Case Report

Diagnostic Dilemma in Left Flank Mass in an Infant: A Rare Case of Intrarenal Neuroblastoma

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Abstract

Neuroblastoma (NBL) and nephroblastoma present with abdominal mass and various imaging features help in differentiating the two; however, localisation is difficult in large masses and at times imaging features can be confusing. Here, we describe the case of large left-sided NBL arising from the adrenal and involving the left kidney with moderate hydronephrosis.

Keywords: Hydronephrosis, mass, nephroblastoma, neuroblastoma

Introduction

Intrarenal neuroblastoma (NBL) is very rare and often misdiagnosed as Wilm's tumour[1] on imaging which poses difficulty in the management. NBL is an aggressive malignancy in contrast to Wilm's tumour which behaves in an indolent manner. It is imperative to differentiate between the two as the prognosis and the management differ.

Case Report

A 5-month-old female presented in the outpatient department with an abdominal lump. The swelling was insidious in onset and gradually increased to the present size. There was associated fever and vomiting on and off. No history of hematuria or trauma was present. Abdominal examination revealed a lump in the left flank which was extending from the subcostal region to the pelvis and was not moving with respiration. Margins of the mass were well-defined superiorly and inferiorly. Complete blood counts were normal except for anaemia (Hb-7.7 g/dL). For localisation of the mass, an ultrasound abdomen was done which revealed a large hetero-echoic mass in the left lumbar region and was seen in relation to the upper pole of the left kidney with hydronephrosis. There was no obvious calcification. On ultrasound, the possibility of Wilm's tumour or NBL was kept because of non-visualisation of the left adrenal gland and upper pole of the left kidney and hydronephrosis. Contrast-enhanced computerized tomography (CECT) of the chest and abdomen were done for further characterisation, to look for the origin, resectability, as well as metastasis. CECT revealed a large heterogeneously enhancing mass arising from the upper pole of the left kidney with extension into the renal pelvis causing moderate hydronephrosis. The mass was crossing midline anterior to the aorta and was causing displacement of vessels to the left side and there was no evidence of intraspinal extension. The left adrenal gland was not visualised separately. No evidence of calcification was seen within the mass. There were multiple subpleural nodules in bilateral lung parenchyma which were suggestive of metastasis [Figure 1]. No evidence of liver or skeletal metastasis was seen. Most of the imaging features were favouring Wilms tumour (renal involvement with hydronephrosis, no vascular encasement, no intraspinal extension, no calcification and lung metastasis) more than NBL. However, age (4 months) was the only parameter which was not supporting the diagnosis of Wilm's tumour. Imaging diagnosis was Wilms tumour and biopsy was advised for confirmation of the diagnosis which revealed a highly

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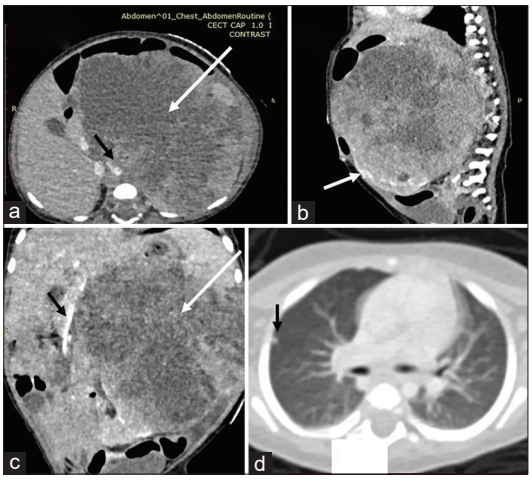


Figure 1: (a-d) CECT abdomen axial and reformatted scan at the level of left kidney shows large heterogeneous mass lesion with area of necrosis (large white arrow) involving left kidney. Vessels are displaced to left side (small black arrow in a and c). Residual enhancing parenchyma is seen at the lower pole. (small white arrow in b and c). (d) CT chest lung window demonstrates pleural-based nodule in right ling parenchyma s/o metastasis (small black arrow in d). CECT: Contrast-enhanced computerized tomography, CT: Computed tomography

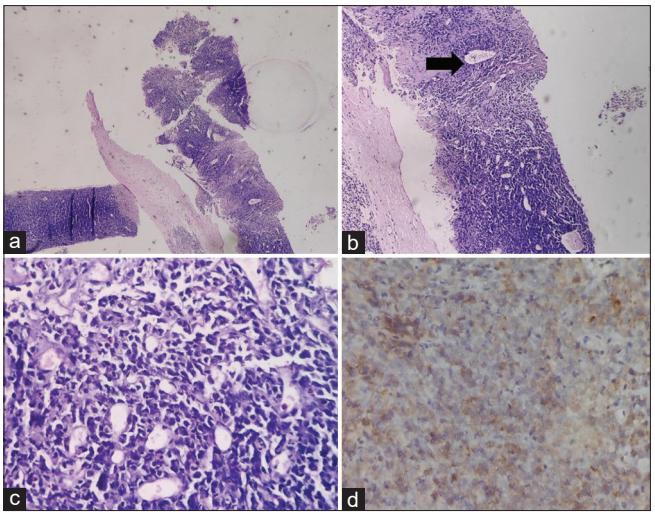


Figure 2: (a-d) (a) H and E ×4. Sheets of tumour cells surrounded by areas of fibrosis. (b) H and E ×10- highly cellular tumour showing perivascular arrangement and pseudorosettes (Black arrow). Marked crushing is seen on the edges. (c) ×40 H and E- small-sized tumour cells showing high n:c ratio, moulding, hyperchromatic nucleus, irregular nuclear margin and scant cytoplasm. (d) ×40 Synaptophysin IHC. Shows cytoplasmic positivity in tumour cells. IHC: Immunohistochemistry

cellular tumour showing the perivascular arrangement and pseudorosettes suggestive of NBL [Figure 2].

Discussion

NBL is the most common extracranial tumour in childhood[1] and manifests usually as an abdominal mass. Whereas Wilms' tumour, also known as nephroblastoma, is the most common renal tumour in childhood and it also presents as an abdominal mass. The clinical features, course of the disease and treatment are very different, thus, early differentiation amongst the two is critical. Both are the disease of early childhood, with NBL presenting earlier in the age group usually >2 years, and Wilms' having a slightly older peak incidence at between 3 and 4 years.

Histologically also, they have different origins with NBLs arising from primordial neural crest cells[1] and Wilms' arising from undifferentiated mesodermal cells.[2]

The most common location for NBLs is the adrenal gland, but it can arise in the sympathetic ganglia of the retroperitoneum, posterior mediastinum, neck or pelvis.[2] The organ of Zuckerkandl is a mass of neural crest tissue adjacent to the mid to distal abdominal aorta and it is another recognised site of disease.

Approximately 99% of Wilms's tumours arise from kidneys with only 0.5% to 1.5% arising from extrarenal sites.[3]

Both of these masses present as abdominal lumps if they are very large,[4] however, NBL can cause hypertension due to the compression of renal arteries or present as neoplastic syndromes such as opsomyoclonus and diarrhoea.[1] Wilm's tumour is either asymptomatic or 25% present with microscopic haematuria and 25% present with hypertension secondary to renin production.[2]

In our case, the child presented with a large abdominal mass with low-grade fever without any history of haematuria.

In our case, the mass was arising from the left kidney causing its inferiomedial displacement with extension into the pelvicalyceal system. Although the left adrenal gland was also not seen separately from the mass, the mass was predominantly intrarenal with suprarenal extension. It showed large areas of necrosis/cystic areas and no calcification. The vessels were also displaced to the right side rather than encased. Moreover, metastases were seen in the lungs. All these findings favoured the diagnosis of Wilm's tumour which was our imaging diagnosis. Age was the only parameter that favoured NBL.

Conclusion

Although NBL and Wilm's tumour have classical imaging features that can differentiate them, there are always exceptions to the rules. As in our case, all the imaging findings mimicked Wilm's tumour, but the final histopathological diagnosis was NBL. Hence, a radiologist must be aware and conscious of these fallacies and tread with caution, as the treatment for these tumours is quite different. The learning point from our case report is that NBL can present as renal mass and the imaging features of NBL and nephroblastoma can overlap and the final diagnosis is made on histopathology.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have

given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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