



Case Report

Survival for 38 years with Animal-type malignant melanoma

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ABSTRACT

Animal-type melanoma is a low-grade malignant tumor with a good prognosis. Although it is common in old grey horses, the tumor is exceedingly rare in humans. Only 173 cases have been reported till 2011. We report a case in a 38-year-old woman who had the neoplasm in the left labium majus since birth. The tumor was composed of heavily melanized polygonal tumor cells with mildly atypical nuclei proliferating diffusely and in nests in the dermis and subcutis with invasion of hair follicles and nerve bundles. Mitotic figures were not seen. Regional lymph nodes were not enlarged.

Present case describes a patient with animal-type melanoma who has had the lesion since birth and who is still living despite having the tumor for 38 years. It attests to the fact that this tumor has indolent biologic behaviour despite being malignant. Mistaking this case for other highly pigmented melanocytic tumors, benign or malignant, is a potential diagnostic pitfall.

INTRODUCTION

Animal-type or Equine-type melanoma, a rare subtype of malignant melanoma, is so named because it closely resembles the identical melanocytic tumour found in grey horses. Although very rare in humans, these tumours occur in up to 80% of old grey horses.¹

While William Dick was the first researcher to describe the tumor in the skin of grey horses in 1832, Darier was the first to describe it in humans in 1925.² Since then a number of names have been given to these tumors in humans viz. animal-type melanoma, equine-type melanoma, pigment

synthesizing melanoma and melanoma with prominent pigment synthesis.³

Animal-type melanoma has been found to have far better prognosis as compared to conventional melanoma even though metastases may occasionally develop. For this reason, Zembowicz et al⁴ proposed a new term 'pigmented epithelioid melanocytoma (PEM) in 2004 for this tumor.

CASE REPORT

A 38-year-old female came with a dark pigmented nodular lesion on the left labium majus which was present since birth. The patient had a history of recurrent discharge from the hairy pigmented lesion. Provisional clinical diagnosis was infected hairy nevus. There was no history

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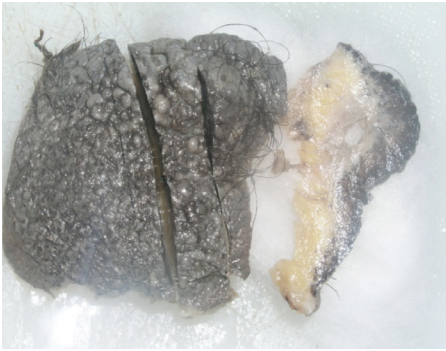


Figure 1: Gross appearance, Animal-type melanoma. The lesion is heavily pigmented.

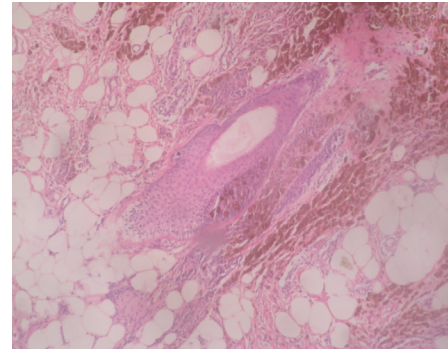


Figure 2: Destructive invasion of hair follicle and subcutaneous fat by heavily pigmented tumor cells (HE stain, X40).

of Carney complex (myxomas, spotty skin pigmentation, endocrine overactivity, and schwannomas). Regional lymphadenopathy was not present.

Grossly, the lesion was uniformly black in colour and measured 7x4.5x3 cm. Cut-sections showed dark pigmentation up to the level of subcutaneous fat (fig. 1).

Microscopically, there was asymmetrical proliferation of mildly atypical, polygonal tumor cells having moderate amount of heavily melanized cytoplasm in the dermis and subcutis (Clark level V). Maturation with descent was not present. Breslow thickness was 11mm. Invasion of hair follicles and subcutaneous fat (fig. 2) was present. Intra and perineural invasion were also seen (fig. 3). Nucleoli were small. The diagnosis was made after ruling out other heavily pigmented benign and malignant melanocytic tumors.

DISCUSSION

Animal-type melanoma is characterized by proliferation of heavily pigmented mildly atypical tumor cells in the dermis with a possible extension into the subcutis. The tumor cells may invade adnexal structures, nerve bundles and lymphovascular spaces. Although the tumor has a tendency to spread to regional lymph nodes, distant metastasis is rare.^{1,2} Among the 173 cases reviewed by Pai-Shan Cheng et al⁵ the positive rate of lymph node metastasis was 22.4%. Regardless of the relatively high rate of lymph node metastasis, there were only 2.9% of cases whose disease had spread beyond the lymph nodes. Even if it metastasizes, its prognosis is better than that of conventional malignant melanoma with the same depth of invasion.⁵ In a review of 18 cases of animal-type melanoma by Orlandi et al⁶, 5-year survival rate was found to be 94%. In conventional melanomas with similar thickness and no ulceration, 5-year survival rate was 70%.⁶

The longest follow-up period (67 months) was reported by Mandal et al.⁷ All 26 patients were free of disease during the follow-up period.⁷ Present case report is the first to describe a patient with animal-type melanoma who has had the lesion since birth and who is still living despite having

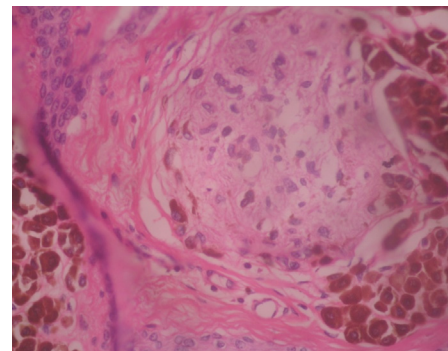


Figure 3: Intra and perineural invasion (HE stain, X400).

the tumor for 38 years. Whether this tumor evolved from a congenital nevus, is open to discussion. These reports succinctly illustrate animal-type melanoma to be an indolent tumor despite being malignant. Therefore, for appropriate management of a patient, it should not be mistaken for other heavily pigmented benign and malignant melanocytic lesions, such as cellular blue nevus, malignant blue nevus, epithelioid blue nevus, pigmented spindle cell nevus, Spitz nevus and deep penetrating nevus.⁸

Cellular blue nevus is a well-circumscribed benign neoplasm that may show a striking pigment synthesis. Hypercellular zone is composed of nests of plump oval cells with bland round nuclei containing delicate chromatin and small blue nucleoli. Hypocellular areas are made up of spindle-shaped cells with pigment-laden macrophages, reminiscent of a common blue nevus, in the background of collagenised stroma.

In a malignant blue nevus, which occurs after malignant transformation of a cellular blue nevus, tumor cells are frankly malignant.⁹

The main distinguishing feature of pigmented spindle cell nevus of Reed is the superficial plaque-like structure of

the lesion composed of spindle-shaped cells, in contrast to deeply invasive extent of animal-type melanoma.¹⁰

The pigmented epithelioid variant of Spitz nevus is a well-circumscribed lesion showing epidermal hyperplasia with hyperkeratosis, intra-epidermal nested and lentiginous proliferation of heavily pigmented epithelioid melanocytes with abundant eosinophilic cytoplasm, uniform vesicular nuclei and prominent eosinophilic nucleoli. Mitoses are confined to the junctional component. Maturation of tumor cells is present.

Deep penetrating nevus shows overlapping features with blue nevus and Spitz nevus. It is wedge-shaped, deeply pigmented dermal tumor with a junctional component. Tumor cells are arranged in nests or bundles and have a short spindle-shaped or, less commonly, round morphology. A thin rim of sustentacular cells is present around the edges of many nests. The dermal component of tumor cells may reach the subcutis.^{2,5}

Epithelioid blue nevus is rare and it occurs in association with Carney complex and also sporadically. Histopathologically, it is indistinguishable from animal-type melanoma. Zembowicz A et al⁴ coined the term 'Pigmented epithelioid melanocytoma' to encompass both of these lesions. PEM is best classified as a low-grade melanoma or borderline melanocytic tumour with metastatic potential.¹¹

CONCLUSION

Because of excellent prognosis of the tumor, 'pigmented epithelioid melanocytoma' is the term preferred to 'animal-type melanoma'. The latter term is inappropriately alarming both to the clinician and the patient. And its diagnosis should be made only after ruling out other heavily pigmented benign and malignant melanocytic tumors.

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