimodal treatment while there are many treatment modalities including surgery, radiotherapy, chemotherapy and biological therapy. The complete surgical resection is mandatory to prolong the survival duration, however, implementation of this technique may be occasionally difficult because of complex anatomical structure of the nasal cavity and sinuses as well as the neighborhood of the tumor to the critical structures (skull base, ocular orbit). Radiotherapy and chemotherapy are performed in the advanced-stage patients (22). Despite the improvements in the treatment, the rates of local recurrence, distant metastasis and 5-year survival are 31-85%, 25-50% and 20-30%, respectively (17). SMMs metastasize less frequently to cervical lymph nodes and more frequently to lungs and brain in contrast to squamous cell carcinomas (9).

Conclusion

Sinonasal malignant melanoma (SMM) is a rarely seen aggressive tumor. Its diagnosis is usually delayed since they have no specific clinical signs or risk factors, present a different histological appearance and mimic many lesions. Sinonasal malignant melanomas are an excellent mimicker and may mimic many tumors that originate from the sinonasal region. A meticulous histopathological examination should be carried out to avoid an inaccurate treatment.

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