Case Report

Indian Journal of Trauma and Emergency Pediatrics
Volume 8 Number 1, January - April 2016
https://dx.doi.org/10.21088/ijtep.2348.9987.8116.8

Unusual Presentation of Ganglioneuroblastoma

Riyaz Ahmed F.K.*, Sudhir D.**

Abstract

Ganglioneuroblastoma are a mix of malignant neuroblastoma and benign ganglioneuroma tissue. These lesions originate from sympathetic cells. It is a rare tumor with an incidence of 5 per lakh children under 10 years of age. We present a case of unusual presentation of this rare tumor.

Keywords: Neuroblastoma; Ganglioneuroblastoma; Gangalioneuroma; Sympathetic Cells.

Case Report

A 3½-year old girl presented to the orthopedician for bilateral CTEV who referred the case to us, as he found mass per abdomen. On enquiry, she had dribbling of urine and constipation since birth passing hard stools only after laxative. She had no other complaints. Birth history was unremarkable and development was normal and family history being non-contributory.

GPE

Normal vitals, anthropometry around 50th percentile, bilateral CTEV. Per abdomen examination revealed distended bladder with no other mass palpable. CNS, CVS and RS were normal. Ultrasound abdomen revealed distended bladder with bilateral hydroureter and hydronephrosis (Grade-III).

A solid mass of the size 7 x 8 cms arising from left sympathetic chain involving multiple ganglia of lumbar and sacral area was found. It was homogenous echogenicity. CT scan revealed solid mass of 7x7 cms of isodense arising from left sympathetic chain with no calcification or increased vascularity. The mass being free from kidney, adrenal gland and psoas muscle of same side, was displacing the kidney anteriorly and medially. Radiography of all bones including spine was normal with no metastasis. USG guided FNAC showed features of GNB. The girl was then subjected for retroperitoneal excision and histopathology confirmed GNB.

VMA and HVA estimation was not done. The patient was discharged on 10th postoperative day with no complications.

Discussion

Ganglioneuroblastoma are a mix of malignant neuroblastoma and benign ganglioneuroma. They are sometimes called 'transitional tumors'. These lesions also originate from sympathetic cells. Histologically they are considered intermediate because they contain primitive neuroblasts along with mature ganglion cells [1]. Ganglioneuroblastoma occur with equal frequency in boys and girls [1]. Ganglioneuroblastoma most commonly occur in infants and in young children; they almost never occur beyond 10 years (median age – 2 years) [1]. Ganglioneuroblastoma tumors are usually found in the adrenal medulla (35%), retroperitoneum (30-35%), posterior mediastinum (20%), neck (1-5%) and pelvis (2-3%) [1].

Ganglioneuroblastoma have a propensity for secreting catecholamines; approximately 90-95% actively secretes vanillylmandelic acid (VMA) and homovanillic acid (HVA). Catecholamine toxicity rarely results [2]. HVA tends to be secreted by more mature and differentiated tumors, whereas VMA is usually a product of less differentiated tumor [3]. Ganglioneuroblastoma often present with pain

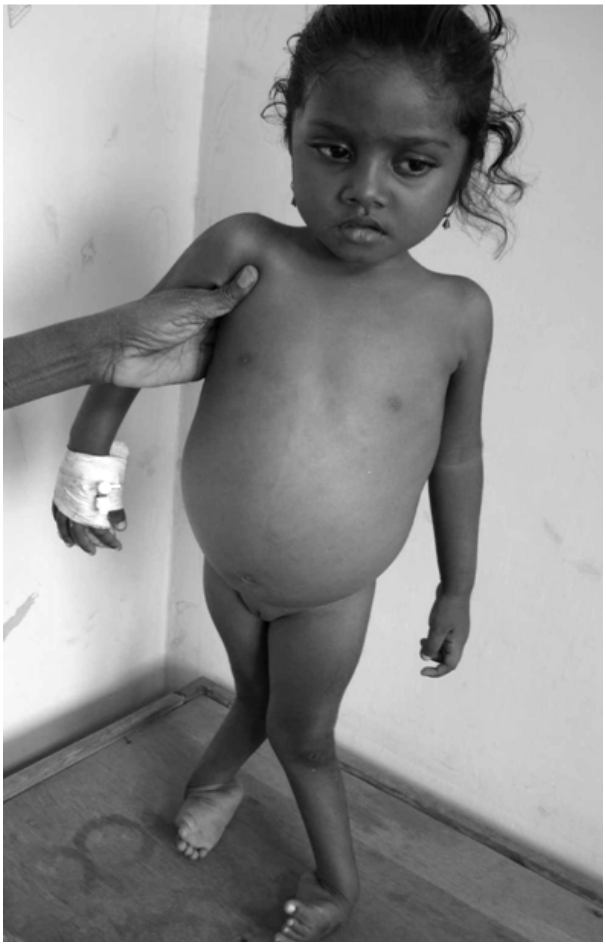
Author Affiliation: *Assistant Professor ****Associate Professor, Department of Paediatrics, VIMS, Bellary, Karnataka.

Reprint Request: Riyaz Ahmed F.K., Assistant Professor, Department of Paediatrics, Vijayanagara Institute of Medical Sciences (VIMS), Ballari, Karnataka 583104.
E-mail: ramspsm@gmail.com

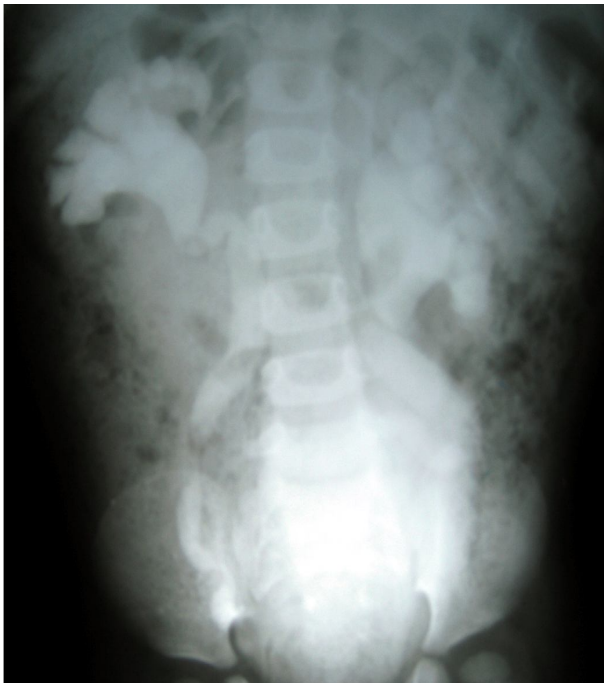
caused by either the primary tumor or by metastatic mass and abdominal distension. Patient may also complain of irritability, weight loss, malaise, shortness of breath, peripheral neurologic symptoms (nerve or nerve root compression) [2]. GNB can metastasize with bone being most common site [3]. GNB is screened by VMA and HVA levels in urine [4]. CT scan is the most commonly used modality to diagnose the GNB [2]. Despite these possible comorbidities, the prognosis for patients with GNB is relatively good. The tumor may spontaneously regress or mature to ganglioneuroma. Regression occur in 1-2% of tumors [3].

Most probable explanation of chronic symptomatology in this patient would be as this patient has GNB involving multiple lumbar and sacral sympathetic ganglia, probably congenital which have completely interrupted the parasympathetic supply to bladder and bowel thereby resulting in autonomic bladder and chronic constipation.

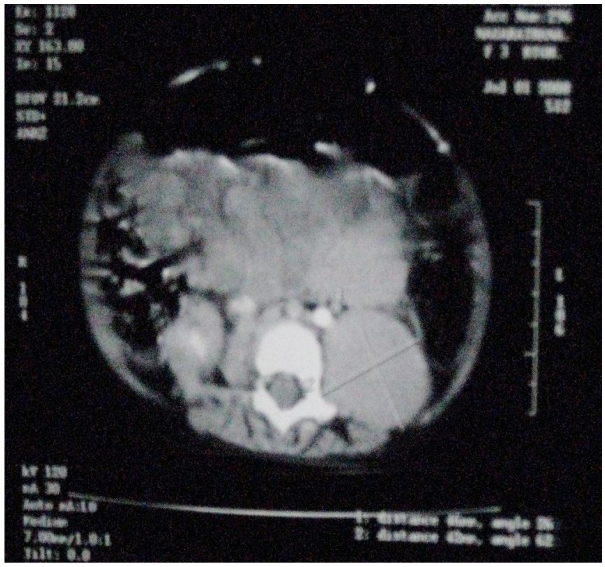
The presence of CTEV in this patient could not be explained. The only association of CTEV with GNB is seen in fetal hydantoin syndrome but here mother was not alcoholic. Hence, association may be coincidental.



Photograph showing grossly distended abdomen with bilateral CTEV



IVP showing bilateral hydronephrosis and hydroureter (grade-III)



CT abdomen showing left sided mass measuring 7x9 cm arising from left lumbar and sacral sympathetic ganglia free from kidney, adrenal gland and psoas muscle

Abbreviations

GNB-Ganglioneuroblastoma

VMA-Vanilyl Mandellic acid

HVA-Homovanillic acid

CT-Computed tomography

CTEV-Congenital Talipes equino varus

References

1. Rha SE, Byun JY, Jung SE, et al. Neurogenic tumors in the abdomen: tumor types and imaging characteristics. *Radiographics*. Jan-Feb 2003; 23(1): 29-43.
2. Lonergan GJ, Schwab CM, Suarez ES, Carlson CL. Neuroblastoma, ganglioneuroblastoma, and ganglioneuroma: radiologic-pathologic correlation. *Radiographics*. Jul-Aug 2002; 22(4): 911-34.
3. http://www.emedicine.com/radio/TOPIC293.HTM (eMedicine Ganglio neuroma and Ganglioneur oblastoma: Article by Andrzej R.Jedynak. March 19, 2008).
4. Brossard J, Bernstein ML, Lemieux B. Neuroblastoma: an enigmatic disease. *British Medical Bulletin*. 1996; 52: 787-801.

Revised Rates for 2016 (Institutional)

Title	Frequency	Rate (Rs): India	Rate ($):ROW
Dermatology International	2	5000	500
Gastroenterology International	2	5500	550
Indian Journal of Agriculture Business	2	5000	500
Indian Journal of Anatomy	3	8000	800
Indian Journal of Ancient Medicine and Yoga	4	7500	750
Indian Journal of Anesthesia and Analgesia	3	7000	700
Indian Journal of Anthropology	2	12000	1200
Indian Journal of Biology	2	4000	400
Indian Journal of Cancer Education and Research	2	8500	850
Indian Journal of Communicable Diseases	2	8000	800
Indian Journal of Dental Education	4	4500	450
Indian Journal of Forensic Medicine and Pathology	4	15500	1550
Indian Journal of Forensic Odontology	2	4500	450
Indian Journal of Genetics and Molecular Research	2	6500	650
Indian Journal of Law and Human Behavior	2	5500	550
Indian Journal of Library and Information Science	3	9000	900
Indian Journal of Maternal-Fetal & Neonatal Medicine	2	9000	900
Indian Journal of Medical & Health Sciences	2	6500	650
Indian Journal of Obstetrics and Gynecology	3	7000	700
Indian Journal of Pathology: Research and Practice	3	11500	1150
Indian Journal of Plant and Soil	2	5500	550
Indian Journal of Preventive Medicine	2	6500	650
International Journal of Food, Nutrition & Dietetics	3	5000	500
International Journal of History	2	6500	650
International Journal of Neurology and Neurosurgery	2	10000	1000
International Journal of Political Science	2	5500	550
International Journal of Practical Nursing	3	5000	500
International Physiology	2	7000	700
Journal of Animal Feed Science and Technology	2	4100	410
Journal of Cardiovascular Medicine and Surgery	2	9100	910
Journal of Forensic Chemistry and Toxicology	2	9000	900
Journal of Microbiology and Related Research	2	8000	800
Journal of Orthopaedic Education	2	5000	500
Journal of Pharmaceutical and Medicinal Chemistry	2	16000	1600
Journal of Practical Biochemistry and Biophysics	2	5500	550
Journal of Social Welfare and Management	3	7500	750
New Indian Journal of Surgery	3	7100	710
Ophthalmology and Allied Sciences	2	5500	550
Otolaryngology International	2	5000	500
Pediatric Education and Research	3	7000	700
Physiotherapy and Occupational Therapy Journal	4	8500	850
Urology, Nephrology and Andrology International	2	7000	700
SUPER SPECIALITY JOURNALS			
Indian Journal of Emergency Medicine	2	12000	1200
Indian Journal of Surgical Nursing	3	5000	500
Indian Journal of Trauma & Emergency Pediatrics	3	9000	900
International Journal of Pediatric Nursing	3	5000	500
Journal of Community and Public Health Nurisng	2	5000	500
Journal of Geriatric Nursing	2	5000	500
Journal of Medical Images and Case Reports	2	5000	500
Journal of Nurse Midwifery and Maternal Health	3	5000	500
Journal of Organ Transplantation	2	25900	2590
Journal of Psychiatric Nursing	3	5000	500
Psychiatry and Mental Health	2	7500	750
OPEN ACCESS JOURNALS			
Global Research in Engineering		5000	500
Global Research in Food and Nutrition		5000	500
Global Research in Library and Information Science		5000	500
Global Research in Medical Sciences		5000	500
Global Research in Space Science		5000	500

Terms of Supply:

1. Advance payment required by Demand Draft payable to Red Flower Publicaion Pvt. Ltd. payable at Delhi.
2. Cancellation not allowed except for duplicate payment.
3. Agents allowed 10% discount.
4. Claim must be made within six months from issue date.

Order from

Red Flower Publication Pvt. Ltd., 48 / 41-42, DSIDC, Pocket-II, Mayur Vihar Phase-I, Delhi - 110 091 (India), Tel: 91-11-22754205, 45796900, Fax: 91-11-22754205. E-mail: redflowerppl@vsnl.net, redflowerppl@gmail.com, Website: www.rfppl.co.in