

## A MALIGNANT MELANOMA METASTASIS WITH UNKNOWN PRIMARY; AN ATYPICAL CLINICAL PRESENTATION; CASE REPORT.

### Primeri bilinmeyen malign melanoma metastazı; Atipik bir prezentasyon; Olgu sunumu.

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Cer San D (J Surg Arts), 2012;5(1): 12-15.

#### ABSTRACT

Malignant melanoma metastasis with unknown primary is a rare clinical entity. A 42-year old man was admitted with the complaint of a rapidly growing mass in his right axillar region. In physical examination, a mass that 24-25 cm in diameter, irregular border, fusiform shaped and enlarged of outward from the axilla was detected. Mass excision and level I-II axillary dissection was performed. Malignant melanoma metastasis was found as result of histopathologic and immunohistochemical staining examination of this mass. This case has been reported by reason of its rare incidence and interesting clinical view.

**Key words :** Malignant melanoma, metastasis, lymph node metastasis, primary unknown

#### ÖZET

Primeri bilinmeyen malign melanom metastazı nadir görülen bir klinik durumdur. Kırkiki yaşında erkek hasta sağ aksiller bölgede hızla büyüyen bir kitle şikayeti ile başvurdu. Fizik muayenede; sağ aksiller bölgeden dışarıya doğru büyümüş, 24-25 cm boyutunda, düzensiz sınırlı ve fuziform şekilli bir kitle saptandı. Kitle eksizyonu ile birlikte level I-II aksiller diseksiyon ameliyatı yapıldı. Histopatolojik ve immünohistokimyasal incelemeler sonucu malign melanom metastazı saptandı. Olgu, nadir görülmesi ve ilginç olduğunu düşündüğümüz klinik görünümü nedeniyle sunuldu.

**Anahtar kelimeler:** Malign melanom, metastaz, lenf nodu metastazı, primeri bilinmeyen tümör

#### INTRODUCTION

Malignant melanoma accounts for 1-3% of all malignancies and 5% of all skin malignancies (1). Like the other malignancies, melanoma has the capability of distant metastasis via lymphatic involvement. Besides, malignant melanoma may present with only lymphatic metastasis or any other systemic metastasis. Primary unknown malignant melanoma is a rare clinical entity. The most widely accepted theory about this clinical entity is the primary tumor that has been regressed (1). On account of the number of patients are small data on clinical course and prognosis are limited. This case has been reported on

account of its interesting clinical view and discussed the prognosis of this rare entity.

#### Case

A 42- year old man was admitted to our clinic with the complaint of rapidly growing mass into last 4 months in his right axillar region. In physical examination, a mass that 24-25 cm in diameter, immobile, irregular border, fusiform shaped and enlarged of outward from the right axillary region was detected (Figure 1). There was no arm edema. All hematological, biochemical and tumoral markers levels were normal. A mass that lobulated contour,

21x23 cm in diameter and heterogen internal structured was revealed in magnetic resonance imaging (MRI) (Figure 2).



**Figure 1:** Physical examination; a mass that 24-25 cm in diameter, immobile, irregular border, fusiform shaped and enlarged of outward from the right axillary region. The skin apex of the mass is thin and bright red color appearance.



**Figure 2:** MRI findings; the mass that lobulated contour, 18x19 cm in diameter and heterogen internal structured; had high signal intensity in T2-weighted images.

Additionally, axillary artery and vein were compressed by the mass, but there were no radiologic signs of invasion. Arterial and venous flows were normal and no thrombus in vascular lumen in doppler ultrasonography (USG). The other diagnostic procedures, including neck, thorax (for lung parenchym and mediastinum), abdomen and pelvic computed tomography (CT) and conventional graphies of the long bones, were normal. Multiple fine needle aspira-

tion biopsies (FNAB) were performed from different areas in the mass owing to it contain large amounts of necrotic areas as radiologically. Malignant cells were revealed in histopathologic examination of this biopsy materials, but a specific diagnosis was not considered. We decided that the surgical excision due to the diagnosis is unclear and the mass volume and shape are very restrictive for maintenance of the normal daily activities of patient. In operation, the mass was easily excised from the thoracic wall and regional vascular structures. Axillary dissection was also performed due to multiple lymphadenopathy. Remaining skin defect after operation was partially closed with non-absorbable sutures (Figure 3). Malignant melanoma metastasis was detected as result of final histopathologic and immunohistochemical staining examination. The primary lesion has not found on the eye, rectum and the other possible locations for malignant melanoma. A skin lesion was excised that 1 cm in diameter, regular border and brown color from the left lumbar region. In histopathologic examination of lesion, melanocyte was not detected and might be the regressed primary focus was thought. The skin defect was closed with full-thickness skin graft after 25 days from the first operation (Figure 3). The patient was discharged postoperatively 32th days uneventfully. Nine-months after the operation, multiple lung metastasis were detected in positron emission tomography (PET) and chemotherapy was planned.

## DISCUSSION

The malignant melanoma has become one of the major health problem and the incidence is increasing worldwide. The malignant melanoma has high metastasis capability. The most common sites of metastasis were the lymph nodes and lungs followed by the liver, brain, bone and adrenal glands, respectively (2). Furthermore, this disease may presenting with only lymphatic metastasis and the incidence of a metastatic malign melanoma with unknown primary varies from 2% to 15% in different series (3,4). Some criteria are needful for diagnosis of primary unknown malignant melanoma metastasis: 1. the eyes, rectum and genital region examination should be normal, 2. should not be the following criteria; orbital enucleation history, surgical operation history for any skin lesions, such as nevus, and surgical operation history for the skin region where lymphatic drainage towards this lymph nodes which have been detected of malignant melanoma metastasis (5). Our patient had none of these criteria.

Several different theories are proposed to explain this confusing clinic entity. The most widely accepted theory is spontaneously regressed primary lesion theory (1). Previously excised and histologically misdiagnosed, concurrent unrecognized melanoma; and de novo malignant transformation of sequestered melanocytes theories are the other theories. Our patient had not been operated in the

past, besides, was not detected any possible primary lesion in clinic, radiologic and endoscopic examinations. Only a skin lesion was detected that possible regressed primary lesion was considered after the histopathologic examination.



**Figure 3:** Postoperative view of the operation area; remaining skin defect after resection was partially closed with non-absorbable sutures (above), two months later the skin graft (below).

The prognosis of patients with systemic metastatic malignant melanoma (stage-IV) is poor and mean survival less than 1 year.<sup>6</sup> Whereas, for stage-III disease, the 5-year survival rates are between 13% and 69%. Tas et al. reported that median overall survival range is 6 - 25.6 months in node positive, stage III malignant melanoma patients in Turkey (7). Also, it is reported that male gender is significantly negatively correlated on overall survival and unlike other studies status (number) of involved lymph

nodes was not found to be significant predictor of prognosis on survival in this study.

Different estimates are proposed about prognosis of the primary unknown melanoma patients. Giuliano et al. reported that the prognosis and overall survival of the primary unknown melanoma patients are similar with those are the primary known melanoma (8). On the other hand, Anbari et al. reported that the survival of the primary unknown melanoma patients are longer than patients who primary known cutaneous melanoma diagnosed with the lymph node metastasis (9). In another study, Velez et al. reported that the overall 5-year survival rate and the median survival interval was 28% and only 17 months respectively, in the primary unknown melanoma (10). On the other hand, between 30 to 45% five-year survival rate has also been reported in those who have the only lymph node metastasis when after regional lymphadenectomy (5). In our case, multiple lung metastasis were detected in 9th months after the operation, progression was detected of this metastatic lesions in 12th months and is being followed as clinically with lung metastasis in postoperative 16th months.

In conclusion, the incidence of malignant melanoma is increasing worldwide and axillary region is the most common localization of lymphatic metastasis area for this disease. For this reason, clinicians must be aware of the different manifestations of metastatic malignant melanoma, even if clinical findings are atypical and primary localization of melanoma is not known.

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