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CARDIAC EXTENSION OF WILMS' TUMOR*

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Wilms' tumor comprises about 20 percent of solid tumors in childhood[1], and it ranks second among extracranial solid tumors[2]. The tumor exhibits a marked tendency to invade vascular structures such as the renal vein and inferior vena cava[2]. Cephalad progression of inferior vena caval involvement and direct cardiac extension of Wilms' tumor can occur. This complication has been documented by some authors preoperatively, intraoperatively, postoperatively or by postmortem observations[2-9]. With the new developments in noninvasive diagnostic methods the number of cases diagnosed preoperatively are increasing. Cases in which intracardiac tumors are diagnosed preoperatively present a technical challenge to the surgeon, and removal of the tumor requires cardiopulmonary bypass. If the tumor extending into the heart is removed intact, and without embolization, and if the patient is otherwise free of nodal or distant metastases, the disease can be considered to be in Stage II[3].

The clinical features of two patients with direct extension of Wilms' tumor into the renal vein and inferior vena cava and to the right atrium are presented in this paper.

Case Reports

Case 1

A four and a half year-old boy was brought to the Istanbul University Children's Hospital with complaints of body swelling and palpitation.

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Physical examination revealed a child with periorbital and pretibial edema. His liver was palpable three cm below the right costal margin. The cervical veins were distended. Maximal cardiac motion was detected in the xiphoid area. A II/VI systolic-diastolic murmur was audible around the same area. His blood pressure was 105/80 mm Hg and pulse rate 135/min. The hemoglobin and leukocyte counts were normal with a sedimentation rate of 22.40, and 60 mm at 30 minutes, one hour, and two hours, respectively. Urinalysis revealed 1+ albuminuria. Microscopic examination of the urine showed six or seven erythrocytes per field; the ECG was normal. The chest x-ray demonstrated cardiomegaly. M-mode and two-dimensional echocardiography revealed a large mass in the right atrium which moved from the right atrium into the tricuspid valve and the right ventricle and back into the right atrium in rhythm with the cardiac systole and diastole. As a result of these findings a tentative clinical diagnosis of a right atrial myxoma was made and surgery was decided. During open heart surgery a mass measuring 6 x 6 x 3 cm with an attached pedicle was discovered which almost completely filled the right atrium. The mass which extended into the inferior vena cava was removed and was diagnosed histopathologically as a mixed type of Wilms' tumor. Following surgery, intravenous pyelography and renal ultrasonography were performed which revealed a small mass in the left kidney (Fig. I). Ultrasonographic examination of the inferior vena cava showed fine linear

Fig. 1: Intravenous pyelography following cardiac surgery demonstrating deformation of the pelvis-caliceal system due to intrarenal tumor.

echogenic structures. Ultrasonography and scintigraphy revealed a normal spleen and liver. Bone marrow examination and bone x-rays demonstrated no metastases. Chemotherapy was initiated which was followed by a left nephrectomy, performed two weeks later. The extracted kidney was larger than normal, and a mass was noted at its upper pole. The renal capsule was intact. The area in and around the inferior vena cava was free of tumoral metastases. Histopathologically, the tumor was visualized in the branch of the renal vein as a thrombus-like mass (Fig. 2). A vena caval venogram using the percutaneous technique was taken postoperatively and revealed a minimal filling defect above the level of the renal vein. Radiotherapy was added to the treatment. After three and a half years, the patient is symptom-free, growing and developing normally.

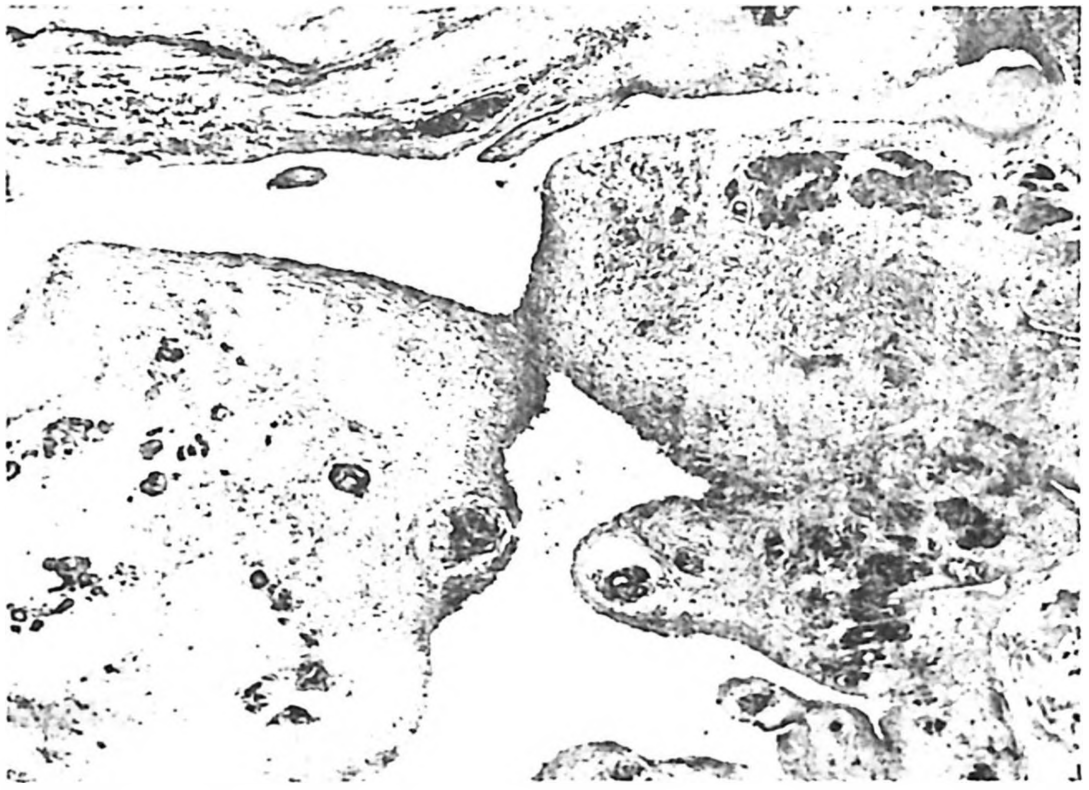

Fig. 2: Formation of the polypoid-shaped thrombus of the tumor in the renal vein branch.

Case 2

A seven-year-old boy was admitted to the Istanbul University Children's Hospital presenting with complaints of tiredness, periorbital edema, jaundice,and a reddish color to the urine. Six months prior to this admission the diagnosis of Wilms' tumor was established and the patient underwent a nephrectomy. He was also given chemotherapy.

Physical examination revealed dyspnea, diffuse edema and obvious jaundice, A pericardial friction rub, and I/VI pansystolic murmur in the xiphoid area were present. The cervical veins were engorged. The liver exceeded the costal margin by four cm. Laboratory examinations revealed a normal blood count and

sedimentation rate. SGOT was 918 U, SGPT 948 U. A chest x-ray demonstrated cardiomegaly and ECG low voltages over the left precordial leads. Two dimensional echocardiography revealed a mass in the right atrium with an attached pedicule. A fairly large amount of pericardial fluid was also present (Fig. 3). The patient's condition was poor. A preoperative diagnosis of extension of Wilms' tumor was established and surgery was planned. During open-heart surgery a 9x5x4 cm yellowish-white mass was removed. Similar to the first case, a pedicule extended into the inferior vena cava. Pathological examination of the mass revealed a Wilms' tumor rich in mesenchymal components (Fig. 4).

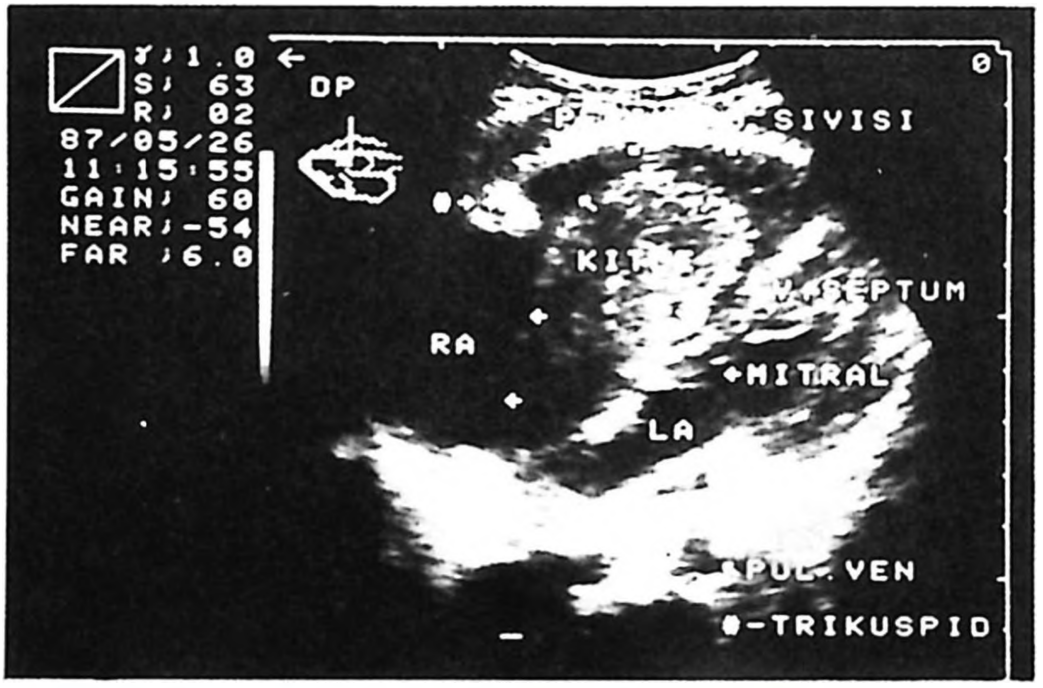

Fig. 3: Echocardiogram showing tumoral mass in the right atrium

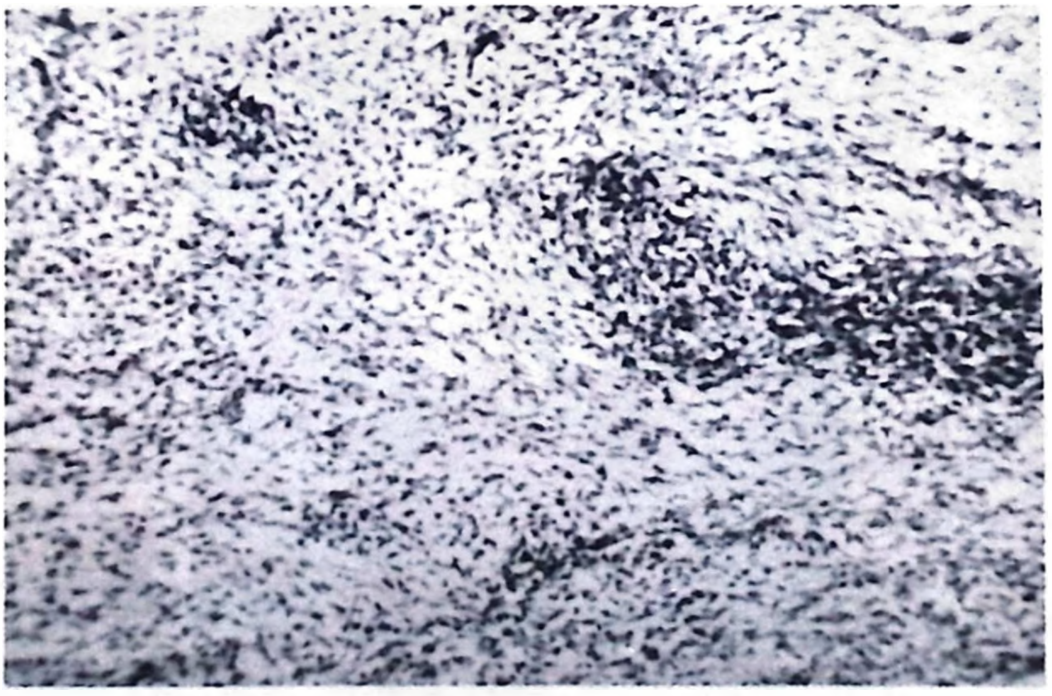

Fig. 4: Mesenchymal appearance and anaplastic cells

Postoperatively, the patient made a very rapid recovery. Work-up and treatment was similar to the first case. Two years postoperatively, the patient is still alive, well and symptom-free.

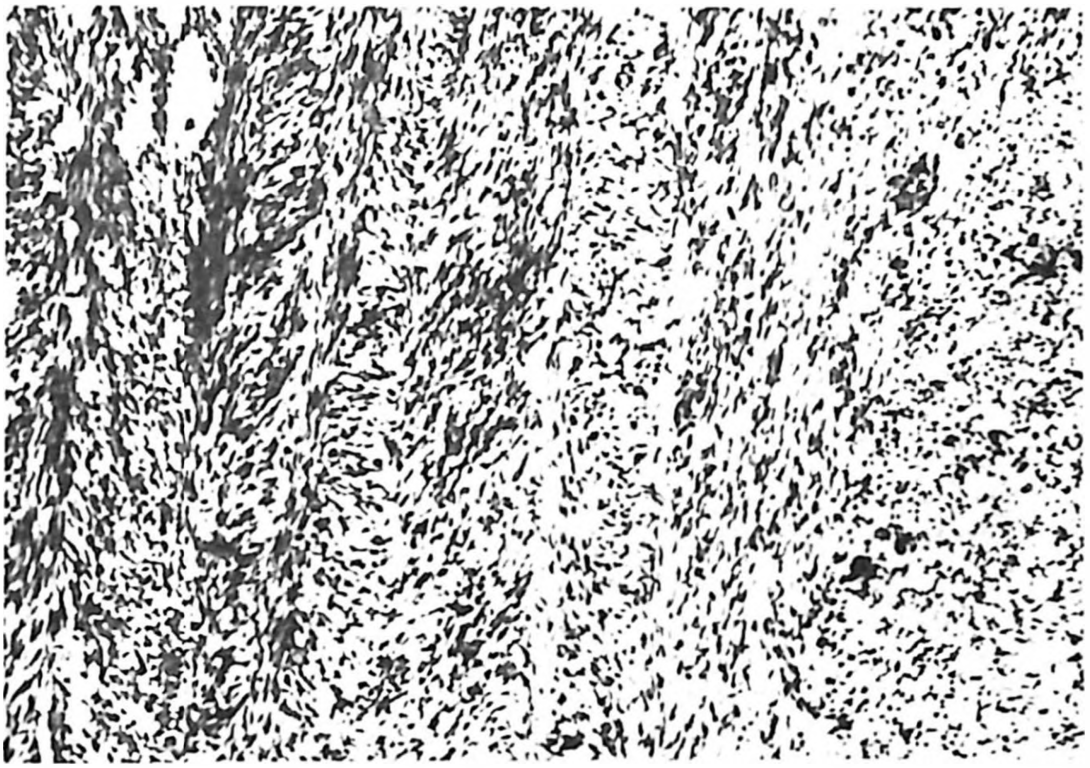

Fig. 4: Sections of the tumoral mass showing embryonic and mesenchymal characteristics in two different representative sections.

Discussion

Neoplastic diseases of the heart, pericardium and great veins, whether primary or secondary are unusual in children[4,5,10,11] Extension of Wilms' tumor into the inferior vena cava and the right atrium also rarely occurs. The National Wilms' Tumor Studies (NWTS) have reported the incidence of this tumor as 15:2280 (0.7 %)[3]. Only six of these 15 cases were diagnosed preoperatively. Of the remaining cases, five were diagnosed during surgery and four postoperatively. Chan et al[4], reported 293 Wilms' tumor cases in a series of 3641 solid tumor cases. Of these, nine had intracardiac metastases and three were diagnosed at postmortem examination. In this study the incidence of intracardiac metastases of Wilms' tumor was about three percent which is four times more than that reported by NWTS.

In our Children's Hospital thirty-two cases of Wilms' tumor have been diagnosed in the past 15 years. The two cases presented in this report are the first in which intracardiac metastases have been diagnosed. To date, there have been approximately forty reported cases of Wilms' tumor with intracardiac metastases[2-9], ten of which were reported prior to 1981. All had right kidney involvement, however, later reports mention both right and left kidney involve-

ment. In our cases, primary renal involvement was on the left side in the first case, and on the right side in the second case.

Cardiac metastases usually present with congestive heart failure and primarily symptoms of right-sided heart failure. Both our cases had findings of right-sided heart failure. In the first case, the mass was diagnosed by echocardiography, but initally the tumor was thought to be a myxoma and the diagnosis of Wilms' tumor was made postoperatively by pathological examinations. A similar case was reported by Holbrook et al[6]. In our second case, a correct diagnosis was made preoperatively since we had the experience of a prior case in which there was a history of surgical removal of a Wilms' tumor. Since echocardiography demonstrated a right atrial mass, emergency surgery was performed. Obviously this patient's condition was more critical because it was complicated by the presence of jaundice and the pathological findings of a congestive liver.

During a nephrectomy acute tricuspid valve obstruction and pulmonary embolism in the renal veins and the inferior vena cava with no metastatic invasion of these veins have been reported.[12,13] Thus, during surgery this has to be taken into consideration.

The best approach to intracardiac extension of Wilms' tumor is open-heart surgery for removal of the mass and extirpation of the pedicule from the inferior vena cava as extensively as possible[3]. Luck et al[7] and Grosfeld and Weber[8] advise a thoraco-abdominal approach for removal of both the intracardiac mass and nephrectomy in one surgical operation[7-8]. The NWTS results show that the two year survival of 15 cases with intracardiac extension is the same as for the noncardiac cases of Wilms' tumor. Also,preoperatively, intraoperatively or postoperatively diagnosed cases did not differ in prognosis[3]. For this reason, the same therapy program that was used in cases without cardiac extension can be applied to patients diagnosed preoperatively, intraoperatively or postoperatively. However, a preoperative correct diagnosis is to the benefit of the patient.

Intracardiac extension of Wilms' tumor is a rare but treatable condition provided that it is diagnosed early by routine echocardiography. All cases of Wilms' tumor should be scrutinized for this possibility using echocardiography which has brought a new perspective to this tumor by its easy applicability as a noninvasive method. This is true not only for diagnosis which saves the patient from costly and potentially risky tests but also for follow-up reevaluations.

Summary

Two children who were brought to the Istanbul Children's Hospital with congestive heart failure caused by extension of Wilms' tumor to the right atrium are presented. In both cases a large mass was noted in the right atrium by

two-dimensional echocardiography. The tumors were successfully removed at open heart surgery, and chemotherapy and radiotherapy were started postoperatively. The patients are both alive and symptom-free; one, three and a half years and the other to years postoperatively.

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