CASE REPORT

Pigmented Epithelioid Melanocytoma: Case Report

Melanocitoma Epitelioide Pigmentado: Relato de Caso

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INTRODUCTION

Pigmented epithelioid melanocytoma (PEM) is a rare melanocytic tumor composed of pigmented epithelioid and dendritic melanocytes with large nuclei. This histologic entity encompasses tumors previously described as epithelioid blue nevus of the Carney complex. Pigmented epithelioid melanocytoma is a specific clinicopathological entity with particular clinical presentation and histological features. We present the case of a 5 year old female patient with a heavily pigmented papule on her right thigh that showed histological findings compatible with pigmented epithelioid melanocytoma and discuss the relevance /clinical significance of sentinel lymph node biopsy as a staging procedure in this particular neoplasm.

DISCUSSION

Sentinel lymph node (SLN) involvement observed in the majority of PEMs along with the tumor once being categorized as an "animal-type" melanoma prompted most previous reports to include...
SLN sampling as a diagnostic procedure. However, the frequent SLN involvement in PEMs does not have the same adverse prognostic significance as in conventional melanoma\textsuperscript{2,6-8} and the positive rate in cases undergoing SNLB may be over-estimated due to a more progressive clinical course or deeper tumor cell invasion.\textsuperscript{9}

Another theory is that PEMs may be part of the neoplasms known as “benign metastasizing” tumors.\textsuperscript{8} These particular kinds of tumors include both cutaneous and non-cutaneous lesions such as benign melanocytic naevi, endometriosis and metastasizing leiomyomas.\textsuperscript{8}

Additionally, based on current studies, a positive sentinel lymph node sampling appears to present little important evidence of the lesion’s biological potential and its risk of developing clinically significant metastatic disease.\textsuperscript{10} Taking that into consideration it can be argued that SLN sampling should not be performed in every patient with PEM.\textsuperscript{8}

There are no clearly established criteria for performing SLNB in PEM patients\textsuperscript{9} and no histological criteria predictive of potential metastatic behavior of the tumor.\textsuperscript{2,10} Current studies suggest that PEM is an indolent melanocytic tumor and long-term follow-ups have not revealed disease-specific lethal cases regardless of tumor deposits in sentinel lymph node sampling.\textsuperscript{1}

Sentinel lymph node sampling was not performed in this case due to lack of evidence proving diagnostic or prognostic value of this practice on this kind of tumor.\textsuperscript{1} A complementary wide surgical excision was performed and the patient will go through a long follow-up period.

In conclusion, while it is considered to be an indolent tumor, PEM patients should undergo a long follow-up period and further studies are needed on whether SLNB should be performed as a staging procedure in every case of PEM.
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REFERENCES