2023 ANNUAL CONFERENCE PESTOLA
PEDIATRIC ENDOCRINE SOCIETY OF TEXAS, OKLAHOMA, LOUISIANA, AND ARKANSAS
FEBRUARY 3-5 | THE SKIRVIN HILTON OKLAHOMA CITY, OK
A HYBRID EVENT

Insulinoma As A Rare Cause Of Behavioral Change In A Teenager

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Introduction

Insulinomas are extremely rare with an incidence of 1-4 cases occurring per million persons per year, and only 10% of patients with insulinoma have multiple endocrine neoplasia Type 1 (MEN1). We describe a case of a boy with MEN1 and insulinoma who presented atypically with abnormal behavior.

Case Presentation

History

•13-year-old African American male presented with abnormal behaviors/movements for 8-12 months, worse since the last 3 days.
•Episodes involved “spacing out,” “talking gibberish,” chewing movements of the mouth, and unresponsiveness.
•Occurred more often in the morning and sometimes improved after eating.
•School performance had declined, and mother and patient denied substance abuse.
•Brought to ER by EMS for unresponsiveness
•In ER, Blood glucose <25 mg/dl. His mental status improved after an initial dextrose bolus.

Physical Examination

- His vitals were stable and examination was unremarkable except some bilateral hand weakness.

Hospital Course

Multiple low Blood glucoses after stopping D10 fluids
↓
Critical sample suggestive of endogenous hyperinsulinism
↓
MRI Pancreas showed pancreatic mass suggestive of insulinoma
↓
Diazoxide was started at 100 mg q8 , ie 5 mg/kg/day
↓
Discharged on Day 2 after starting Diazoxide
↓
Laparoscopic distal pancreatectomy done 2 weeks later
↓
No low blood glucoses after the procedure
Diazoxide discontinued after surgery

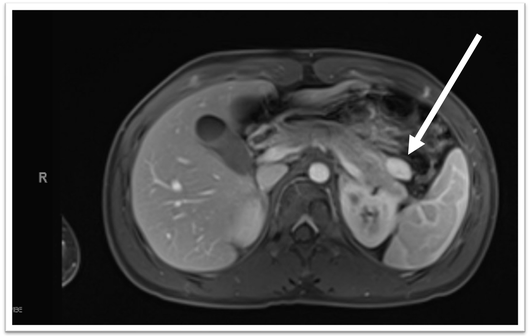
Genotyping

Genetic testing for MEN1 done due to multifocal lesions on pathology - heterozygous point mutation in the menin gene (c.783+1G>A)- Suggestive of MEN1

Table 1. Hormonal work up

Test	Normal reference Values	1st Critical Sample	2nd Critical Sample
Cap BG	70-106 mg/dl	46 mg/dl	48 mg/dl
Serum BG	70-106 mg/dl	55 mg/dl	57 mg/L
BOH	0.-2.8 mmol/L	<0.1 mmol/L	0.1 mmol/L
Insulin	**3-25 mU/L**	**56.6 mU/L**	**17.6 mU/L**
C- peptide	**0.4-2.2 ng/ml**	**-**	**2.3 ng/ml**
Cortisol	5.9-22.5 mcg/dL	-	6.02 mcg/dL
Growth hormone	0.05-11 ng/ml	-	9.65
Sulphonyl urea panel		Negative	Not sent
Proinsulin	0-10 pmol/L	-	21.9 pmol/L

Figure 1. 2 cm mass in the pancreatic tail, with imaging features most consistent with an insulinoma.



Discussion

- Insulinomas are the **most frequent functioning pancreatic Neuroendocrine tumors (NET) in children** with Multiple Endocrine Neoplasia (MEN1).
- Present with hypoglycemic symptoms that improve with glucose intake, thus fulfilling Whipple’s triad.
- Medical treatment with frequent carbohydrate meals, diazoxide, and/or somatostatin analogs is not always successful.
- Surgery is considered the standard of care.
- Our patient is doing well after the laparoscopic distal pancreatectomy, no residual neurological deficits, and had no further symptoms of hypoglycemia.
- Pathology revealed multifocal neuroendocrine tumors with islet cell hyperplasia.
- Initial screening for hyperparathyroidism and other neuroendocrine tumors was negative.

Conclusion

This case highlights the importance of assessing for hypoglycemia in uncommon presentations, such as abnormal behavior, that can be suggestive of hypoglycemia. For our patient, identifying hypoglycemia led to the diagnoses of insulinoma and MEN1, both of which have significant implications for patients and their families.

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