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Case Report

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WILMS TUMOR WITH LEFT ATRIAL EXTENSION: A RARE CASE REPORT

Dr. Parag Gharde[1], Dr. Amita Sharma[2], Dr. Bharti Sharma*[3] and Dr. Ritesh Kumar Sheorain[4]

[1]Professor, Department of Cardiac Anaesthesiology, AIIMS, New Delhi.
[2]Senior Resident, Department of Cardiac Anaesthesiology, AIIMS, New Delhi.
[3,4]Junior Resident, Department of Pathology, PGIMS, Rohtak.

***Corresponding Author: Dr. Bharti Sharma**

Junior Resident, Department of Pathology, PGIMS, Rohtak.

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ABSTRACT

Introduction: Wilms tumor is second most common abdominal solid tumor in children, also known as nephroblastoma. Intra atrial extension of Wilms tumor occurs in only about 1 to 3% of cases and its isolated metastasis to left heart in absence of vena cava extension is extremely rare, thus this case is unique. ***Case report:*** We reported a rare case of an isolated left heart metastasis of Wilms tumor in a 7 years old male child who presented with chief complaints of generalized weakness and loss of appetite for one month and chest pain associated with dyspnea for 10 days. ***Discussion:*** Wilms tumor is curable in majority of the cases. Metastatic tumors of the heart are 20 to 40 times more common than primary tumors. Isolated metastasis of Wilms tumor to left heart in absence of vena cava extension is extremely rare, thus this case is unique. ***Conclusion:*** Left sided heart metastasis of Wilms tumor is an extremely a rare condition. Preoperative chemotherapy and resection with an appropriately skilled team is the treatment of choice.

KEYWORDS: Wilms tumor, left atrium, intracardiac mass.

INTRODUCTION

Wilms tumor is second most common abdominal solid tumor in children. It is also known as nephroblastoma. Intravascular extension of Wilms tumor is a well-recognized phenomenon. Extension into the vena cava occurs in only 4 to 8% of cases and intra atrial extension of Wilms tumor occurs in about 1 to 3% of cases. It can grow for a long time without any characteristic symptoms, causing only abdominal pain, fever, vomiting or nausea.[1,2]

Several imaging modalities including computed tomography, magnetic resonance imaging and doppler ultrasound can demonstrate intravascular tumor. Cardiac ultrasound is presently the modality of choice for imaging space occupying lesions of heart.[3,4]

We reported a rare case of an isolated left heart metastasis of Wilms tumor in a 7 years old male child.

CASE REPORT

A 7 years old male child presented with chief complaints of generalized weakness and loss of appetite for one month and chest pain associated with dyspnea for 10 days. Further history revealed right nephrectomy 3 years back for removal of Wilms tumor. Following that patient received multiple cycles of radiotherapy and chemotherapy and he was symptom free for last 1 year.

For present complaints, patient was advised to undergo series of examination including CECT thorax, USG Doppler for inferior vena cava thrombus, echocardiography.

On examination his vitals were stable. His heart sounds were normal with no added sounds or murmur. Bilateral breath sounds were also normal. Per abdomen examination revealed no organomegaly. Hemogram was normal. Renal and liver functions were with in normal limits.

Chest X ray showed large homogenous opacity in the left upper lung field (figure 1). CECT thorax revealed large well defined hypodense mass in the middle mediastinum of about 36 X 50 mm size.

Transthoracic echocardiography revealed a large 4.5 X 3 cm homogenous mass attached to the free wall of left atrium (LA) via a narrow stalk and prolapsing into the left ventricle (LV) causing mildly enlarged left atrium and left ventricle. Severe pulmonary arterial hypertension and right ventricular dysfunction were documented. Intraoperative transesophageal echocardiography (TEE) showed a large 5.25 X 2.86 cm homogenous mass extending from free wall of left atrium with a narrow stalk of 1.24 cm. Mass was prolapsing into left ventricle. (figure 2).

Cardiac team removed the intracardiac mass from left atrium by performing sternotomy and cardiopulmonary bypass (figure 3) and the mass biopsy was taken.

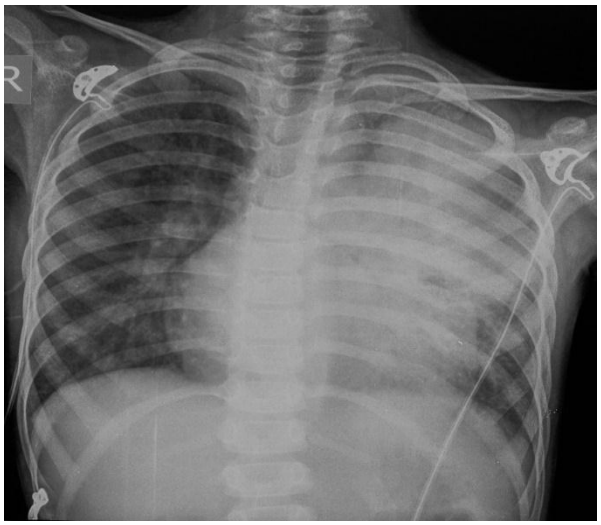

Figure 1: Chest X ray showing mediastinal mass extending into left lung field.

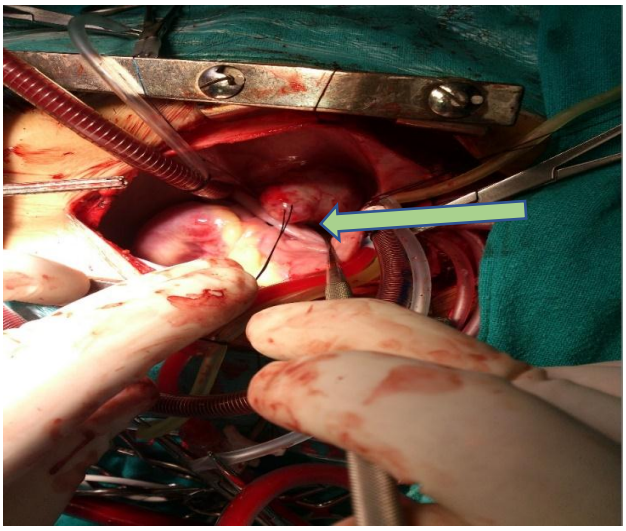

Figure 2: Intracardiac mass (green arrow) from left atrium (LA).

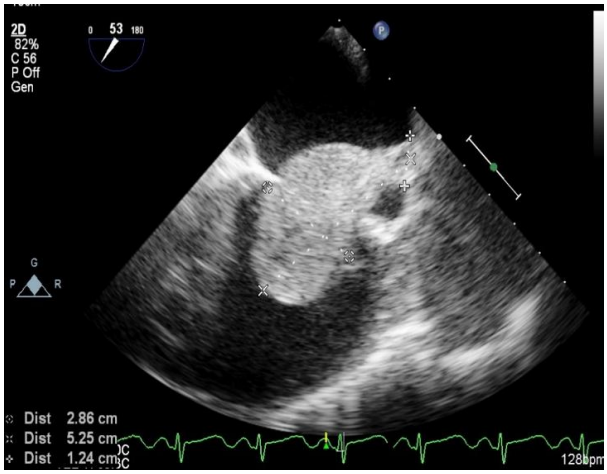

Figure 3: TEE showing LA mass with stalk prolapsing into LV through mitral valve.

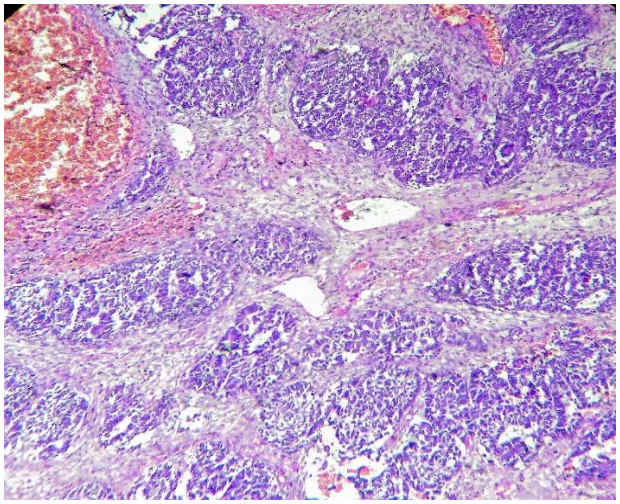

Figure 4: Photomicrograph showing Wilms tumor histology (H&E stained, 100X).

Histopathological examination of resected mass biopsy was found to be consistent with the histology of Wilms tumor. Microscopic examination showed malignant small round cells with immunopositivity for WT 1 and desmin and negative for myogenin. (figure 4).

DISCUSSION

Wilms tumor is curable in majority of the cases. More than 90% of patients survive 4 years after the diagnosis.[5] Metastatic tumors of the heart are 20 to 40 times more common than primary tumors. These metastasis involve the pericardium, myocardium and endocardium in decreasing order of frequency.[6]

Various routes of metastasis to heart includes hematogenous, lymphatic or direct invasion. Wilms tumor metastasis to heart mainly occur via inferior vena cava route. Isolated metastasis of Wilms tumor to left heart in absence of vena cava extension is extremely rare, thus this case is unique.

CONCLUSION

Isolated metastasis of Wilms tumor in left side of heart in absence of inferior vena cava extension is extremely a rare condition. Several imaging modalities including computed tomography, magnetic resonance imaging and doppler ultrasound used for imaging space occupying lesions of heart. Preoperative chemotherapy and resection with an appropriately skilled team is the treatment of choice.

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