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Retrospective analysis of prognostic factors in pediatric patients with adrenocortical tumor from unique tertiary center with long-term follow-up

Pediatric adrenocortical tumors (PACTs) represent rare causes of malignancies. However, the south/southeast regions of Brazil are known to have a high incidence of PACTs because of the founder effect associated with a germline pathogenic variant of tumor suppressor gene TP53. We aimed to retrospectively analyze the types of variables among hormone production, radiological imaging, tumor staging, histological and genetic features that were associated with the occurrence of malignancy in 95 patients (71% females) with PACTs from a unique center. The worst prognosis was associated with those aged > 3 years ($p < 0.05$), high serum levels of 11-desoxicortisol ($p < 0.001$), tumor weight ≥ 200 g ($p < 0.001$), tumor size ≥ 5 cm ($p < 0.05$), Weiss score ≥ 5 ($p < 0.05$), Wieneske index ≥ 3 ($p < 0.001$) and Ki67 $\geq 15\%$ ($p < 0.05$). Furthermore, patients with MacFarlane stage IV had an overall survival rate almost two times shorter than patients with other stages ($p < 0.001$). Additionally, the subtractions of BUB1B-PINK1 (< 6.95) expression ($p < 0.05$) and IGF-IR overexpression ($p = 0.0001$) were associated with malignant behavior. These results helped identify patients who are likely to have an aggressive course; further multicenter prospective studies are required to confirm our results. In conclusion, PACTs with these patterns of prognostic factors could be treated using an adjuvant approach that may improve the overall survival in such patients.

Keywords: pediatric adrenocortical tumor; prognostic factors; pediatric cancer; TP53

Biography:

Maria Candida is a Professor at the University of Sao Paulo, Brazil, where she also serves as the Head of the Adrenal Unit of the Discipline of Endocrinology and Metabolism at Hospital das Clínicas, Medical School, USP. Additionally, she holds the position of Assistant Physician and is the Principal Investigator for Adrenocortical Cancer Studies at the Cancer Institute of the State of São Paulo (ICESP). She actively conducts research as a member of the Laboratory of Hormones and Molecular Genetics (LIM42 – HC-FMUSP). Her exceptional contributions have been recognized with a CNPq Senior Research Productivity Scholarship.