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Poster Presentation

Pancreatoblastoma Presenting as a Rapidly Enlarging Pancreatic Mass in a Child: A Diagnostic Challenge

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ABSTRACT

Introduction. Pancreatoblastoma is a rare tumor predominantly affecting pediatric population and accounts for 0.5% of all pancreatic tumors. Overlapping histologic features with other neoplasms in a limited sample poses a diagnostic challenge in rendering final diagnosis for appropriate management.

Case Description. A 6-year-old female presented with abdominal mass. Imaging revealed a pancreatic mass initially measuring 8.9 cm enlarging to 20.8 cm in the span of 3 months with elevated serum AFP levels. Biopsy and subsequent immunohistochemistry was interpreted as malignant round cell neoplasm. Neoadjuvant chemotherapy was initiated and underwent distal pancreatectomy. Grossly, there was an ill defined, tan, solid mass measuring 10.0 x 8.0 x 8.0 cm and histology revealed lobules separated by dense fibrous bands with acinar and trabecular patterns and featured prominent squamous morules. Patient was discharged stable with normalization of serum AFP and started adjuvant chemotherapy.

Discussion. Biopsy showed round cells which are beta-catenin positive and synaptophysin, chromogranin A negative with an impression of solid pseudopapillary neoplasm versus pancreatoblastoma. Thorough sampling of the resection specimen showed lobules of malignant cells positive for pancytokeratin and E-cadherin confirming epithelial differentiation and the diagnostic squamous morules showed CD10 positivity and a distinct pattern of both nuclear and cytoplasmic beta-catenin expression supporting the activation of Wnt pathway that is typical of pancreatoblastoma.

Conclusion. Limited tissues due to large tumor size and core biopsy sampling did not reveal the characteristic squamous morules, thus having a diagnostic challenge. Comprehensive evaluation of the resected specimen addressed this gap by having more tumoral areas and specific tissues subjected to immunohistochemistry studies.

Key words. *pancreatic tumor, pancreatoblastoma, pediatric, serum AFP, squamous morules*