Doege-Potter Syndrome: A Case of Tumor-Induced Hypoglycemia

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Introduction:
Doege-Potter syndrome is a paraneoplastic syndrome characterized by hypoglycemia in association with solitary fibrous tumor. Hypoglycemia results from tumor secretion of IGF-2, a hormone with structural similarity to insulin. Definitive treatment of SFT is surgical resection; however, in unresectable cases, systemic therapy and selective embolization can be considered to slow tumor progression, while glucocorticoids can be used to treat hypoglycemia. This report presents the case of a patient with Doege-Potter syndrome secondary to SFT of the brain with metastases to the liver. He had experienced disease progression despite multiple chemo and immunotherapy agents, and was treated with oral steroids and transarterial chemoembolization (TACE) to his liver lesion.

Case Presentation:
A 54 year-old man with solitary fibrous tumor (SFT) of the brain and metastases to the liver developed acute onset right-sided hemiparesis, confusion, and dysarthria. EMS was called, and blood glucose was noted to be 38. The patient was given D50 during transport to the hospital with improvement in his blood sugar and resolution of his neurologic symptoms. He described slightly diminished oral intake over the preceding week but was able to eat a full meal a few hours prior to symptom onset. He had no history of diabetes or recent medication changes. In the emergency department, his initial glucose after D50 was 102 but then dropped to 50 on subsequent check. He was started on D5LR at 125cc/hr and admitted to the hospital. Non-contrasted head CT scan showed no abnormalities. Over the next 48 hours, he had recurrent hypoglycemia when the D5LR infusion was weaned. Lab workup revealed normal liver function tests, Hgb A1c of 4.2%, low serum insulin level (<1.0), low serum C-peptide (0.5 ng/mL), and low insulin-like growth factor 1 (IGF-1) (21 ng/mL). Cortisol stimulation test demonstrated appropriate rise in cortisol levels. Infectious workup including UA, CXR, and blood cultures were unrevealing. CT abdomen/pelvis demonstrated multiple hepatic metastases from SFT.

Final Working Diagnosis:
Doege-Potter syndrome secondary to solitary fibrous tumor

Management/Follow-Up:
In this case, the patient had previously received bevacizumab/temozolomide, pazopanib, sorafenib, and trabectedin, and had disease progression with all of these agents. Due to need to control the SFT in order to wean the glucose drip, the patient was evaluated by surgical oncology and interventional radiology. His disease was not considered resectable, and the patient underwent transarterial chemoembolization (TACE) of the left lobe of the liver. He was started on oral steroids with the hope of suppressing IGF-2 (3, 7). Over a few weeks, the steroids were weaned. The patient later received a second TACE to the right lobe of the liver; however, within two months he began to experience recurrence of his hypoglycemia and was restarted on an increased steroid dose. He continues to follow with his primary oncologist and will undergo radiation therapy to his largest liver lesion in an attempt to treat both his disease progression and hypoglycemia.

Learning Objectives
Describe the workup and differential diagnosis of hypoglycemia
Explain the pathophysiology of Doege-Potter syndrome
List current treatment strategies for Doege-Potter syndrome and solitary fibrous tumor