

## Response to Capecitabine and Temozolomide in an Adolescent Female with Metastatic, Non-functional Pancreatic Neuroendocrine Tumor

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**Background:** Pancreatic neuroendocrine tumors are rare with an estimated overall incidence of <1/100,000. Aggressive surgical resection is advocated as first line therapy. However, approximately half of tumors are metastatic at presentation limiting the ability for complete resection.

A 13-year-old African American female presented with two years of intermittent, dull, epigastric pain. CT scan revealed a 2 cm mass in the pancreatic tail and multiple hepatic lesions. Imaging with PET scan, bone scan, and spine MRI revealed involvement of multiple thoracic and lumbar vertebrae. Liver lesion biopsy was consistent with metastatic pancreatic neuroendocrine tumor (mPNET). Octreoscan revealed no uptake within the pancreatic mass. The family initially declined chemotherapy due to concerns of quality of life. Her pain subsequently worsened requiring narcotics. Fourteen months after diagnosis, she suffered a pathologic fracture of her right femoral neck requiring hemiarthroplasty. Given symptomatology and progressive disease, they agreed to a chemotherapeutic regimen, 15 months after diagnosis, with capecitabine 750 mg/m<sup>2</sup>/dose twice daily for 14 days and temozolomide 250 mg/m<sup>2</sup>/dose daily on days 10-14. She had no adverse events attributed to therapy. CT scan after two cycles showed decreased size of a left breast lesion (2.3 cm to 2 cm), a right anterior mediastinal mass (4.2 x 3.4 cm to 2.5 cm x 1.7 cm), and a left hepatic lobe lesion (14.6 cm to 12 cm). Representative images are shown in Figure 1. The patient received two additional cycles of therapy prior to further disease progression and death 20 months after diagnosis.

**Conclusions:** This case highlights the hesitance of patients to receive chemotherapy for a rare, and slow growing, but fatal disease when little literature exists in the pediatric population. In addition, it demonstrates the feasibility, disease response, and minimal toxicities associated with this regimen in a pediatric patient with extensive mPNET treated late in the disease process.

**Figure 1:** A and B are contrasted enhanced axial CT images obtained prior to treatment demonstrating a necrotic anterior mediastinal mass and extensive hepatic metastasis with a large dominant conglomerative mass in the left hepatic lobe, respectively. C and D depict visible decrease in size of these lesions after 2 cycles of chemotherapy.

