Case report

Inferior vena cava thrombus shrinkage in patient with adult Wilms' tumor: A Case report

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ABSTRACT

Introduction and importance: Adult Wilms' tumor is infrequent in adults and usually has poor prognosis. We report a case of inferior vena cava thrombus shrinkage in patient with adult Wilms' tumor.

Case presentation: A 27-year-old male came to hospital due to pain in his left flank and repeated hematuria since one year ago. An inhomogeneous mass and thrombus in IVC and abdominal aorta near renal artery were found from his abdominal CT scan with contrast, so cytoreductive nephrectomy was performed. However, the patient was unstable during operation so the thrombus couldn't be operated. Post-op immuno-histo-chemistry evaluation confirmed the diagnosis of adult Wilms Tumor. A follow-up CT scan with contrast showed shrinkage of thrombus size with no sign of peripheral congestion. This phenomenon was rare in adult Wilms tumors, especially when the patient didn't undergo thrombus removal or adjuvant chemotherapy.

Clinical discussion: Adult Wilms' tumor tends to invade blood vessels in the form of thrombus, as shown in this case. Up to 4% of patients with thrombus had it around vena cava [3,4]. The recommended treatment is open surgery. Patients with chronic thrombus obstruction might have several venous collateral pathways to maintain blood flow to the right atrium. Generally, there are three major alternative pathways found in such complications.

Conclusion: This study presented a patient with adult Wilms' tumor that experienced post-operative thrombus shrinkage without doing adjuvant chemotherapy or thrombus removal during surgery.

1. Introduction

Wilms' tumor or nephroblastoma is a malignant embryonal tumor of the kidney made up of embryonic cells and commonly found among children below five years old [1,2]. It is infrequent in adults and usually has poor prognosis. Adult Wilms' tumor can be diagnosed using criteria outlined by Klinton et al. such as 1) tumor is identified as primary renal neoplasm 2) presence of primitive blastemic spindle or round cell component 3) formation of abortive or embryonal tubules or glomerular structures 4) absence of renal cell carcinoma around tumor area 5) histologically confirmed 6) age above 15 years old [2].

Adult Wilms' tumor has a strong tendency to invade blood vessels in the form of a thrombus. Previous studies found that thrombus extension along the renal vein into the inferior vena cava occurred in 4–10% of all patients [3]. Patients are usually asymptomatic and diagnosis is only made based on imaging examination. A multidisciplinary approach should be taken as spontaneous shrinkage of thrombus in adult Wilms' tumor has never been reported [3]. Due to its rare occurrence, there is no general consensus regarding the therapy of this disease in adults [3]. The case report has been reported in line with the Surgical Case Report (SCARE) guidelines [5].

2. Case presentation

A 27-year-old male came to the hospital due to pain in his left flank. The pain has been persistent, dull, and unaffected by physical activity in the past year. The patient also had red-colored urine since the pain started. He denied any history of weakness, nausea, vomiting, swelling of both extremities, shortness of breath, and loss of consciousness. Comorbidities such as hypertension, diabetes mellitus and prior urologic surgery were also denied. The patient is an active smoker, one pack a day. There is no history of allergy. His daily urine production is approximately 1500 cc a day.

All vital signs were within normal limits. From the physical

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a. IVC Thrombus by the *diameter of 4 cm and **length 10 cm seen at the level of right renal vein up until reaching opening of portal vein on IVC.

b. *Macroscopic examination of the tumour, **Inhomogenous mass in CT Scan with contrast.

Fig. 1. a. IVC Thrombus by the *diameter of 4 cm and **length 10 cm seen at the level of right renal vein up until reaching opening of portal vein on IVC. b. *Macroscopic examination of the tumor, **Inhomogenous mass in CT Scan with contrast.

Fig. 2. A, Anatomical Pathology appearance using H.E. staining. B, Immunohistochemistry appearance.
examination, localized tenderness and positive ballottement were only found around the left flank region. Both laboratory and imaging diagnostic tests were performed. The laboratory findings revealed an elevation in urea, creatinine and erythrocyte sedimentation rate (ESR). Chest X-ray of thorax gave normal result. An 85 mm × 82 mm × 52 mm bulging mass filled with fluid was found on abdominal ultrasound in the superolateral left kidney, which then was suspected as a hematoma. The abdominal CT Scan with contrast examination showed an enlarged left kidney with a bulge on the upper pole and an inhomogeneous mass around 13.97 cm × 8.89 cm × 8.81 cm. After contrast administration, the mass was assumed solid and cystic as it was greatly demarcated. Moreover, a thrombus in IVC was also identified (Fig. 1).

Left cytoreductive nephrectomy with regional dissection of lymph nodes excluding hematoma was performed. The hematoma was left out because patient was unstable during procedure. Therefore, the treatment course was done conservatively. Pathology reported 1 kg mass with the size of 18 cm × 12 cm × 9 cm in which 10 cm tumor and 5 cm normal renal parenchyma were identified. Histologically, four patterns were found; a relatively monotonous spherical cell proliferation, partially arranged tubules, pleomorphic nuclei, and clear cytoplasm with bits of cell necrotic. The perineal fat in one area was infiltrated with a left clear cell type renal cell carcinoma (nuclear grade III). Diagnosis of adult Wilms’ tumor was confirmed through immunohistochemistry examination. No residual tumor was found (Fig. 2).

Follow up was carried out with two post-op abdominal CT Scan with contrast examinations. Follow-ups were done in 3- and 6-month post-op, with the result of relatively unchanged thrombus size. The patient had no complaints or complications after the procedure, and the CT scan showed satisfactory results that showed shrinkage of thrombus size with no sign of peripheral congestion (Fig. 3).

3. Discussion

Adult Wilms’ tumor tends to invade blood vessels in the form of thrombus, as shown in this case. Up to 4% of patients with thrombus had it around vena cava [3,4]. The recommended treatment is open surgery. If the tumor is inoperable, neoadjuvant chemotherapy can be suggested to help shrink the tumor down. However, it’s not the case for this patient. The tumor shrunk down with conservative treatment. Compared to the size pre-surgery, post-op CT scan with contrast showed a great reduction of tumor size. Moreover, he had no complaints throughout the whole follow up period. The improvement was fascinating since such results
were almost unheard of in cases in which tumor was left untreated surgically [3]. There was only one study by Ritchey et al. that had the same approach and outcome [4].

Patients with chronic thrombus obstruction might have several venous collateral pathways to maintain blood flow to the right atrium. Generally, there are three major alternative pathways found in such complications. But again, it was not seen in this patient. Since there was no sign of peripheral congestion, the involvement of collateral vein was suspected. A previous study by Kalapurkal et al. stated that a multidisciplinary approach similar to the NWTSG management algorithm would increase the survival rate in adult patients with well-differentiated Wilms’ tumor. Few other studies had also reported better survival rate in adults Wilms’ tumor if treated with combined therapy algorithm in pediatrics since there is no guide created for the adult group yet [6].

4. Conclusion

This study presented a patient with adult Wilms’ tumor that experienced post-operative thrombus shrinkage without doing adjuvant chemotherapy or thrombus removal during surgery. No sign of peripheral congestion was found, suggesting the involvement of collateral vein. Such case is rarely found in adult Wilms’ tumor.

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Ethical approval

Ethical approval for this study was obtained from Padjadjaran University (LB.02.01/X.6.5/187/2021).

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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