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Case Report

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Intra-Abdominal Solitary Fibrous Tumor secretes high molecular weight IGF-II, causing Hypoglycemia

Abhishiek Setia*, Martha Bilbatua, Sarahi Herrera-Gonzalez and Mindy Griffith

Department of Internal Medicine, Jersey City Medical Center, Jersey City NJ, 07302, USA

***Corresponding author**
Abhishiek Setia, Department of Internal Medicine, Jersey City Medical Center, Jersey City NJ, 07302, USA

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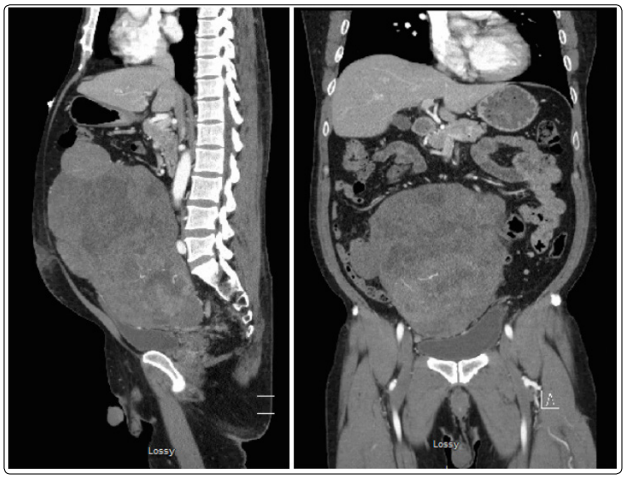
Background

Hypoglycemia is a common event that has many well-known causes. Nevertheless, Non-islet cell tumor hypoglycemia (NICTH) is not one of them. As a matter of fact, NICTH is a rare but serious paraneoplastic syndrome. Solitary Fibrous tumor (SFT) is a rare spindle cell neoplasm derived from mesenchymal cells that in just 5% of cases releases "The Big" Insulin-like Growth Factor (IGF) II leading to hypoglycemic events [1].

Case Report

Our patient is a 53-year-old male with Hypertension that presented to the Emergency Department with a 3-week history of intermittent episodes of confusion witnessed by family members and coworkers, as well as abdominal discomfort. He reported some abdominal fullness and weight gain, but denied tenderness, nausea, vomiting or diarrhea. Physical exam revealed a large hard abdomino-pelvic mass that on Computed Tomography (CT) was revealed to be a heterogeneous abdomino-pelvic mass with calcifications and some areas of hyper vascularity measuring approximately 15 x 23 x 26 cm, extending superiorly to the undersurface of the transverse colon and inferiorly into the right pelvis with mass effect upon the bladder (Figure 1).



Patient was found to have multiple episodes of hypoglycemia with glucose levels of 20-40 mg/dL and was started on dextrose infusion and a high starch and protein diet. Laboratory test were relevant for Insulin level of 2.3 mUI/mL (2-19.6 mUI/mL), C- peptide level of 0.44 ng/mL (0 .8-3.85 ng/mL) and IGF-II of 1,230 ng/mL (267-616 ng/mL). A CT guided biopsy was done and showed a low grade Spindle Cell Tumor.

Total mass excision with repair of the bladder and colonic wall, as well as appendectomy were done by exploratory laparotomy with no surgical complications. No further episodes of hypoglycemia were noted post surgically. The final surgical pathology report demonstrated a SFT.

Discussion

SFT, present as pedunculated and well-circumscribed masses in association with serous cavities, given its preference for the pleura, only 15-18% of the cases have described extra-thoracic tumors. STF are mostly asymptomatic and are found incidentally, unless they are associated with cough, chest pain, and abdominal pain [2]. Hypoglycemia, seen in approximately only 5% of cases of this already rare tumor, was described by Doege and Potter in the 1930's, hence its name [4]. It has been reported mostly in masses >10 cm and tends to resolve after complete surgical resection, if technically possible and if no metastatic disease is present [4].

The tumor secretes a precursor molecule of IGF II, known as "Big IGF-II" for its high molecular weight, which binds IGF I and insulin receptors in glucose avid tissues and the mass itself causing episodic hypoglycemia. During said episodes, there has been documented suppression of Insulin, pro-insulin, C peptide, Growth hormone (GH) and IGF I, for which both endogenous and exogenous insulin or insulin secretagogues are generally ruled out as causes of such severe hypoglycemic episodes during initial workup. If association of a spindle cell tumor with hypoglycemia is seen, a useful tool to prove causality is a IGF II:GF I ratio >10, that is now identified as a pathognomonic finding of this syndrome given that elevation of IGF II works as negative feedback decreasing GH production and IGF I, subsequently [3, 4].

As mentioned above, surgical resection of the tumor is curative of hypoglycemia in most cases. While awaiting for surgical intervention, episodes are prevented and treated with frequent high glucose meals, with or without continuous or as-needed dextrose infusions, which prove helpful in overnight events. Oral corticosteroids have also been

used in preventing hypoglycemic episodes and appear to normalize IGF levels [1, 4]. Recurrence of tumor after resection is frequent and the mass should be completely excised in the first attempt in an effort to prevent this from occurring, especially when a second intervention would be difficult in some debilitated patients [5].

Conclusion

SFT are rare tumors associated with an even more uncommon paraneoplastic syndrome causing hypoglycemia known as Doege-Potter syndrome. We present a patient with Whipple's triad, biochemical proof of elevated IGF II and a spindle cell neoplasm in a particularly unusual location, the abdomino-pelvic cavity. As per seen before in the literature, our patient's hypoglycemia improved with intravenous glucose infusions and resolved after surgical resection of the tumor, proving that this peculiar tumor and its associated syndrome can be readily treated.

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