A RARE CAUSE OF PEDIATRIC HYPOGLYCEMIA IN A BOY: A MALIGNAT INSULINOMA

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P2- 147

BACKGROUND

Insulinoma is a rare neuroendocrine tumor, usually benign, but can be life-threatening in causing hypoglycemic accidents. It presents in individuals aged between 8 and 82 years and can occur sporadically or constitute a part of multiple endocrine neoplasia type 1 (MEN-1).The nonspecific-symptoms and small size of these tumors led to difficulties of diagnosis and localization.

CASE REPORT

Here we describe the case of a 11-year-old boy, who came to our attention for a 1-month history of confusion, amnesia and diplopia, occurring during severe hypoglycemia. His past medical history revealed three simple febrile seizure in the first two years of life.
At presentation, patient was vitally and hemodynamically stable. Cardiovascular, respiratory, gastrointestinal and neurological examinations were normal.
Initial laboratory investigations showed low blood glucose (48 mg/dL) and high blood insulin levels (62.74 μIU/ml) with detectable C-peptide values. Complete blood count, hepatic, renal, serum electrolytes, thyroid, parathyroid and prolactin profiles were normal. Urinary sample was negative for ketones. During hospitalization, glucose monitoring showed fasting low glucose levels (32 mg/dl). As very low serum glucose levels were associated with inappropriately high serum insulin and detectable C-peptide levels, the diagnosis of endogenous hyperinsulinism was made.
In view of hypoglycemic symptoms and possible insulinoma, patient underwent to radiological investigations. The abdominal contrast–enhanced MR showed a 40 mm, well-demarcated lesion within the head of pancreas associated with mild dilatation of pancreatic duct and retropancreatic lymphadenopathy (34x10 mm). No distant metastasis was identified.
After diagnosis confirmation, a continuous infusion of 10% glucose was started to maintain normal serum glucose levels . The patient underwent duodenocephalopancreasectomy after two weeks.
The definitive histological examination revealed a neuroendocrine tumor, well differentiated (NET-G2 sec. WHO 2010) of 4,5 cm infiltrating vascular and perineural districts, with metastasis in 2 pancreatic-duodenal lymph nodes among the 9 that had been surgically removed. Immunohistochemical staining was positive for synaptophysin (replicative index Ki67:12%). The most recent abdominal revalutation did not reveal any recurrence of disease and currently the patient is in good conditions and with normal glycemic levels .

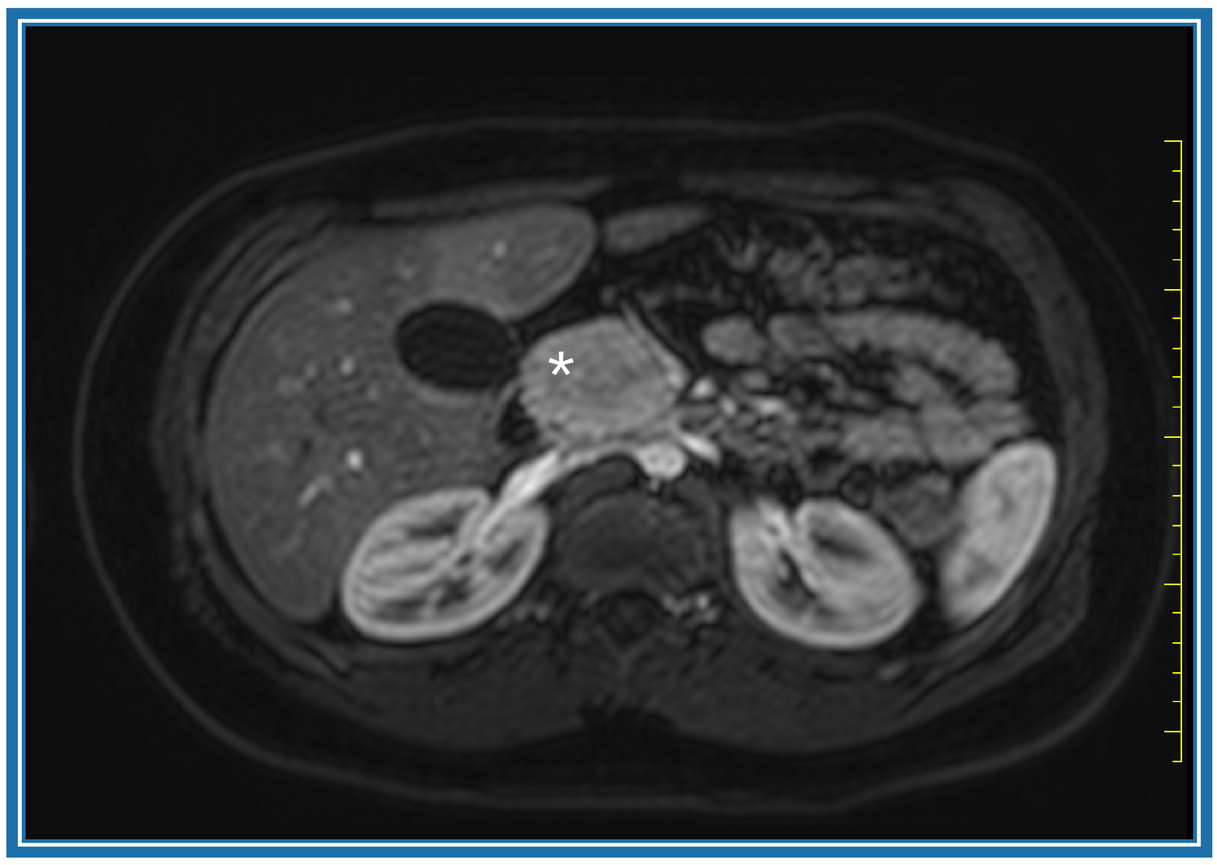

Figure 2. Magnetic resonance imaging of the tumor within the head of pancreas.

CONCLUSIONS

Insulinoma in childhood is generally rare. Even if the vast majority of them (90%) are benign, a small percentage (10%) may have a malignant behavior. Clinicians must maintain a high index of suspicion for insulinoma in the presence of young patients with frequent hypoglycemic symptoms because early recognition is important to ensure proper surgical treatment and prevent serious adverse neurological consequences.

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