PRIMARY SYNCHRONOUS UVEAL AND SKIN MALIGNANT MELANOMA – case report

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SUMMARY – Simultaneous occurrence of primary uveal melanoma and skin melanoma is rare. We presented a 66 year old female patient with primary synchronous choroidal and skin malignant melanoma on right upper-arm. The patient has a negative family history for cutaneous or uveal melanomas and did not display the dysplastic nevus syndrome phenotype. Six years after enucleation of left bulbous and skin excision of malignant melanoma the patient presented with lymphatic metastasis in right axillary lymph nodes. After two years metastases in mesenteric lymph nodes and solitary nodule in spleen appeared. Nine years after initial presentation of melanomas patient developed metastases in ingvinal lymph node. Metastatic spread was most probably due to lymphatic dissemination of more biologically aggressive primary skin malignant melanoma. Ten years after initial diagnoses the patient is alive and without new metastases.

Key words: *uveal melanoma, primary cutaneous malignant melanoma, simultaneous occurrence, risk factors*

Introduction

Primary malignant melanoma of the eye or skin are not common tumors and their coincidence is even most uncommon.[1] The cause of uveal melanoma is unknown but several risk factors for the development have been identified: age, sex, genetic factors and possibly environmental factors.[2,3] We presented a rare case of a patient with synchronous occurrence of primary uveal and cutaneous melanoma.

Case report

A 66-year- old female patient with symptoms of retinal detachment and with atypical pigmentary skin lesion was admitted to our Hospital in April 1993. The patient did not display the dysplastic nevus syndrome phenotype and has a negative family history for cutaneous or uveal melanomas. After enucleating of left bulbous macroscopically heavily pigmented, mushroom-shaped choroidal tumor mass was seen measuring 1,7 cm in largest diameter

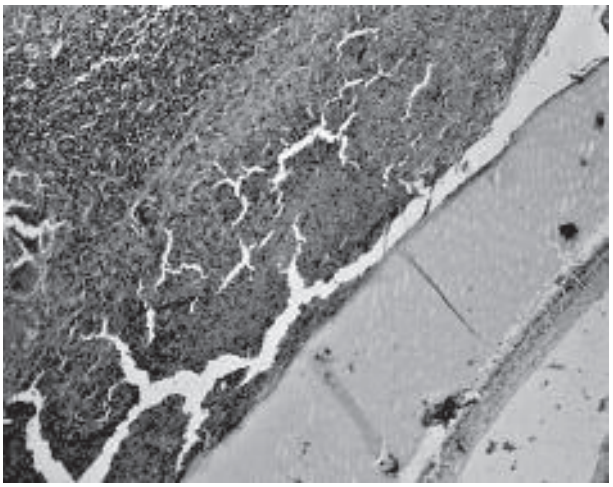

Figure 1. Choroidal melanoma composed predominantly of epithelioid cells with propagation to retina (H&EX200)

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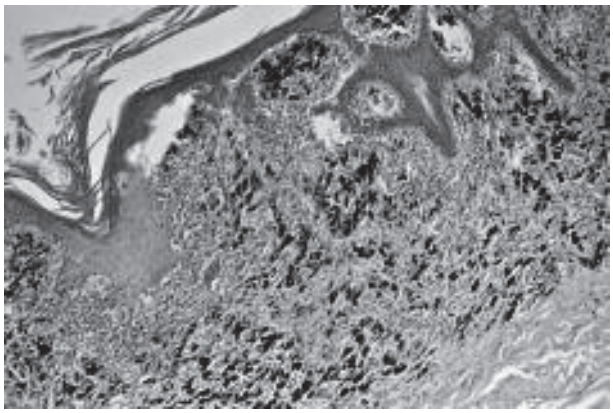

Figure 2. Skin melanoma composed of polymorph, heavily pigmented atypical melanocytic cells (H&E X 200)

on the base and 0,7 cm in elevation. Histology revealed malignant melanoma infiltrating from choroid to iris and ciliary body on one side composed predominantly of epitheloid type cells with melanin pigment in cytoplasm (Figure 1). Tumor stage was pT3. In same time skin excision of irregularly pigmented lesion measuring 2,5 cm in largest diameter on right upper-arm was performed and pathohistological analysis confirm the diagnosis of superficial spreading malignant melanoma Clark III, Breslow III (Figure 2). In January 1999 after extirpation of right axillary lymph nodes metastatic melanoma was seen histologically in one lymph node (Figure 3). Eight months later patient was admitted in Department of Surgery for operation of metastatic mesenteric lymph nodes and spleen with

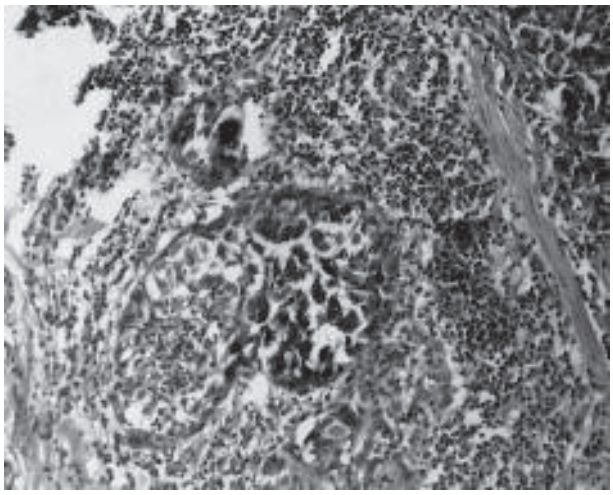

Figure 3. Metastatic melanoma to axillary lymph node (H&EX 200)

two metastatic nodes measuring in largest diameter 4,5 cm and 2,5 cm histologically consisted of loosely cohesive atypical melanocytic cells. In February 2002 extirpation of metastatic lymph node from left ingvinal region was performed.

Discussion and conclusion

Simultaneous occurrence of uveal and cutaneous melanoma is rare and the literature date are not nummerous.[2] Bataille V. and coworkers described five cases of coexistent primary ocular and cutaneous melanoma, three of ocular melanoma were uveal and two conjuctival. All uveal melanomas were presented in females patient aged between 56 and 76 years.[1] This correlate with sex and age of our patient. Uveal and cutaneous melanomas diffear in tumor biology, immunophenotypes and the demographic correlates of their occurrence.[4] It is well established that uveal melanoma especially those that did not spread out of uvea rarely develop hematogenous metastases. In presented case the lymphogenous metastases to right axillary, mesenteric, inguinal lymph nodes and spleen were probably derived from cutaneous melanoma. The problem is to identify the source of metastases because there is no histologicaly cellular difference between morphology of cutaneous and uveal melanoma. Some authors recently proposed that the association of intraocular melanoma with cutaneous melanoma and dysplastic nevus syndrome in the same patient and in different members of the same family is not coincidental.[5] Therefore, until the relationship between intraocular and cutaneous melanomas is not fully understood the skin of patients suspected of having intraocular melanomas should be examined routinely for evidence of atypical melanocytic lesions.[6] lt has also been revealed that the presence of cutaneous dysplastic naevi in patient with uveal melanoma is associated with an increased incidence of the prognostically worst forms of uveal melanoma (epithelioid or mixed cell type melanomas). The presence of cutaneous dysplastic naevi is not only a risk factor but also a prognostic factor for uveal melanoma.[7]

In largest retrospective study from Singh AD and al. was reviewed 4500 medical charts of the patients with uveal melanoma for family history and association to cutaneous melanoma. Familial uveal melanoma most often (63%) affects first-degree relatives, rarely affects more than two persons in a family, and may be associated with a generalized inherited predisposition to cancer.[8,9] Patients with familiar uveal melanoma are at four time's greater risk to develop a second primary malignant neoplasm than peo-

ple in the general population. [8,10] But still rare case of coexistent uveal and skin melanoma raise the possibility of some shared etiologic factors in pathogenesis of multicentric malignant melanoma.

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Sažetak

ISTOVREMENI PRIMARNI UVEJALNI I KOŽNI MALIGNI MELANOM – PRIKAZ SLUČAJA

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Istovremeni melanom kože i uveje je rijedak. U radu je prikazan tijek rijedkog slučaja istovremene pojave melanoma žilnice i primarnog melanoma kože desne nadlaktice u bolesnice stare 66 godina. U obiteljskoj i osobnoj anamnezi bolesnice nije bilo melanoma niti sindroma displastičnog nevusa. U bolesnice su se pojavile limfogene metastaze u limfne čvorove desne aksile šest godina nakon enukleacije lijeve očne jabučice i ekscizije kožnog melanoma. Dvije godine nakon toga su se pojavile metastaze u mezeneterijalne limfne čvorove i solitarne metastaze u slezenu. Devet godina nakon pojave melanoma razvila se metastaza u limfni čvor ingvinuma. Metastatski rasap nastao je limfogeno i najvjerojatnije potječe od primarnog k utanog melanoma koji uvijek ima lošiju prognozu od uvejalnog melanoma. Deset godina nakon pojave primarnog melanoma uveje i kože bolesnica je živa i bez pojave novih metastaza.

Ključne riječi: *uvejalni melanom, primarni kožni melanom, istovremena pojava, faktori rizik*