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Case Report

UNUSUAL PULMONARY PRESENTATION OF WILMS TUMOR MESTASTASIS DETECTED BY ABDOMINAL ULTRASOUND WITH REVIEW OF THE LITERATURE

[1]Okpala O. C., [1]Okafor C. O., [2]Osuigwe A., [1]Obasikene C. N., [1]Ume E. O.
[1]Department of Radiology, College of Health Sciences, Nnamdi Azikiwe University, Nnewi Campus
[2]Department of Surgery, College of Health Sciences, Nnamdi Azikiwe University, Nnewi Campus

ABSTRACT
We report a case of 9 years old boy that presented at 5 years with nephroblastoma. He had an unusual metastatic deposit at the right lung base after 3 years following surgery and courses of cytotoxics. Metastasis to the lungs following Nephroblastoma is common[1] but the presentation as an elevation of the right hemidiaphragm on a chest radiograph and a right basal mass are rare. The objective of the report is to highlight the fact that elevation of the diaphragm in patients with nephroblastoma may well be hiding a metastatic deposit.

Key words: *Wilms tumor, Metastasis, Ultrasonography.*

INTRODUCTION
Wilms tumor is the most common renal malignancy of childhood under 5 years. The annual incidence in the United States of America is about 7.5 million children under 15 years of age. The annual incidence in Nigeria is not known. The tumor has been diagnosed in the new born. It is both heritable and sporadic. It is inherited as an autosomal dominant trait with incomplete penetrance[2].

Presentation is most often seen as a protuberant abdomen. Radiological evaluation usually include:-

(a) Chest radiography
(b) Ultrasonography and
(C) Computerized Tomography.

These modalities will be able to establish the presence of a renal mass and a normal functioning contralateral kidney. They are useful in demonstrating the presence or absence of pulmonary metastasis.

Nephrectomy is the primary treatment and the decision for further therapy will depend on the stage of the tumor. Relapse occurs most often within 4 months of initial diagnosis and most common site of metastasis reported include; mandible, brain, paratesticular and duodenum. In the case reported here, the authors highlight the fact that it can mimic any of the causes of elevated hemi diaphragm.

CASE REPORT
C.O. is a 9 year old boy who presented at the age of 5 years in 2003 with abdominal mass and painless haematuria following trauma to the abdomen. Clinical examination suggested a diagnosis of nephroblastoma. Routine investigations ordered included urinalysis, which essentially showed no white blood cell but a red blood cell count of 1000. There was no bacteria growth seen in the culture. Serum urea / creatinine were normal, white cell count 5.3 x 10^9 d/l and platelet of 189 x 10^9/L were recorded. Geneotype is AA.

Abdominal ultrasound revealed an enlarged right kidney, 110mm in its longest axis with a mixed echo complex mass within the mid portion and extending to the superior pole. No caliectasis was noted. The left kidney 83mm in its longest axis had a normal echo texture. Other intra abdominal organs, liver gall bladder, pancreas and the spleen were normal. An impression of wilms tumor was made and intravenous urography advised.

For correspondence: Okpala O.C.

At Intravenous Urography (IVU), the right kidney showed a bulge on the superior lateral aspect of the nephrogram. There were no calyces demonstrated even after 2 hours. The left kidney outline was normal and the collecting system as well as the urinary bladder outline. An impression of a right renal mass at the upper pole was made with impaired renal function.

At surgery, the right kidney was enlarged with necrotic mass at the upper pole which ruptured on mobilization. Involvement of the lymph nodes was not clear except within the mesentery. A right nephrectomy was done and a mesenteric lymph node biopsy as well. Cytotoxic therapy was instituted with cyclophosphamide, actinomycin D and vincristine. Histology of the specimen consists of the kidney, rupture at the one of it's poles with accompanying friable grayish white tissue, measuring 10cm x 9cm x 6cm.

Microscopy of the histologic section showed sheets of primitive ovoid cells with deeply eosinophillic nucleic stroma. These primitive cells were seen forming tubular structures in some areas. The lymph node tissue showed hyperplastic changes but it is spared of infiltration by these primitive malignant cells. A diagnosis of nephroblastoma was made.

After 6 courses of cytotoxics, a follow up abdominal ultrasound, showed normal organs and no lymph nodal enlargement. Follow up chest x-ray was normal but for the elevated right hemidiaphragm, that was dense as well. The carina was not well defined and mediastinum was displaced to the left. Bony ribcage was intact.

An impression of right sided pleural effusion and a possible right lobe basal mass was made. A repeat abdominal ultrasound following the above chest x-ray findings showed a huge homogenous roundish mass seen "sitting" on the dome of the right hemi diaphragm, the central portion was hypoecoic and curvilinear. There was no associated pleural effusion. The liver was normal and the intra abdominal organs as well. There was no ascities.

DISCUSSION

Wilms tumor (Nephroblastoma) is a renal tumour that develops in childhood and one of the most common malignant tumors in children under 5 years[2].

The incidence of this disease is unknown in Nigeria and under reporting is a major contributory factor. Metastasis of this tumour has been reported involving many parts of the body [3,4,5,6,7].

Tracking metastasis of this tumour needs an efficient painstaking approach and the relevant facilities. With the dearth of pathologists Nigeria, it's often difficult to collaborate this effort. In a series reported from department of pediatric surgery at the India institute Of Medical Sciences, over 11 years, 101 cases of wilms tumour were seem. Of the thirteen that had metastasic disease at onset, 24 patients presented with relapse at a latter date. Our patient did not present with metastasis at onset. Risk factor associated with relapse was found to be unfavourable histology. Lymph node involvements, age more than 6 years, diffuse spill, capsular and vascular invasion and anaploidy. There was report of spill at surgery but no lymph node involvement was noted except in the mesentery. Mainstay of the management in most centers in Nigeria involves surgery and chemotherapy but elsewhere "judicious" use of other options are in place, chemotherapy and radiotherapy to the metastasic sites, second look surgery, resection of the pulmonary metastasis and use of CIS-Platinum. Diaphragmatic elevation of no obvious cause must be further investigated in such patients with a history of nephrectomy following a diagnosis of nephoblastoma with other imaging modalities other than the usual chest radiograph. The reporting session by radiologists should not stop at the common differential diagnosis of hemidiaphragmatic elevation like hepatic tumour, amoebic liver assess, subphrenic collections, phrenic nerve plasy[7]. In centres where investigative modalities are limited to plain radiographs, ultrasound can go a long way in making a diagnosis of Wilms Tumour metastasis. It

has been used to detect intracaval involvement.
In conclusion, we highlight the need to further investigate cases of wilms tumour for metastasis following resection with abdominal ultrasound. At follow up visits, this could be the first indication of metastasis especially if there is diaphragmatic elevation.

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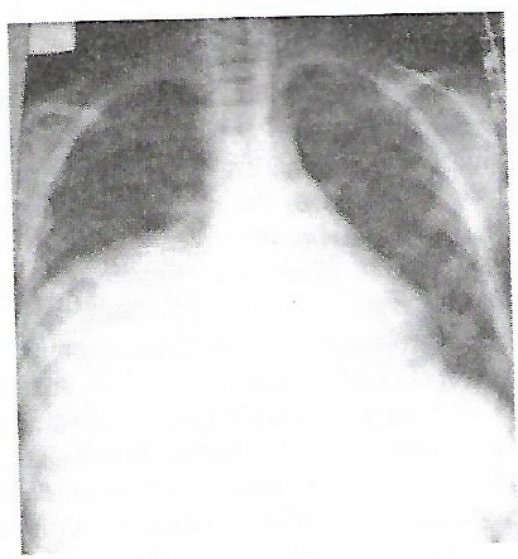
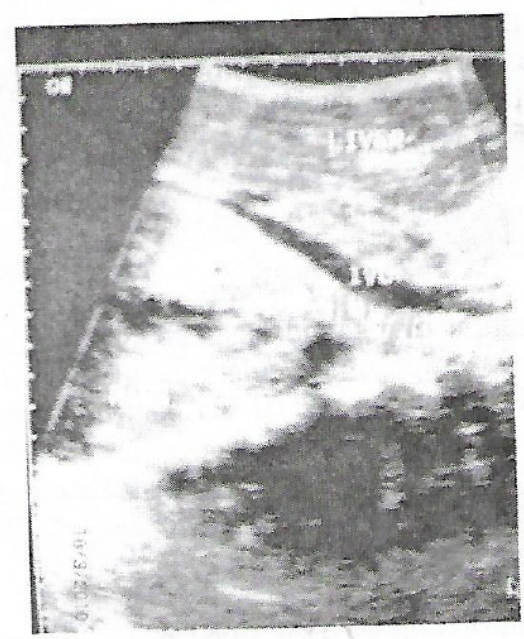


Chest X-ray



Liver