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Diabetes Mellitus in Neurofibromatosis I: An Unusual Presentation

Neurofibromatosis (NF) are a set of genetic disorders resulting from abnormalities of neural crest development or 'neurocristopathy'. They can affect the development of non-nervous tissue such as bones, soft tissue, endocrine glands and skin. We report a case of NF I with diabetes mellitus. To the best of our knowledge diabetes mellitus in association with NF 1, has not so far been reported in children. A 9-year-old male presented with general debility, fever off and on since birth, polydipsia, polyuria, and polyphagia for the last 3 months. There was no history of diarrhea, steatorrhea, jaundice, abdominal pain, headache or seizures. He was found to be both underweight and short statured. Multiple (>15) Cafe-au-lait spots measuring 2 mm to 20 mm and axillary freckling was present. Two large naevi with underlying neurofibromas covered the lower back and gluteal region. Eye examination revealed bilateral Iris Lisch nodules, and pigmentary changes in the fundus. The left lower limb was found to be 6cm longer than the right. Systemic examination did not reveal any abnormality. The child was normotensive, had a cheerful isposition, average intelligence and showed no psychological or behavioral problems. Investigations revealed a random blood glucose 830 mg/dL, and fasting blood glucose 307 mg/dL. Urine examination showed specific gravity 1.040, glucosuria 4+, and traces of ketone bodies. No abnormality was detected on ultrasonography or CT scan of the abdomen or duodenal endoscopy. MRI abdomen could not be performed due to financial constraints of the patient. The patient was put on insulin therapy and normoglycemia was maintained on 0.8 units of insulin / kg / day.

The diagnosis of NF type I is based largely on clinical criteria set up by National Institute of Health, 1987(1). The condition is associated with protean manifestations as well as some serious complications. In 1983 a syndrome of multiple endocrine neoplasia was described as MEN type III which included duodenal carcinoid (often producing somatostatin) and NF type I or pheochromocytoma(2). Unlike other endocrinal abnormalities, diabetes mellitus is rarely seen in association with NF I. It is attributed to occurance of somatostatinomas in pancreas and duodenum. These are rare gut-pancreatic endocrinomas that secrete somatostatin. Inhibition of insulin release produces the diabetic state which is easy to control because of concomitant suppression of glucagon release. Most somatostatinomas in NF I are duodenal in location(3). Duodenal

somatostatinomas are rare neuroendocrine generally non- functioning well differentiated tumors with a low grade of malignancy and are less frequently metastatic(4). These rare neoplasms have been reported only in adults and old patients. In contrast to pancreatic tumors, duodenal tumors are seldom associated with 'somatostatin syndrome' (diabetes mellitus, diarrhea, and cholelithiasis). They are smaller in size, and give rise to symptoms like abdominal pain or lump, gastrointestinal hemorrhage and obstructive jaundice(3). A case has, however, been reported where partial somatostatinoma syndrome was found along with duodenal carcinoid(5). In yet another case all the 3 features of somatostatin syndrome were present in association with the same tumor. Most tumors located in the head of pancreas are malignant with metastasis to liver. Pancreatic tumors and metastasis are equally well detected by CT and MRI. They are echo poor and can sometimes be missed on USG (especially those located in the tail).

Endoscopic evaluation is often required for detection of duodenal carcinoids as they can be missed on MRI or other radiographic techniques. Clinical and hormonal features of somatostatinoma syndrome are not requisite for diagnosis, analysis of plasma somatostatin immunoreactivity might lead to a higher detection rate of the tumor. Hypersomatostatinemia demonstrated by Calcium-pentagastrin test or tolbutamide test is very useful for diagnosis. Tumor is further confirmed by immunohistochemical studies. Somatostatin receptor scintigraphy and octreotide therapy is a newer modality for diagnosis and treatment.

Zeeba Zaka-ur-Rab,
Kamlesh Chopra

Correspondence to:
Dr. Zeeba Zaka-ur-Rab,
Assistant Professor,
Department of Pediatrics,
Himalayan Institute of Medical Sciences,
Swami Ram Nagar, Dehradun 288 140,
Uttaranchal,
India.

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