Adrenal Cortical Carcinoma

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• CASE REPORT •

Cutaneous Adrenal Cortical Carcinoma Metastasis in a 6 year-old Boy

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We report the case of a six year-old boy with a history of adrenal cortical carcinoma presenting with cutaneous metastasis. Due to the low incidence of cutaneous metastasis arising from adrenal cortical carcinoma, its diagnosis can be challenging based solely on histological analysis. Yet, the clinical history in combination with an immunohistochemical panel consisting of melan-A, alpha inhibin, D11, caretinin, neuron specific enolase, synatophysin, and chromogranin, can be useful in differentiating it from other tumors with similar cytomorphology. [P R Health Sci J 2010;3:330-331]

Key words: Adrenal cortical carcinoma, Cutaneous metastasis, Immunohistochemistry

A drenal cortical carcinoma (ACC) is an uncommon aggressive malignancy with a bimodal age distribution with a first peak in childhood and a second in the fifth decade. Its prognosis largely depends on tumor stage. The most common sites of metastasis include liver, local lymph nodes, lungs, peritoneum, and bone. There are only rare case reports of ACC with cutaneous metastasis (1). We report the case of a six year-old boy with history of adrenal cortical carcinoma who developed cutaneous metastasis and confirm the usefulness of a previously described immunohistochemical panel to help differentiate it histopathologically from other tumors with similar cytomorphology.

Case Report

A six year-old boy with history of precocious puberty and stage IV ACC status post resection, with lung, brain, and heart metastasis, was admitted to the hospital for his fifth cycle of chemotherapy with etoposide, doxorubicin, and mitotane. The dermatology service was consulted for an asymptomatic lesion over the left lower border of the scapula that had been present for two weeks. On examination, there was an isolated 1.0 cm diameter, firm, subcutaneous nodule with hyperpigmentation (Figure 1).

Histopathological examination revealed a neoplasm located in the deep dermis composed of nodules of large cells with a compact eosinophilic cytoplasm (Figure 2). The differential diagnosis at this point included a metastatic lesion from ACC versus renal cell carcinoma vs melanoma. Immunohistochemistry revealed weakly positive neuron-specific enolase (NSE), focally positive alpha inhibin and synatophysin, yet negative chromogranin. The clinical, histological, and immunohistochemical findings were consistent with metastatic adrenal cortical carcinoma.

Discussion

Given the rarity of adrenal cortical carcinoma, cutaneous metastasis are uncommon compared to other neoplasms that spread to the skin. Therefore, it is unusual to entertain it in the differential diagnosis of cutaneous metastatic tumors. Given the lack of specific immunohistochemical stains, a panel with markers used in tumors with similar cytomorphological features (adrenal cortical adenoma, pheochromocytoma, metastatic renal cell carcinoma, and metastatic melanoma) is often helpful in differentiating ACC (Table 1).

Alpha inhibin, a glycoprotein expressed by sex cord stromal tumors, is sensitive for ACC. Yet, it is not very specific as it also stains sex cord stromal tumors and pituitary adenomas (1-2). A103, a monoclonal antibody against epitopes on recombinant Melan-A, constitutes a more specific stain. Melan-A is an antigen recognized by cytotoxic T-cells initially thought to be restricted to melanocytic tumors, but later on recognized to be present on steroid hormone-producing neoplasms including adrenocortical tumors (2). It is positive in both ACC and melanoma, yet the former is S-100 and HMB-45 negative (3).

D11, a molecule that labels the nuclei of adrenal cortical cells, can be utilized to identify ACC, yet it lacks specificity...
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inhibin, D11, calretinin, NSE, synaptophysin and chromogranin can be helpful in establishing the diagnosis.

Resumen

Reportamos el caso de un niño de seis años de edad con historia de carcinoma adrenocortical que presenta con una metástasis cutánea. Debido a la incidencia baja de metástasis cutáneas que surgen del carcinoma adrenocortical, establecer su diagnóstico puede ser un reto basado solo en el examen histológico. El historial clínico en combinación con un panel de inmunohistoquímica consistiendo de melan-A, inhibina alfa, D11, calretinina, enolasa neural específica, sinatofisina y cromogranina, pueden ser útiles para diferenciarlo de otros tumores con citomorfología semejante.

References