

# A Rare Case of Ganglioneuroblastoma with Presentation of Diarrhea and Resistant Hypokalemia

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## Abstract

**Background:** Neuroblastoma is the most common extracranial solid tumor in children and the most common malignancy in infant. Some tumors release vasoactive intestinal peptide (VIP), causing a profound secretory diarrhea. **Case Report:** In this report we describe a 33 month old girl with intermittent diarrhea from 9 month ago and hypokalemia resistant to treatment. She had increased VMA in 24hour urine. After complete workup she had a mass in her thorax that resected and her diarrhea and hypokalemia was improved. **Conclusion:** In children with resistant diarrhea we should consider tumors secreting VIP.

**Keywords:** Neuroblastoma, Diarrhea, Hypokalemia.

## INTRODUCTION

Neuroblastoma (NB) is the most common extracranial solid tumor in children. The site of primary tumor can be in any part of the sympathetic chain or the adrenal medulla [1]. The tumor can be found in the abdomen (75%), mediastinum (20%) and the neck (5%) [2]. It can be primary or metastatic [3]. The signs are palpable mass in the abdomen or neck, loss of appetite and vomiting (in abdominal tumors), shortness of breath, pulmonary infections, and dysphagia (in Chest tumors), anemia and petechiae (in bone-marrow involvement). The initial presentation can be metastatic lesions of liver, lung or bone. Excretion of catecholamine and VIP can lead to sweating, pallor, hypertension and watery diarrhea [4]. Patients diagnosed under 1 years of age (40%) have a much better prognosis than those diagnosed in later childhood [5]. The diagnosis is difficult because of wide range of clinical presentations. The methods to diagnosis are ultrasound, CT scan and urine VMA analysis. Biopsy from the metastatic lesions can confirm the diagnosis [6]. Depending on the stage of tumor the treatment includes surgical excision, multi agent chemotherapy, and bone marrow transplantation [6]. Patients with diarrhea as the main symptom, namely diarrheal NB that is rare [7]. Diarrheal NB is not easy to

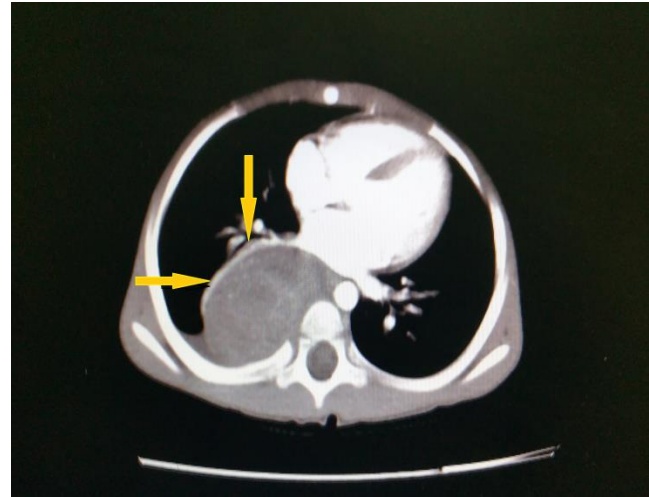
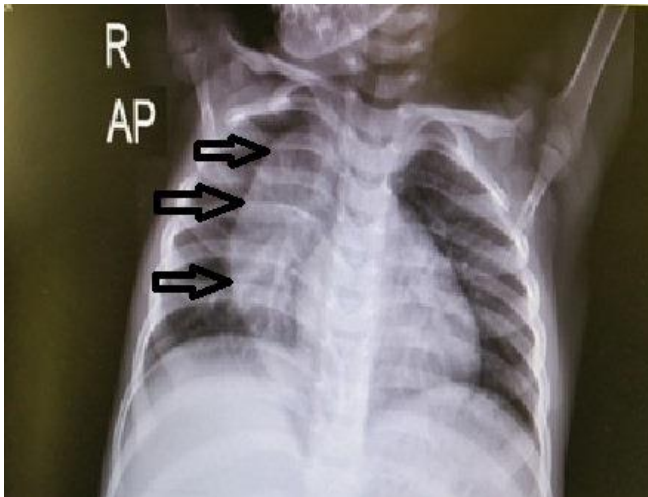
diagnose in the early stage of the disease [8]. The initial clinical manifestations are intractable diarrhea, hypokalemia, and dysplasia. In these patient's VIP has increased. Potassium supplementation plays a vital role in the treatment. On the condition of appropriate treatment, diarrheal NB has a good prognosis [8]. With surgical excision of the tumor, diarrhea resolute completely [9].

## CASE REPORT

We are reporting a 33-month-old girl with the complaint of intermittent diarrhea from 9 month ago. Her diarrhea was watery and voluminous, 3-4 times a day. Also, she had sometimes vomiting and abdominal pain and blowing. She had 4 kilogram decrease in weight duration of 9 month. In laboratory tests she had hypokalemia and hyponatremia resistant to treatment. Her diarrhea was improved with NPO and Octerotide. Her sonography revealed distended loops with much fluid. In her 24hour urine had increased VMA. In her endoscopy esophagus and stomach and D1 were normal but D2 had decrease in the size of villus and scalloping in some points. In her colonoscopy had nodularity in descending colon and decrease of vascular pattern. A chest CT was performed for her. The findings were a large posterior mediastinal right paravertebral soft tissue mass

about 49x41x65mm with fine calcified foci, minimal reactive pleural effusion without evidence of lung involvement that was most likely suspected mediastinal neuroblastoma. In MRI the mass was hyposignal in T1 and hypersignal in T2 with spreading to spinal cord from neural foraments that had changed the site

of techal sac but had not pressure effect on the neural cord. At the end the mass was excised totally from right thoracotomy and her diarrhea and hypokalemia was improved. The pathology of the tumor was ganglioneuroblastoma.



**Figure 1:** large posterior mediastinal right paravertebral soft tissue mass with fine calcified foci, minimal reactive pleural effusion and no evidence of lung involvement

## DISCUSSION

Neuroblastoma (NB) is the most common extracranial solid tumor in children. It can be found in the abdomen (75%), mediastinum (20%) and the neck (5%) [2]. It can be primary or metastatic [3]. Fever, limb pain, abdominal mass, abdominal pain, and lower limb weakness are the most common clinical manifestations in the most patients. Diarrheal NB is rare and it is not easy to diagnose in the early stage of the disease [8]. Secretion of large amounts of VIP leads to Intractable diarrhea, hypokalemia, and achlorhydria [11], such as our patient. In 1952, Hawfield and Daisly reported a case of adrenal neuroblastoma with chronic diarrhea that it improved after removal of the tumor [12]. Other investigators also reported ganglioneuroma and ganglion NB with watery diarrhea, hypokalemia, and achlorhydria syndrome in 1973 [13]. VIP secreting tumors include pheochromocytoma, mast cell tumors, non B cell hyperplasia, and medullary thyroid carcinoma. Diarrheal NB was first reported by Ghishan et al [14] in 1979. VIP secreting tumors characterized by watery diarrhea, hypokalemia, and alkalosis (WDHA syndrome). Our patient had not alkalosis. This syndrome is most common in children aged one year to three years, and should be included in the differential diagnosis of pediatric chronic diarrhea. VIP was first isolated in 1970 from pig small intestines [15.] The central nervous system and the sympathetic ganglion neurons are also rich in VIP [8]. VIP is a polypeptide of 28 amino acid residues. It stimulates intestinal water and electrolyte secretion by activating adenylate cyclase and cyclic AMP in intestinal cells [16]. Also, it is distributed in the digestive tract, the central and peripheral nervous systems. [17] VIP also inhibits gastric acid secretion, promotes hepatic glycogenolysis and hyperglycemia and dilates peripheral systemic blood vessels [18]. The signs can be secretory diarrhea, dehydration, and flushing and weight loss. [19] Our case also had these signs. Laboratory abnormalities include hypokalemia, achlorhydria and metabolic acidosis [19, 20]. In resectable stage 1 (confined to the site of origin) and stage 2 (extending beyond the site

of origin but not crossing the midline), surgery alone is usually sufficient without the need for chemotherapy or radiotherapy. In stage 3 (tumor extending beyond the midline) and stage 4 (remote metastases to bones or lymph nodes), chemotherapy and/or radiotherapy are required. Our case was in stage one of tumor and improved after excision of the tumor. We suggest that in a patient with chronic diarrhea without any reason, VIP-secreting tumor should be considered and gastrointestinal, abdominal and thoracal ultrasound or CT scan were done to discover a mass. The serum level of VIP should be measured. Ganglioneuroblastoma and ganglioneuroma are the commonest tumors in children.

## CONCLUSION

Ganglioneuroblastoma may have many atypical presentations, so awareness and keeping it in mind have dramatic importance in early detection and preventing of its serious consequences.

### Conflict of Interest

None declared

### Financial Support

None declared

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