

Amelanotic malignant melanoma of the perineum: a case report

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Summary

The case of a patient diagnosed and surgically treated for amelanotic malignant melanoma of the perineum, accompanied by several local relapses, metastases to iliac-femoral lymph nodes, and distant metastases to both lungs is presented. Survival, up to date, equals 12 years. Amelanotic malignant melanomas are very rare tumors. Perineal and perianal localization of these tumors is especially rare. References cite about 500 cases with survival limited to between six months and one year after diagnosis..

Key words: Amelanotic malignant melanoma; Survival; Perineum.

Introduction

Malignant melanoma is one of the most malignant human tumors. It is characterized by extremely fast growth and fast formation of hematogeneous and lymphogeneous generalized metastases. While 80% of all primary malignant melanomas are located in the skin and 19% in the eye, the remaining one percent comprise malignant melanomas localized in mucous membranes of the oral cavity, esophagus, bowels, rectum, nose, urethra, and vagina. They are seldomly found in the vulva, perineum, palms or soles, i.e., in places not exposed to ultraviolet solar radiation. Amelanotic melanomas comprise only two percent of all melanomas [1] and are most commonly subungual [2]. Urogenital and perianal localization of such tumors is extremely rare.

Amelanotic malignant melanomas are the most aggressive of all histological types of malignant melanomas. Tumor cells in these histological types of tumors are undifferentiated to such a degree that they are not able to create melanin. This renders them highly malignant. Amelanotic melanoma diagnostics is a special problem because macroscopic forms of these tumors can be very diverse; due to the lack of pigment, melanoma is often excluded from consideration in the process of early diagnostics [3]. On the other hand, deep tumor invasion into dermis and hypodermis, or even deeper, is present in a significant percentage of cases. Data cited in references show a positive correlation between depth of invasion and final outcome of the disease.

Case Report

The case is presented of a patient whose survival after diagnosis and surgical treatment of amelanotic malignant melanoma primarily localized in the perineum, followed by several relapses and distant metastases equals 12 years. Discomfort which caused the 60-year-old patient to report for a medical

exam were tingling sensation in the perineal area, occasional contact bleeding, and occasional bleeding during defecation. These discomforts appeared several months before the first visit to the physician during which a small, protuberant tumor lesion was visualized, around ten mm in size, macroscopically polypoid in appearance, localized on the skin of the perineum some two cm above the anus. Macroscopic appearance of the lesion did not contain any elements which would lead to suspect that the malignant tumor, especially not malignant melanoma, was present. The lesion was surgically removed. Amelanotic malignant melanoma was verified by histopathological examination; it was characterized by dominantly trabecular, partly solid organization with strong invasion into dermis and hypodermis, Breslow III, Clark level V.

Considering this unexpected histopathological finding, computed tomography (CT) exam of the abdomen and pelvis was performed postoperatively. Lymphadenopathy was found in the right inguinum, while one month after the control check-up following the surgical removal of the tumor, a local relapse was detected. A dissection of the right inguinum was performed accompanied by lymphadenectomy and excision of the local relapse. Metastases were verified in lymph nodes. Histopathological examination of the local relapse also confirmed the existence of amelanotic malignant melanoma, infiltrating only hypodermis, without any deeper invasion. The patient underwent regular quarterly check-ups over the next two years. A metastatic lesion in the upper lobe of the left lung was diagnosed by radiographic exam during the third year; it was subsequently surgically removed. Metastasis of amelanotic malignant melanoma was confirmed. The tumor did not affect the visceral pleura and edges of the resection did not contain malignant cells. A month later a local relapse appeared in the perineum and was surgically removed. Over the following three years, the patient remained symptom-free. However, a solid metastasis was subsequently discovered in the right lung as well. Resection was performed again, this time of the right lung and metastasis of the amelanotic malignant melanoma was histopathologically verified; edge of the resection did not contain malignant cells. One year later, another local relapse in the perineum appeared and was surgically removed. Histopathological examination showed that the depth of invasion of this relapse was even smaller than that of previous local melanoma relapse in the perineum. One possible explanation for the smaller depth of invasion of the relapse is the existence of scar tissue in deeper

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layers formed as a result of surgical removal of primary tumor and several local relapses. Since then the patient has been subjected to regular check-ups. There are no symptoms and the malignant disease has been in remission phase for the last five years. Survival period since the moment of primary diagnostics of this tumor until the present day is 12 years.

Discussion

Perineal amelanotic malignant melanoma is a very rare tumor. References up to date cite only about 500 cases [4, 5]. Recent epidemiological analyses indicate, however, that the incidence of malignant melanoma in the world is increasing. Supported by references, a clear adult female predominance with involvement of the lower genito-urinary tract was identified [6]. Literature reviews describe three cases of amelanotic malignant melanoma of the vagina, two cases each originating from the cervix, ovary, and urethra; such cases were also identified in cerebral cortex (frontal and fronto-parietal area), rectum, nasal cavity, tongue, breast, and tonsillar sinus [7-9].

Early detection, accurate staging system that rates the level of invasion according to Clark and Breslow, surgical therapy, regular check-ups, and prompt treatment of relapses are all important for survival [10-14].

Prognosis is dependent on the Breslow's level at time of diagnosis. In amelanotic melanoma, the cues leading to diagnosis are often absent, leading to reports of missed diagnoses and poor prognoses.

According to data presented by American Joint Committee on Cancer, the five-year survival of patients suffering from malignant melanoma is under ten percent [15]. References cite cases of patients with rare malignant melanoma localizations whose survival ranged from six months to one year after being diagnosed. Cases of survival around five years are extremely rare, while ten or more years survival are only sporadic.

It is a fact that malignant melanomas can arise in unconventional areas. Gross tumor appearance and the unavailability of an immunohistochemical panel may result in their misdiagnosis.

Considering the fact that malignant melanomas are predominantly tumors of the skin, an organ easily accessible for regular self-examination, education of the entire population with emphasis on self-examination of skin, and all new lesions appearing on it is very important. The relationship between sun ultraviolet (UV) radiation and appearance of "de novo" tumors on completely healthy skin is well-known. However, it is very important to perform exams on those skin regions not exposed to UV radiation, yet posing as possible predilection sites for development of various macroscopic forms of amelanotic melanoma, the most aggressive malignant tumors charac-

terized by very poor prognosis and short survival rate. Early diagnostics and early surgical treatment can prolong remission and survival periods even with these most aggressive human malignant tumors.

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