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Retroperitoneal Ganglioneuroblastoma Presenting as Chronic Diarrhea and Refractory Hypokalemia in a Child

Abstract

Neuroblastoma is the most common extracranial solid tumor in children. Neuroblastoma presenting with diarrhea as the predominant symptom is rare and can be a diagnostic challenge to the paediatrician. We present a twenty months old child with Retroperitoneal Ganglioneuroblastoma who presented with chronic diarrhea and refractory hypokalemia. Tumor removal resulted in cessation of diarrhea and correction of hypokalemia.

Keywords: *Chronic diarrhea, ganglioneuroblastoma, hypokalemia*

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Introduction

Neuroblastoma may develop at any site of sympathetic nervous system tissue. The signs and symptoms of neuroblastoma reflect the tumor site and the extent of disease. The symptoms may mimic many other disorders, a fact that can result in a delayed diagnosis.[1] Intractable diarrhea due to tumor secretion of vasoactive intestinal peptide (VIP) is one of the paraneoplastic syndromes associated with neuroblastoma. We present a child in whom a detailed evaluation for intractable diarrhea and hypokalemia led to the diagnosis of a retroperitoneal ganglioneuroblastoma.

Case Report

A 20-month-old girl child was admitted with complaints of watery diarrhea of 3-month duration. Diarrhea was associated with abdominal distension and documented weight loss. The loose stools were watery, 5–6 times a day, not associated with blood or mucus. As there was an increase in the frequency of loose stools associated with decreased urine output and lethargy for 1 day, she was brought to us. On examination, the child was hypotonic, undernourished, and weighing 7.8 kg. Signs of some dehydration were present, and the abdomen was distended. Laboratories done at admission showed severe hypokalemia (1.8 mEq/dl), metabolic acidosis (11 mEq/dl), and normal chloride (100 mEq/dl). The child's dehydration was corrected with isotonic saline, and potassium infusion was given under cardiac monitoring. Serial monitoring of serum electrolytes was done. During the hospital stay, diarrhea persisted and serial laboratories showed persisting hypokalemia (<2.5 mEq/dl) and acidosis (<15 mEq/dl). Dyselectrolytemia persisted despite adequate hydration along with potassium and bicarbonate supplementation. Detailed etiological workup for chronic diarrhea was done. Stool microscopy was normal. Stool viral and bacteriological profile was negative. Ultrasonography (USG) abdomen was normal. Upper gastrointestinal (GI) endoscopy and colonoscopy were normal. Duodenal and intestinal biopsy Histopathology (HPE) was normal, and there was no evidence of villous atrophy or inflammation or autoimmune enteropathy. Urine electrolytes were normal, and there was no evidence of renal loss. Serum magnesium was normal. Screening for underlying immunodeficiency was negative. As detailed etiological workup was negative, we considered the rare possibility of diarrhea secondary to a VIP-secreting tumor. Clinically, intermittent flushing was noted. Blood pressure and heart rate were normal. Twenty-four-hour urinary vanillylmandelic acid (VMA) was normal. Although USG abdomen, chest X-ray, and 24-h urinary

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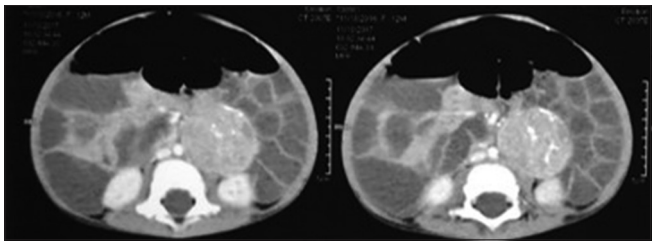

Figure 1: Computed tomography abdomen showing heterogeneous mass lesion with calcification in the left para-aortic region

VMA were normal, in view of strong clinical suspicion, we proceeded with positron-emission tomography (PET) scan. PET scan identified a well-defined metabolically active soft-tissue density lesion in the retroperitoneum in the para-aortic region. Calcification was noted within the lesion, and there was no para-aortic lymphadenopathy or focal liver lesions or bony metastasis. Contrast-enhanced computed tomography (CT) abdomen [Figure 1] confirmed a 37 mm × 38 mm × 49 mm heterogeneous mass lesion in the left para-aortic region below the left renal vessels. Bone marrow biopsy showed trilineage hematopoiesis. Laparotomy and *in toto* excision of tumor mass were done. Histopathology of the tumor mass was consistent with ganglioneuroblastoma-mixed nodularity of Schwannian stroma-poor type. N-Myc amplification was negative. After tumor removal, diarrhea settled, hypokalemia and acidosis improved, and there was documented weight gain.

Discussion

Neurological symptoms such as opsoclonus myoclonus are the most common paraneoplastic syndrome in patients with neuroblastoma.[2] Watery diarrhea, hypokalemia, and achlorhydria syndrome was first described by Verner and Morrison as a rare cause of chronic secretory diarrhea secondary to a VIP-secreting tumor.[3] Diarrhea can be a symptom of paraneoplastic syndrome of neuroblastoma. There is evidence that this symptom is related to VIP production in tumor cells stimulating intestinal aberrant secretions.[4] In a review by Zhang *et al*., out of 9 children who presented exclusively with symptoms of paraneoplastic syndrome, ganglioneuroblastoma was found to present predominantly with GI symptoms as a paraneoplastic syndrome.[5] CT and MRI are the recommended imaging modality for diagnosis and staging of tumor.[6] In our child, the tumor was not identified on USG abdomen. The presence of dilated bowel loops could have interfered with the imaging of the retroperitoneal mass. In view of strong clinical suspicion, we proceeded with PET scan which identified the tumor mass in the abdomen. Tumors, which present with diarrhea as paraneoplastic syndrome, are generally biologically favorable. The prognosis is good following appropriate treatment.[7] In our child, excision of the tumor mass resulted in complete recovery.

Conclusion

In any child who presents with chronic secretory diarrhea, hypokalemia, and failure to thrive, the possibility of an underlying VIP-secreting tumor should be considered. A high index of clinical suspicion is needed as early diagnosis results in good clinical outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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