Case Report

Cystic neuroblastoma in an infant: A case report

Jasmine Bajracharya[1], Sagar Upadhyay[2], Prabhat Poudel[2], Binay Khadka[2]

Abstract

Neuroblastoma is a neoplasm of the peripheral sympathetic nervous system. Depending upon the primary location of the tumor, it can present as incidental finding, hypertension, abdominal pain, and distention with nonspecific clinical signs. Our patient was four-months-old when he developed excessive crying, increased frequency of micturition, and loose bowel movements. On CT scan, a cystic mass was found in the left suprarenal region and was surgically removed. Immunohistochemistry of the mass confirmed the diagnosis of cystic neuroblastoma. The outcome was good after surgical excision and chemotherapy. Early diagnosis and management of cystic neuroblastoma will result in a good outcome.

Keywords: Abdominal mass; Cystic; Infant; Neuroblastoma

Author affiliations:

[1]Department of Pediatric Surgery, Nepal Medical College Teaching Hospital, Kathmandu, Nepal

[2]Department of General Surgery, Nepal Medical College Teaching Hospital, Kathmandu, Nepal

Correspondence:

Dr. Prabhat Poudel,
Nepal Medical College Teaching Hospital,
Kathmandu, Nepal

Email: theprabhatpoudel@gmail.com

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Introduction

Neuroblastoma is a pediatric neoplasm that emerges anywhere in the peripheral sympathetic nervous system. More than 95% of cases are detected by 10 years of age.[1] They occur once in every 7500 to 10000 live births and comprise 10% of all childhood tumors.[2] They have both solid and cystic forms; cystic neuroblastoma is extremely rare with only a few cases described in the medical literature.[3] Hereby, we report a case of cystic neuroblastoma in a four-month-old male child.

Case Report

A four-month-old male child was brought to the pediatrics outpatient department (OPD) with a complaint of excessive crying at night with increased frequency of micturition and multiple episodes of loose motion. Birth and family history was insignificant. On examination, the child was playful without pallor, icterus, cyanosis, edema, or dehydration. The abdomen was soft and non-tender with mild distension without palpable mass.

Ultrasound of his abdomen and pelvis revealed a well-defined hypoechoic lesion measuring 4.2x3.2x3 cm in the left suprarenal region without any evidence of calcification and cystic area, causing mass effects in the upper pole of the left kidney.

Vanillylmandelic acid (VMA) in 24-hour urine was negative. We performed a contrast-enhanced computed tomography (CECT) of his abdomen that revealed a suprarenal mass (**Figure 1**) measuring 4.6x3.7x4.6 cm, with a displaced left kidney, without hydronephrosis. The mass

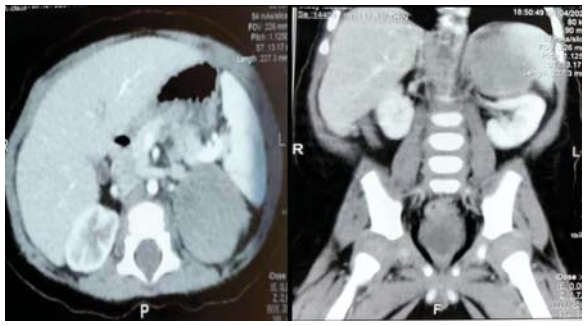

Figure 1. CECT of the abdomen revealing a suprarenal mass with displaced kidney without hydronephrosis

appeared heterogeneous with some areas of calcifications, insinuating itself beneath the aorta, abutting the left psoas muscle, and compressing the left kidney. There was no evidence of invasion of surrounding organs.

The patient underwent an excision of the tumor in the left suprarenal gland revealing a cystic mass (**Figure 2**), and the specimen was sent for Histopathological examination (HPE). The patient had an uneventful postoperative period and was discharged on the fourth postoperative day.

In HPE, an encapsulated and well-circumscribed adrenal medulla tumor consisting of an anastomotic network of vessels with a flattened endothelium and intraluminal collection of round lymphocyte-like nuclei and scant cytoplasm with foci of rosette formation was observed. Immunohistochemistry (IHC) was advised to confirm the diagnosis. The IHC reports came back with negative CD34, GFAP, ALK-1, desmin, and CD45. And a final diagnosis of cystic neuroblastoma was made.

The patient received two cycles of Cyclophosphamide. N-MYC amplification test which was negative. A CECT of the abdomen and pelvis done 6 months after surgery revealed enhancing lymph nodes in the left paraaortic region, stenotic left pelvic ureteric junction, and moderate left hydronephrosis. Computed Tomography guided Intravenous urography (CT-IVU) revealed a few left paraaortic nodes with left PUJ stenosis and moderate to gross left hydronephrosis with obstructive uropathy. We also performed a TC-MDP whole-body scan, it was normal without any evidence of skeletal metastasis.

Figure 2. Cystic mass excised from the left suprarenal gland (scale has been added later for the size reference)

Technetium diethylenetriaminepentaacetic acid (TC-DTPA) diuretic renogram revealed a poor functioning left kidney with 10% differential function and a normal functioning right kidney with 90% differential function. Upon urologic evaluation, the left VUJ was not accommodating a 4.5Fr Ureterorenoscope (URS). Complete occlusion at the left proximal ureter was seen in retrograde pyelogram (RPG).

The patient was planned for a percutaneous nephrostomy (PCN) and a repeat DTPA scan was done before PCN which revealed a five percent differential function of the left kidney. He underwent PCN and was scheduled for a left nephrectomy but intraoperatively the kidney appeared to be salvageable, so Anderson-Hynes open pyeloplasty with double-J(DJ) stenting was performed. A DTPA scan done 3 months after the pyeloplasty showed that the differential function of the left kidney improved to 15% with DJ stent in situ.

The patient is doing well on follow-up visits, the most recent visit was 7 months following the surgery.

Discussion

Neuroblastoma is the most frequently occurring extracranial childhood tumor and all suprarenal masses in the infantile period are often presumed to be neuroblastomas. Although solid neuroblastomas are relatively common, cystic neuroblastoma is extremely rare, with very few cases published in the literature.[3] Neuroblastomas could be hereditary or could arise due to random mutations. The median age at the time of diagnosis of neuroblastoma is 18 months, with 40% of patients diagnosed in infancy and 90% of patients below 10 years of age.[4] Our patient, however, was only four months old at the diagnosis.

The clinical signs and symptoms of neuroblastoma are directly related to the location of the primary tumor and the sites of metastatic disease. More than half of the primary neuroblastomas occur in the medulla of the adrenal glands.[5] Localized disease often presents as an incidental finding but large abdominal tumors can cause hypertension due to compression of renal arteries, abdominal distension, and pain. Tumor metastases present with organ-specific symptoms. Non-specific clinical symptoms include fever, weight loss, and fatigue. Chronic diarrhea caused by the secretion of vasoactive intestinal peptides can often be the first clinical manifestation.

The diagnosis of solid neuroblastoma could be made with the help of clinical, histopathological, and radiological findings.[6] Cystic neuroblastomas can often be identified in prenatal ultrasound but should be differentiated from adrenal hemorrhage, mesenchymal hamartoma, and subdiaphragmatic extrapulmonary sequestration.[7] Diagnostic evaluation is based not only on a thorough history

and physical history, but also on biochemical, histological, and radiographic analyses. Histological confirmation is necessary to establish a diagnosis of neuroblastoma.

Treatment modalities include simple observation, surgical resection, chemotherapy, radiation therapy, stem cell transplantation, and immunotherapy, depending on tumor type, location, grade, and stage at the time of diagnosis. Our patient had a tumor excision and two cycles of cyclophosphamide after confirmation of the diagnosis. The prognosis is very good in localized cystic neuroblastoma; it has the highest rate of spontaneous regression of all human malignant neoplasms, yet one of the poorest outcomes when occurring as a spreading disease in children.[8] Hydronephrosis occurs in 20% of pediatric non-renal abdominal and pelvic tumors. 60% of these cases resolve with the treatment of the primary tumor and 13% require specific urological intervention for urinary tract involvement or compression, the main predictive factor being the primary stage of the disease.[9]

Conclusion

Cystic neuroblastoma is a very uncommon diagnosis. Presentation in a very young infant could be nonspecific, but an early diagnosis and timely management can result in excellent outcomes. Hydronephrosis is a known complication of intra-abdominal neoplasms that can resolve spontaneously with the removal of the tumor.

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