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CASE REPORT

PRIMARY RENAL NEUROBLASTOMA: A CLINICOPATHOLOGICAL CASE REPORT

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ABSTRACT:

Neuroblastoma is the second most common solid pediatric malignancy, with >90% of tumors diagnosed in children <10 years old. The Primary Intra Renal Neuroblastoma (IRNB) is a rare condition. Intra renal Neuroblastoma typically results from direct renal invasion from an adrenal Neuroblastoma, but true intra renal Neuroblastoma originates either sequestered adrenal rests during the fetal life or intra renal sympathetic ganglia. The distinction of this rare tumor from Wilms' tumor is an important challenge since both tumors have major differences in prognostic and therapeutic response. This case highlights a primary renal neuroblastoma in a 18 months old male child presenting with unilateral abdominal mass. The diagnosis of primary renal neuroblastoma was confirmed by clinical findings, radiology and histopathology followed by immunohistochemistry.

KEY WORDS: Neuroblastoma, pediatric, histopathology, radiology

INTRODUCTION

Primary renal neuroblastoma is a rare but aggressive tumor and comprises of 10% of all childhood malignancies.(1) Renal invasion by neuroblastoma occurs by direct penetration through the renal capsule and/or lymphatic perivascular spread. (2)Renal invasion occurs in approximately 20.4% of cases of abdominal neuroblastoma. A higher incidence of hypertension (66–100%) has been associated with intra renal neuroblastoma as compared to 27% reported in the literature for neuroblastoma, probably because of compression of renal vessels, increased rennin release from the kidney, and a high circulating level of catecholamine.(5,6) It can mimic Wilms' tumor in its clinical presentation. The distinction of this rare tumor from Wilms' tumor is an important challenge since both tumors have

major differences in prognostic and therapeutic response. (8) The factors that affect survival are age and health of child, extent of the disease, size, type and location of the tumor, metastasis, tumor response to therapy, and overall child's tolerance to medications.(9,10) This case highlights a primary renal neuroblastoma in a 18 months old male child presenting with unilateral abdominal mass. The diagnosis of primary renal neuroblastoma was confirmed by clinical findings, radiology and histopathology followed by immunohistochemistry.

CASE REPORT

An 18 months old baby presented to the pediatric OPD in a highly irritable state with complains of vomiting, abdominal swelling and loss of feed uptake. On examination showed a right sided abdominal mass extending slightly below the costal margin and measuring approximately 4x4 cms. Baby's complete haemogram showed Hb- 8.7 gm%, TC- WNL, PLT- WNL. Peripheral smear showed normocytic hypo chromic anemia. The liver and renal function tests were all within normal limits. A computed tomography was done which showed a right sided intra renal mass involving part of the kidney measuring 5.6x5.8 cms. Tumor did not extend beyond the midline. Capsular invasion was also not seen. (Fig 1)Patient had a right sided nephrectomy and specimen sent for histopathological examination. Gross showed an intra renal growth with peripheral part of normal kidney and areas of hemorrhage. Hilar structures were not identifiable. Capsule was intact. Adrenal gland was not received with the specimen. (Fig 2) Histopathology showed small round cells with hyper chromatic nucleus and scant cytoplasm arranged in sheets with scanty stroma. Infiltration of lymphocytes was seen. Perivascular arrangement of cells was seen. Thrombotic blood vessels and areas of hemorrhage seen. (Fig 3) Immunohistochemistry was done for Chromogranin and Synaptophysin which showed positive membrane staining consecutively. Our final diagnosis was Primary intra renal neuroblastoma. (Fig 4, 5)

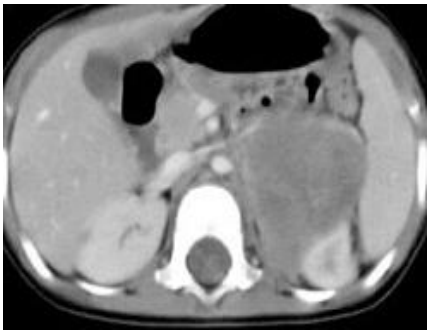

Fig 1: CT finding

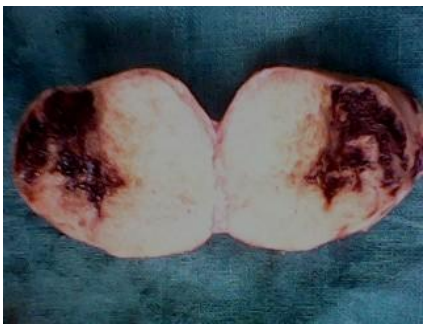

Fig 2: Gross of resected right kidney.

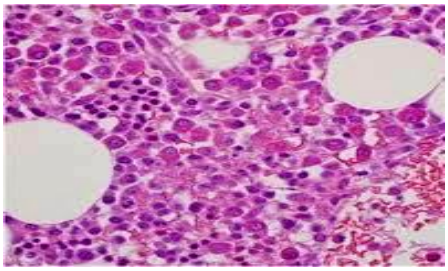

Fig 3: Histopathology (10X)

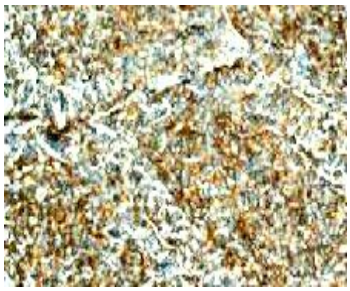

Fig 4: IHC , Chromogranin (10X)

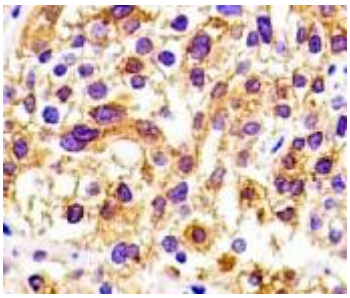

Fig 5: IHC, Synaptophysin (40X)

DISCUSSION

Neuroblastoma occurs in the sympathetic nervous system and principally affects the adrenal glands and retro peritoneum, with a similar pattern of distribution in children and adults. The presentation of the neuroblastoma can be asymptomatic and varies depending on the tumor size and site, involvement of blood vessels.(9) The treatment modalities for children have included surgery, chemotherapy, and radiotherapy. Patients can be stratified into favorable and unfavorable risk groups according to the histopathology characteristics. (10) A high proportion of intra renal neuroblastoma is of unfavourable histology as defined by the International Neuroblastoma Pathology Classification and have a higher incidence of anaplasia (32%) when compared to both their adrenal counterparts and to Wilms' tumor.(7) In the present case histopathology favored poorly differentiated variant without proper rosettes or stroma. The factors known to have a positive prognosis in children include age <1 year, a low disease stage, a lack of MYC amplification, and hyper diploidy. (10, 11) Complete surgical resection is the most effective therapy for localized disease. In our case patient recovery was uneventful post surgery and post chemotherapy. (10) A higher incidence of hypertension (66–100%) has been associated with intra renal neuroblastoma as compared to 27% reported in the literature for neuroblastoma, probably because of compression of renal vessels, increased rennin release from the kidney, and a high circulating level of catecholamine. Lall et al. reported hypertension in all their cases of intra renal neuroblastoma. (11) In our case patient was not hypertensive on presentation. There are some reports about association of

neuroblastoma and disseminated intravascular coagulation (DIC). (12)This patient did not have any such complication. The distinction of intra-renal neuroblastoma from Wilms' tumor is important as both tumors have different prognostic and therapeutic responses. (8) Clinical and histopathological findings along with Immunohistochemistry helped us to rule out Wilms' tumor. Urine catecholamine levels would become very useful to distinguish intra-renal neuroblastoma from Wilms' tumor. (13) In our case the patient was not tested for urine catecholamine.

CONCLUSION:

Primary intra renal neuroblastoma is an extremely infrequent tumor in kidney. Clinical, radiological, and pathological correlation is very essential for diagnosis and appropriate management of this type of unusual cases. The present case emphasizes the need for a clear histopathology diagnosis to establish the appropriate investigations, treatment options, and routine follow-up protocol.

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