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Extrarenal Nephroblastoma

Gaxa Luvo, Hlatshwayo Bafana

ABSTRACT

Introduction: As much as the nephroblastoma is almost exclusively known to be a renal tumor, some of the extrarenal locations, although rare, have been described in the literature. The sites of this rare entity (extrarenal nephroblastoma) reported entail the inguinal canals, retro-peritoneum, mediastinum, chest wall, ovaries, cervix, uterus and the prostate gland. Case Report: We report a case of a 13-month-old boy who was referred from the peripheral hospital presenting with severe abdominal distention of three months duration. The patient was critical ill on arrival and had to be actively resuscitated. The patient was emaciated. No known past medical history reported and the patient had normal developmental milestones. On physical examination a large abdominopelvic mass was palpated. The patient also had sepsis confirmed with blood culture and hypoglycemia with a blood glucose measurement of 1.4 mmol/l saturation was 96% in room air and had episodes of gasping respiration, the heart beat was ranging between 110–140 bpm. The patient had pallor with hemoglobin 8.4 g/dL and had cold peripheries. Computed tomography scan and biopsy were performed and the diagnosis of an extrarenal large abdominopelvic nephroblastoma was made. The patient was later, after stabilization, treated with chemotherapy. The patient now remains in a critical general condition while on chemotherapy. Conclusion: A nephroblastoma may present in some extrarenal sites which is quite an uncommon encounter, the most common of these rare sites being the retro-peritoneum. The final diagnosis is mainly histopathological. There are no specific management protocols and as a result the extrarenal tumors are managed in the same manner in which the intra-renal tumors are managed with a general good prognosis.

Keywords: Computed tomography scan, Extrarenal nephroblastoma, Histopathology

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INTRODUCTION

Nephroblastoma also known as the Wilms' tumor arises almost exclusively from the kidney and it is one of the most common malignant solid pediatric neoplasms and the tumor represents about 6% of all childhood carcinomas [1, 2].

Simultaneous involvement of both kidneys by nephroblastoma occurs in 5% of cases and diagnosing the tumor as extrarenal is extremely rare and exceptional with cases ranging between 0.5–1% reported [2].

There are no specific clinical criteria used to diagnose the extrarenal nephroblastoma because the clinical presentation is dictated by the location of the tumor [1]. A radiological examination plays a critical role in the assessment of these lesions for the extent but the ultimate diagnosis is histopathological. The extrarenal nephroblastoma responds very well to the same chemotherapy that is used for the intrarenal nephroblastoma.

CASE REPORT

A 13-month-old male baby presented with severe abdominal distention of three months duration. The birth history showed that the patient was born through a normal vaginal delivery with an uneventful birth history. The birth weight was 2.65 kg and 44.3 cm in length. The Apgar score was 10/10. The patient had normal developmental milestones. On physical examination the patient was very wasted. The patient also had sepsis and hypoglycemia with a blood glucose measurement of 1.4 mmol/l. The saturation was 96% in room air and had episodes of gasping respiration, the heart beat was ranging between 110–140 bpm. The patient had pallor with hemoglobin 8.4 g/dL and had cold peripheries. The cardiovascular system examination showed an S1S2 murmur. Central nervous system examination revealed that the patient was lethargic and not opening the eyes. The patient had to be resuscitated on arrival by giving a bolus of 50% dextrose ordered and given stat over 10 minutes. Emergency blood transfusion was also given on 0.9% saline bolus.

Later the patient became active, moving all the limbs, awake, looking around and started breathing normally. Four hourly glucose monitoring was then ordered while the patient was wheeled to the ward. Post blood transfusion and dextrose vital signs were as follows:

Blood pressure 108/57, pulse 109, saturation 100%, and blood glucose 3.5 mmol/l. Hemoglobin post blood transfusion 10.9 g/dl and the platelets were still low $126x10^9$/L. Urea and creatinine results were within normal limits. The abnormal results in the liver function test were those of lactate dehydrogenase measuring 1899 U/L and the normal higher range 430 U/L.

The inflammatory marker severely elevated (C-reactive protein 58 mg/l) the highest normal range is less than 10 mg/l. The weight of the patient at presentation was 8 kg.

Computed tomography scan showed a large abdominopelvic mass clearly separable from the kidneys and heterogeneously enhancing. Biopsy was also performed and the diagnosis of an extrarenal nephroblastoma was made.

The histopathology report of this case was as follows:

Microscopy section showed a representative tissue core of a tumor composed of small round dark cells compatible with blastoma

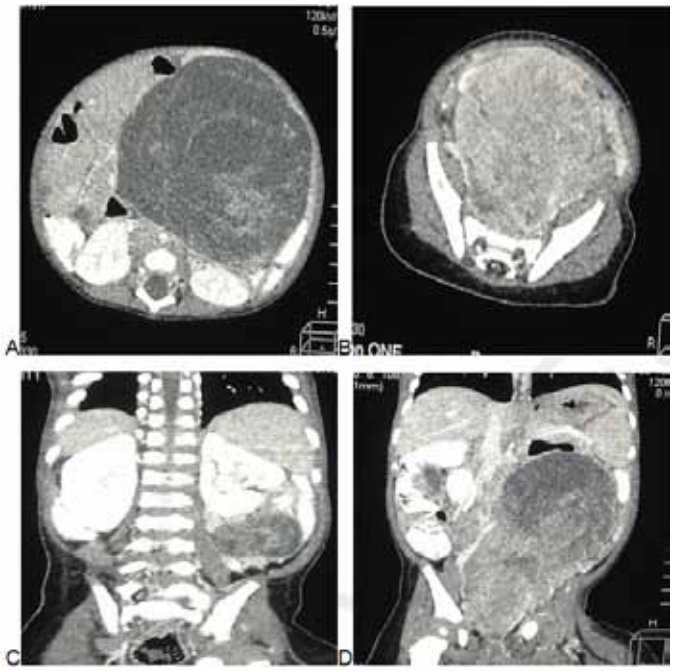

Figure 1: (A) Axial view at the renal level showing a large heterogeneously enhancing soft tissue mass that is clearly separable from the left kidney and displacing it superiorly. The mass displaces the bowel superolaterally and crosses the midline to the contra-lateral side, (B) Axial view at the pelvic level showing the mass at the pelvis displacing the pelvic structures, (C) Coronal view also showing a clear separation of the mass from the left kidney, and (D) Coronal view showing the extent of the mass from the pelvic region to the left infra-renal space.

The above described features are those of a nephroblastoma.

When stable the patient was started on chemotherapy and the chemotherapy drugs that were given were as follows:

Vincristine 0.7 mg intravenously as a stat dose and actinomycin 0.3 mg intravenously as a stat dose and the patient was also covered with rocephin 400 mg intravenously two times a day.

The patient is now in a continuous course of chemotherapy, however, his general condition remains unsatisfactory and appears to be deteriorating.

DISCUSSION

Nephroblastoma also known as the Wilms' tumor arises almost exclusively from the kidney and it is one of the most common malignant solid pediatric neoplasms and the tumor represents about 6% of all childhood carcinomas [1, 2].

Simultaneous involvement of both kidneys by nephroblastoma occurs in 5% of cases and diagnosing the tumor as extrarenal is extremely rare and exceptional with cases ranging between 0.5–1% reported [2].

Some of the sites of this rare entity (extrarenal nephroblastoma) reported entail the inguinal canals,

retroperitoneum, mediastinum, chest wall, ovaries, cervix, uterus and the prostate gland [3].

In literature up to date only 200 cases of the extrarenal nephroblastoma including both the children and adults have been reported and there are only 25 cases in 52 years that have been reported in the Japanese literature between 1955 and 2007 [4].

The exact origin of the extrarenal nephroblastoma is not well understood although various hypothesis and theories implicate metanephric derivatives, intermediate blasterma and mesonephric rests as the culprits [5].

It is well known as a rule that to diagnose extrarenal nephroblastoma there must be a complete absence of a primary renal lesion [6].

Extrarenal nephroblastoma does not have a specific clinical presentation as the tumor site largely dictates the clinical signs and symptoms of this condition [7].

The consensus regarding the criteria for the pathologic diagnosis of the extrarenal nephroblastoma is accepted to involve the following [8] :

1. Extrarenal site of the primary carcinoma
2. Primitive blastematous spindle or round cell component
3. Abortive or embryonal tubular or glomeruloid structures and
4. No evidence of teratoma or renal cell carcinoma

It is not clear whether the prognostic features that apply to the intra-renal nephroblastoma also apply to the extrarenal Wilms' tumor [9].

As much as there is no specific management protocol for extrarenal Wilms' tumor it is generally known that all surgically treated patients need postoperative chemotherapy and the drugs used for renal nephroblastoma are equally effective in treating the cases of extrarenal nephroblastoma [9].

Patients with un-resectable gross residual tumor and those with distant metastasis are treated with radiotherapy [9].

Histopathology is the ultimate investigative tool to clinch the diagnosis of extrarenal nephroblastoma.

CONCLUSION

Extrarenal nephroblastoma is an extremely rare and an exceptional condition that needs a multi-disciplinary approach. The ultimate diagnosis is achieved histopathologically. Staging and management of the extrarenal nephroblastoma does not differ from that of intra-renal nephroblastoma.

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Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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