TUMOR IN HORSESHOE KIDNEYS: A CASE REPORT AND LITERATURE REVIEW

[1]Bobby Sutojo, [2]Ikhlas Arief Bramono, [2]Edward Usfie Harahap, [2]Rachmat Budi Santoso, [3]Galuh Ayu Treswari, [4]Rizky Ifandriani Putri, [5]Hardini Intan Setiawati Mahdi.

[1]Department of Urology, Faculty of Medicine/Universitas Indonesia, CiptoMangunkusumo General Hospital Jakarta.
[2]Urology Division, Dharmais National Cancer Hospital, Jakarta.
[3]Department of Radiology, Dharmais National Cancer Hospital, Jakarta.
[4]Department of Pathology Anatomy, Dharmais National Cancer Hospital, Jakarta.
[5]Department of Pediatric, Dharmais National Cancer Hospital, Jakarta.

ABSTRACT

Objective: *This study aims to report a renal tumor in a horseshoe kidney successfully treated with an open left nephrectomy.* ***Case(s) Presentation:*** *A 3 years old girl with a lump in the left abdomen and intermittent hematuria was referred to our hospital. Whole abdominal contrast CT scan showed the presence of a left renal mass in a horseshoe kidney that infiltrates the left renal artery and is attached to the spleen. The patient underwent preoperative chemotherapy followed by surgical resection.* ***Discussion:****In a horseshoe kidney, renal tumors are uncommon. The limited kidney mobilization and access to the renal hilum and the abnormal and highly variable vasculature make surgical treatment of a tumor in a horseshoe kidney extremely difficult.* ***Conclusion:*** *Open surgery continues to be the treatment of choice for horseshoe kidney tumors due to their anatomical complexity, particularly in cases where the tumor is difficult to eradicate.*

Keywords: *Renal tumor, horseshoe kidney, surgical management.*

ABSTRAK

Tujuan: *Penelitian ini bertujuan untuk melaporkan tumor ginjal pada ginjal tapal kuda yang berhasil diobati dengan nefrektomi kiri terbuka.* ***Presentasi Kasus:*** *Seorang anak perempuan berusia 3 tahun dengan benjolan di perut kiri dan hematuria intermiten dirujuk kerumah sakit. CT scan kontras seluruh perut menunjukkan adanya massa ginjal kiri di ginjal tapal kuda yang menyusup kearteri ginjal kiri dan menempel pada limpa. Pasien menjalani kemoterapi praoperasi dan diikuti dengan reseksi bedah.* ***Diskusi:****Pada horseshoe kidney, tumor ginjal jarang terjadi. Mobilisasi ginjal yang terbatas dan akses kehilus ginjal, serta pembuluh darah yang abnormal dan sangat bervariasi, membuat perawatan bedah tumor pada ginjal tapal kuda menjadi sangat sulit.* ***Simpulan:*** *Operasi terbuka terus menjadi pengobatan pilihan untuk tumor ginjal tapal kuda karena kompleksitas anatominya, terutama pada kasus dimana tumor sulit untuk diangkat.*

Kata kunci: *Tumor ginjal, horseshoe kidney, penatalaksanaan bedah.*

Correspondence: Bobby Sutojo, c/o: Department of Urology, Faculty of Medicine/Universitas Indonesia, CiptoMangunkusumo Hospital, JalanDiponegoro No. 71, Jakarta 10320, Indonesia. Email: bonifasius.bobby@gmail.com. Mobile Phone: +62812-8830-7383.

INTRODUCTION

Nephroblastoma, also known as Wilms' tumor, is the most frequently diagnosed pediatric renal mass, comprising 87% of all renal masses and 7% of all malignant tumors diagnosed in children.[1] This tumor is diagnosed at a median age of 3 years.[2] Approximately 1 in 500 people is affected by horseshoe kidney, with a male preponderance.[3] The incidence of Wilms' tumor in horseshoe kidneys is estimated to be between 0.4% and 0.9% of the total incidence of Wilms' tumors.[4]

Surgical planning is difficult due to anatomical factors when a tumor arises in a horseshoe kidney. Open surgery remains the standard treatment for such tumors. Therefore, horseshoe kidney tumor removal remains difficult. Here we reported a renal tumor in a horseshoe kidney successfully treated with an open left nephrectomy.

CASE(S) PRESENTATION

A 3-year-old girl was referred to our hospital with a lump in the left abdomen. Intermittent pain

and hematuria were also complained by her parents. An abdominal computed tomography (CT) scan was performed from the previous hospital and showed an isthmus connecting the right and left kidneys. A large mass was observed, measuring 12x12x11 cm, located in the left part of the horseshoe kidney. This mass showed heterogeneous enhancement with less enhancement relative to the normal kidney parenchyma.

Laboratory studies were done and showed complete blood counts, liver function tests, renal function tests, and urinalysis results were normal. Chest x-ray showed a metastatic nodule on the lung. Due to the large kidney tumor in this child, the initial diagnosis of Wilms' tumor was established. Because of the size of the tumor and the metastatic nodule on her lung, the patient underwent 6 weeks of chemotherapy with vincristine 0,8 mg, doxorubicin 30 mg, and dactinomycin 185 mcg. An evaluation CT scan was done and showed a reduction of mass to the size of 8x6x5 cm with infiltration to the left renal artery and attachment to the spleen. Aortocaval, left and right parailiac, and superior perimesenteric lymphadenopathy were also found (Figure 1). Chest x-ray showing metastatic nodule was diminished.

The patient was consulted to urology division for surgical resection. On arrival, physical examination, show a palpable mass in the left hemiabdomen with bulging on the left flank region. A left radical nephrectomy was done under general anesthesia. A transverse incision was made on the left side of the abdomen. The left kidney tumor was freed from the colon and spleen (Figure 2). The left kidney is connected to the right kidney resembling a "horseshoe kidney" (Figure 2). Left ureter was identified and ligated. The renal artery and vein were also ligated. The left kidney was resected, and the renal isthmus was cut and sent for a frozen section by anatomical pathology examination. Bleeding was controlled, the intrabdominal drain was left, and the wound was closed layer by layer.

The post-operative anatomic pathological report showing extensive necrosis of kidney tissue with the impression of tumor necrosis. The kidney

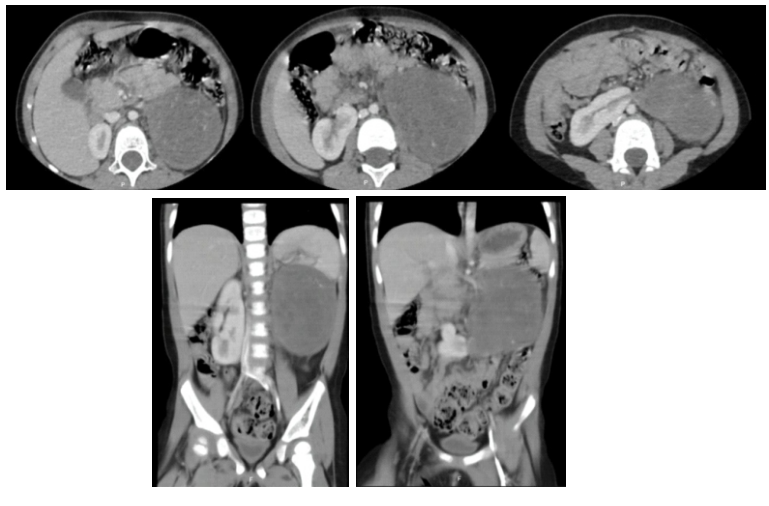

Figure 1. Contrast abdominal CT scan post chemotherapy.

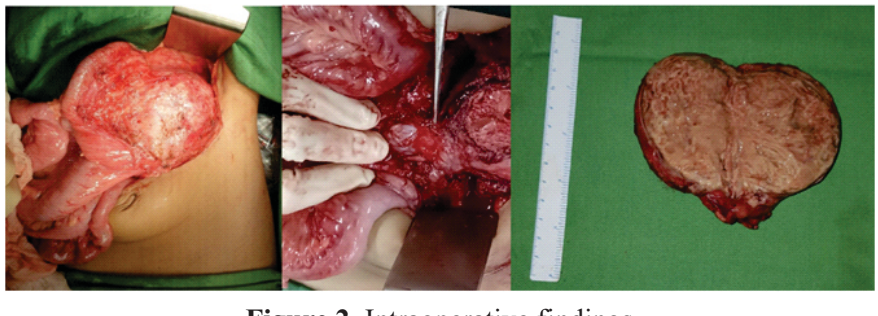

Figure 2. Intraoperative findings.

stromashowed a granulomatous reaction, presumably due to previous therapy, consisting of extensive fibrotic tissue filled with inflammatory cells of lymphocytes, foam cells, and DatiaLanghans cells. The ureteral incision margin did not contain a malignant tumor mass. (Figure 3)

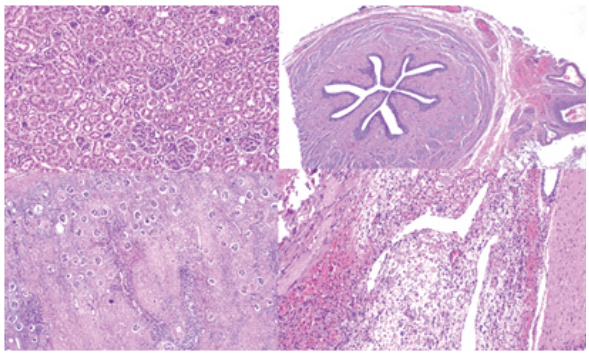

Figure 3. Histopathological Features of Tumor Tissue.

DISCUSSION

Horseshoe kidneys were first described by da Carpi in 1522 during autopsies and are the most common type of kidney fusion defect, with a reported incidence of 1:500.[3,5] Horseshoe kidney is characterized by abnormalities in renal position, rotation, and vascular supply.[6] Multiple etiological factors may contribute to the development of a horseshoe kidney, including intrauterine environment, genetic/chromosomal predisposition, and structural factors that influence the development and migration of the kidneys. The clinical relationships between horseshoe kidneys and secondary renal pathology, associated syndromes, and subsequent malignancy are significant.5In previous studies, most patients present with local symptoms, such as lumbar and abdominal pain, hematuria, and a palpable abdominal mass.

In a recent case series by Mano et al., all cases of renal cell carcinoma were discovered incidentally and were localized to the kidney at the time of diagnosis, whereas patients with upper tract urothelial carcinoma presented with hematuria.[7] In this case, the patient was presented with abdominal mass and intermittent hematuria. The majority of patients diagnosed with urothelial carcinoma exhibit hematuria.[8]

The disease's highly variable presentation is the greatest variable to consider for evaluating surgical treatment for tumors in this type of malformation. In 95% of cases, both moieties are fused symmetrically at their lower poles to form a "U" shape. It may be situated on the sacral promontory or, more commonly, between the third and fifth lumbar vertebra, caudal to the inferior mesenteric artery, and anterior to the great vessels.9 Vasculature varies considerably between individuals. In 63% of cases, there are three or more renal arteries with different origins (aorta, iliac, inferior mesenteric), and the isthmus is usually supplied by its blood vessels. Inferior vena cava anomalies (double, left, or pre-isthmic IVC) frequently accompany this condition.[5,9,10] The limited kidney mobilization and access to the renal hilum and the abnormal and highly variable vasculature make the surgical treatment of a tumor in a horseshoe kidney very challenging.[9-10]

Surgical indications for horseshoe kidney tumors are comparable to those for anatomically normal kidneys at this time. Surgical planning should include evaluating several essential factors: vascular and excretory system anomalies, the characteristics of the isthmus, the size, location, and complexity of the tumor, and the possibility of involvement of both renal units.[9] Imaging tests with intravenous contrast play a vital role in diagnosis and surgical planning, and new technologies such as 3D reconstruction provide high-quality data.[9,11] Arteriography, which was once performed on nearly all patients, has been supplanted by tests such as CT and MRI. Its main utility may lie in the embolization of the desired segment to reduce the risk of bleeding and facilitate surgery.[12]

Open transperitonealheminephrectomy, with an incision adapted to the location and size of the tumor, has been the conventional treatment for horseshoe kidney tumors. However, this type of surgery can present significant complications, notably bleeding.[13-14] Recent series suggest that the tumor may be treated with partial nephrectomy with minimal blood loss and preservation of renal function. However, relatively high complication rates are reported. Numerous authors have recently described laparoscopic surgery for a tumor in a horseshoe kidney.[15] We treated the patient with an open left nephrectomy in this case. The laparoscopic approach is unsuitable for our case because the tumor infiltrates the left renal arteries and is attached to the spleen. The types of incisions described are the midline transperitoneal, flank, and ilioinguinal approaches. Currently, there are no series comparing

these various incision approaches. In this case, we used a transversal incision along the abdomen.

Histopathological examination in this patient showed extensive tumor necrosis within kidney tissue. Non-viable cells were also seen, with pyknotic and hyperchromatic nuclei. The stroma showed a granulomatous reaction, presumably due to previous therapy, consisting of extensive fibrotic tissue filled with inflammatory cells of lymphocytes, foam cells, and Datia Langhans cells. There were foci of calcification between them and hyperemic vascular proliferation. Although the histopathological findings are inconclusive, we suspect this tumor is a Wilms tumor. Nephroblastoma (Wilms' tumor) is the most frequently diagnosed pediatric renal mass (87% of all renal masses) and 7% of pediatric malignant tumors.[16] The median age at diagnosis of this tumor is three years,[2] of which is relevant to our patient. The estimated incidence of Wilms' tumor in horseshoe kidneys is between 0.4% and 0.9%.16 Up to 13% of patients with Wilms' tumor develop a bilateral tumor.[17]

National Wilms' Tumor Study (NWTS) and the Societe Internationale D'oncologie Pediatrique (SIOP) have made significant contributions to the treatment of Wilms' tumor (SIOP). In 2001, NWTS and several other pediatric oncology cooperative groups merged to form the Children's Oncology Group (COG). Important components of Wilms' tumor treatment include surgical removal of the tumor, combination chemotherapy, and radiotherapy. The primary distinction between the NWTS/COG and SIOP treatment protocols is that SIOP routinely administers chemotherapy before surgery.[18-19] Different treatment strategies are due to different staging systems.[20] In most instances, the COG staging system is based on the pathology analysis from a primary nephrectomy. The staging of SIOP is based on the outcomes of preoperative chemotherapy.[19,21] The recommended algorithm for managingWilms' tumor as per NWTSG is given in Table 1.

The SIOP protocol algorithm for managing Wilms tumor is depicted in Table 2. Postoperative therapy consists of chemotherapeutic drugs that vary according to the stage of the tumor with or without lymph node involvement and the use of radiotherapy according to the stage and grade of the tumor; the specifics are shown in Table 3.

In this case, we give chemotherapy with vincristine 0,8 mg, doxorubicin 30 mg, and dactinomycin 185 mcg prior to nephrectomy

Table 1. Management of Wilms' tumor according to the NWTS protocol.

Stage	Treatment
Stage I FH/UH	18 weeks of DAM/VCR
Stage II FH	18 weeks of DAM/VCR
Stage III + IV FH	24 weeks of DAM/VCR/DOX, RT tumor bed + involved sites
Stage II–IV UH	24 weeks of DAM/VCR/DOX/CPM/Etoposide, RT tumor bed + involved sites

Table 2. Management of Wilms tumor according to the SIOP protocol.

Clinical staging		
Localized	4 weeks of DAM/VCR	Surgical staging
Metastatic	6 weeks of DAM/VCR/EPI	(Histological diagnosis)

DAM- Dactinomycin; VCR-Vincristine; EPI-Epirubicin.

Table 3. Post-operative regiment, according to SIOP.

Localized	Stage	Treatment
	Stage I, Low grade	none
	Stage I, Intermediate grade + anaplasia	18 weeks DAM/VCR 28 weeks DAM/VCR/EPI
	Stage II– (no lymph nodes)	28 weeks DAM/VCR/EPI + RT tumor bed.
	Stage II + and III High -grade Metastatic IV	34 weeks EPI/IF/VP16/CARBO + RT
Metastatic	IV	As per the local stage for tumor + treatment of metastases – RT and/or excision.

DAM- Dactinomycin; VCR-Vincristine; EPI-Epirubicin; IF-Ifosfamide; VP-16, Etoposide; Carbo-Carboplatin; RT-Radiotherapy.

because the patient is in stage IV disease. The COG recommendation suggests surgery as the initial treatment before chemotherapy. The COG only recommends preoperative chemotherapy under the following conditions: inoperable Wilms' tumor; solitary kidney; synchronous bilateral Wilms' tumor; tumor thrombus in the inferior vena cava extending above the level of the hepatic veins; tumor involving adjacent structures such that removal of the kidney tumor necessitates removal of other organs such as the spleen, pancreas, or colon; and with extensive pulmonary metastases.[19]

Whereas, the SIOP guideline recommends preoperative chemotherapy for all patients with Wilms tumor. Patients with unilateral localized tumors are suggested to be given 4-week vincristine (weekly) and dactinomycin (biweekly). Vincristine–dactinomycin for no longer than 9–12 weeks is recommended for patients with bilateral tumors, with the addition of doxorubicin for reinforcement in some patients. In addition, patients with metastasis are administered 6-week vincristine–dactinomycin regimen (similar to the one described above), and doxorubicin on weeks 1 and 5.[22]

Reevaluation is required in week 4 for non-metastatic Wilms' tumor and week 6 for metastatic Wilms' tumor. The investigation of choice involves contrast-enhanced CT of the thorax, abdomen, and pelvis. Abdominal radiology should note (as in the initial CT scan): (a) the maximum dimension of the tumor; and its size; (b) the presence and extent of necrosis; (c) the presence of thrombus; (d) lymph node status; (e) liver nodules (number, size, location); and (f) relationship with the aorta and inferior vena cava. As with the baseline chest radiograph, the chest radiograph should indicate the status of chest metastases as (a) present/absent, (b) unilateral/bilateral, and (c) the number of metastases on each side.[23]

Surgery is ideally performed during week 5 for non-metastatic Wilms' tumor and week 7 for metastatic disease. The optimal treatment consists of a radical nephroureterectomy and lymph node sampling. The surgery should be a transabdominal/ transperitoneal approach. The surgical notes must indicate whether the procedure was performed outside or within Gerota's fascia (per the institutional practice). Even without clinical or radiographic evidence of lymph node enlargement, lymph nodes should be sampled and examined histologically. One paracaval supra-hilar lymph node, one paracaval infra-hilar lymph node, one paraaortic supra-hilar lymph node, one paraaortic infra-hilar lymph node, both iliac lymph node, and one mesenteric lymph node are the minimum seven lymph nodes that must be sampled.[23]

Postoperative chemotherapy should be initiated as soon as the ileus subsides following surgery and is determined by postoperative histology and stage. The COG recommends routine postoperative or adjuvant chemotherapy for all patients with Wilms tumor, except those at very low risk: age 2 years old at diagnosis, stage I favorable histology, tumor sample weight 550g, and confirmed negative lymph nodes.[24] The SIOP recommends postoperative chemotherapy for all patients with Wilms tumors, except those with low-risk stage I tumors.[19] Radiotherapy should be initiated within 9–14 days of surgery unless medically contraindicated.[23] The COG recommends that all patients with stage III tumors receive postoperative radiation to the tumor bed.

The SIOP recommends whole-abdominal radiotherapy for patients with intermediate or high-risk histology tumors with significant preoperative or intraoperative tumor rupture or macroscopic peritoneal deposits; pulmonary radiotherapy is indicated for lung metastases lacking complete response until week ten postoperatively. Patients with a complete response to induction chemotherapy with or without surgery are not required to undergo pulmonary radiotherapy. Patients diagnosed with possible metastases or high-risk histology must undergo pulmonary radiotherapy. Regardless of histology, whole-lung irradiation is recommended for patients who did not receive lung irradiation during the initial treatment.[19] In this case, because the patient is in stage IV disease with pulmonary metastasis, the patient requires neoadjuvant chemotherapy and radiotherapy.

In addition to those previously described, there are treatments for special conditions such as stage V disease, Wilms tumor in infants, and recurrent cases.

CONCLUSION

Open surgery continues to be the treatment of choice for horseshoe kidney tumors due to their anatomical complexity, particularly in cases where the tumor is difficult to eradicate. Although controversy exists as to whether chemotherapy should be administered before or after surgery, we chose to administer chemotherapy prior to open

surgery to reduce tumor size and associated morbidity. Wilms tumor was suspected to be the cause of this kidney tumor.

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