

## A 4 Month Male Child With Sacrococcygeal neuroblastoma: A Case Report.

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**Abstract:** Neuroblastoma is the most common extracranial solid tumor of childhood. Sacrococcygeal neuroblastomas are reported to be rare. We report a case of a poorly-differentiated neuroblastoma case, detected in a 4-month-old male who presented with retention of urine and totally resected by surgery. Biopsy revealed features consistent with neuroblastoma. MRI pelvis showed large well defined mass in presacral region displacing rectum and bladder anteriorly with indistinct fat planes.

**Keywords:** Neuroblastoma, Sacrococcygeal neuroblastomas, Tumor In Infants

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Date of Submission: 26-10-2017

Date of acceptance: 23-12-2017

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### I. Introduction

Neuroblastoma (NBL) is a tumor derived from the primitive neural crest cells and seen at the adrenal medulla and sympathetic ganglia. It is the most common extracranial solid tumor. The most common site of origin during infancy is the abdomen, especially the suprarenal glands (Brodeur, 2006). Pelvic neuroblastomas are rather rare. Herein, we are presenting a 4-month-old male NBL case who presented with complaints of urinary retention.

### II. Case Report

A 4-month-old male came to surgery OPD with complaints of urinary retention since 8 days for which he was catheterised. Physical examination revealed a vague soft tissue mass in the pelvic region. The skin covering the lesion had blue-purple color change. Findings of a neurologic examination and the other systems examinations were normal. In addition, laboratory studies revealed no hematologic abnormalities. Levels of tumor markers such as carcinoembryonic antigen and alphafetoprotein were normal. Lumbosacral vertebral column MRI revealed a large well defined mass lesion measuring 4.3x4.8x6.5 cm in presacral region. The lesion is displacing rectum and bladder anteriorly with indistinct fat planes with overlying lumbo-sacral spine to be normal with no signs of destruction. There is mild left sided dilated pelvicalyceal system likely due to compression of left ureter by mass lesion with no extra pelvic extension of mass. (Fig. 1a-c). The preoperative diagnosis of the patient was teratoma. After being separated from the sacrococcygeal joint level, the lesion was totally excised all together with coccyx and the intrapelvic part. There appeared vaguely circumscribed and partly capsulated tumor arranged in nests, lobules and festoons on microscopic examination and the tumor cells were mostly small blue cells having round to oval nuclei with speckled chromatin (salt and pepper) and inconspicuous nuclei. So the tumor was interpreted as poorly-differentiated neuroblastoma. Following the pathology report, the vanilmandelic acid (VMA) and homovanilic acid (HVA) levels were evaluated and found to be normal. Being younger than 1-years of age, the tumor having poorly-differentiated type, tumor having no n-myc amplification and being removed totally were low risk prognostic factors in our pelvic neuroblastoma case.



**Post operative:-** Excised tissue

### **III. Discussion**

Neuroblastoma accounts for approximately 7.5% of all childhood malignancies (Crucetti et al., 2000). Prevalance is 1 in every 7000 live birth. Neuroblastoma is commonly seen at 0-4 years and the mean age of diagnosis is 22 months. 36- 40% of cases are younger than 1 year, whereas 89% is younger than five years and 98% is younger than ten years of age (Shusterman and George, 2006). The tumor is seen mostly in the first month of the life. Neuroblastomas are tumors of the sympathetic chain or any sympathetic ganglia. Most primary tumors occur in the abdomen (65%), and half of the tumors occur in the adrenal gland. Other common places are thorax, neck and pelvis. Pelvic sites of origin accounts for 2-6% of all solid NBs and the primary site of origin is unknown in 1% (Knoedler, 1989; Crucetti, 2000; Brodeur, 2006 ). Neuroblastomas of the pelvis are calculated to account for 0.25% of all neonatal neoplasms (Crucetti, 2000; Massad, 1986). A few cases were reported in the literature. D'Alessio et al. (2006) reported a case of pelvic (presacral) cystic NBL in a 2-month-old boy. Tanaka et al. (2005) reported a case of neonatal neuroblastoma mimicking Altman type III sacrococcygealteratoma. Watanabe et al. (2008) described a case of a 2-month-old girl with large presacral NB. And also Unal et al. (2010) reported a case of malignant transformation of an unresected sacrococcygealteratoma to neuroblastoma in a 6-years-old girl. The symptoms of neuroblastoma arising from the sacral region may include abdominal distention and urinary retention. Our patient had complaint of urinary retention for the past 8 days. The differential diagnosis should be defined clearly preoperatively especially for sacral cystic neuroblastomas. Teratomas, cystic lymphangiomas, meningomyeloceles and chordomas may arise from this region in infants. But today computed tomography and magnetic resonance imaging helps in diagnosis (Tanaka et al., 2005; D'Alessio et al., 2006; Watanabe et al., 2008). Urine VMA and HVA levels are reported to be normal or slightly higher than normal in the literature diagnosis (Tanaka et al., 2005; D'Alessio et al., 2006; Watanabe et al., 2008). In our case, following the pathology report, the VMA and HVA levels were evaluated and found to be normal. Preoperative studies were not performed because the preoperative diagnosis of the patient was teratoma.

The mortality rate of pelvic neuroblastomas is quite low even if the tumor is incompletely resected. Unfortunately morbidity related to the tumor bulk pressure and probable postoperative neurologic deficits are rather high (35%) as well (Knoedler et al., 1989). We suggest that neuroblastoma should be considered in the differential diagnosis of infantile sacrococcygeal tumors even if they are asymptomatic and urinary levels of VMA and HVA should be examined.

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\*Dr. Sanjay Pandey. "A 4 Month Male Child With Sacrococcygealneuroblastoma: A Case Report."  
IOSR Journal of Dental and Medical Sciences (IOSR-JDMS) 16.12 (2017): 05-07