Wilm's Tumor with Hypertensive Encephalopathy

Wilm's tumor is the second most common malignant retroperitoneal neoplasm in children(1). Hypertension in this tumor is present in 63% of cases reported in a series(2). However, severe hypertension is unusual in Wilm's tumor(3). We describe a child with Wilm's tumor who presented as hypertensive encephalopathy.

A 2-year-old male child was admitted with history of intermittent hematuria and a mass in the left lumbar area of 2 months duration, markedly irritable for 3 weeks and alteration in sensorium for 2 days prior to admission. No history of seizures was elicited. Examination revealed a drowsy child with mild degree of pallor. Blood pressure (BP) recorded with an appropriate size BP cuff was 220/170 mm Hg. Cardiovascular and nervous system examination was normal. Abdominal examination revealed a large left sided retroperitoneal mass. Examination of the fundus revealed Grade II changes of hypertensive retinopathy without papilledema or retinal hemorrhages. Injectable drugs like sodium nitroprusside or diazoxide were not available at that moment. Child was started on oral nifedipine 0.25 mg/kg/dose every 6 hours and captopril 0.5 mg/kg/day in divided doses. Sub-lingual (S/L) nifedipine (0.25 mg/kg/dose) was also used at times to maintain a diastolic blood pressure below 90 mm Hg. Oral hydralazine 3 mg/kg/day was subsequently added as BP was not fully under control. However, despite these drugs, diastolic BP was episodically rising up to 120 mm of Hg though systolic BP was under control.

Investigations revealed a normal urine examination and renal function tests. Abdominal ultrasound revealed a large echogenic mass in left kidney, with a patent inferior vena cava. The liver was normal. Intravenous pyelography (IVP) revealed distortion and displacement of calyces of left kidney. Chest skiagram excluded obvious pulmonary metastasis. Immediate surgery in form of a radical nephrectomy was done but some residual tumor was left as disease was in Stage III. Histopathology revealed Wilm's tumor with features of unfavourable histology.

On the first post-operative day, minimum diastolic blood pressure was 90 mm Hg, which further decreased to 70 mm Hg on the second day. Nifedipine (both S/L and oral) was stopped. Irritability disappeared and sensorium returned to normal on the third post-operative day. Subsequently other antihypertensives were also taken off. Blood pressure has remained normal after 8 weeks of follow up. Chemotherapy IV vincristine 1.5 mg/m^2 and actinomycin-D10mg/kg) and abdominal radiation with 2000 rads were given. The child remains normotensive on follow up.

The BP should be checked carefully in every case of suspected Wilm's tumor. At times, as exemplified by the present case, these subjects may have to be treated as an emergency. With a BP recording of 220/170 mm Hg along with central nervous sys-

tem symptoms which reversed once BP was controlled, a label of hypertensive encephalopathy can be given to the case. The absence of papilledema or hemorrhages precludes the diagnosis of malignant hypertension.

The mechanisms of hypertension in Wilm's tumor include compression of renal vasculature or parenchyma and renin production by the tumor itself(4). To know the pathogenesis of hypertension is very important to decide the antihypertensive agents. Early surgical removal of the tumor is advocated.

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REFERENCES

1. Green DM. The diagnosis and management of Wilm's tumor. Pediatr Clin North Am 1985, 32: 735-754.
2. Sukarochana K, Tolentine W, Kierewetter WB. Wilm's tumor and hypertension. J Pediatr Surg 1972, 7: 573-578.
3. Case Records of the Massachusetts General Hospital: Case 44061. N Engl J Med 1958, 258: 289-295.
4. Othersen HB. Wilm's tumor (Nehroblastoma). *In:* Pediatric Surgery Eds. Welch KJ, Randol PHJG, Ravitch MM, O'Neill JA, Rowe MI, 4th edn. Chicago, Year Book Medical Publishers Inc, 1986, pp 293-302.

Turner's Syndrome in a Neonate

Turner's syndrome, though relatively not so rare, is very uncommonly diagnosed in the early neonatal period. This report described one baby diagnosed as Turner's syndrome clinically at birth.

A full term female baby, a product of non-consanguinous marriage, weighing 2750 g, was born normally. Examination revealed edema of the dorsum of both upper and both lower extremities extending up to the base of the terminal phalanges (*Fig. 1*), webbing of the neck with loose posterior nuchal skin folds (*Fig. 2*); triangular face with micrognathia; large abnormal auricles, high arched palate, widely spaced nipples and hypoplastic nails.

There was no evidence of coarctation of aorta or hypertension. A skeletal survey and ultrasonography of the abdomen did not reveal any abnormality. Karyotyping revealed a genotype of 45 XO . The baby's FSH, LH and TSH levels were 160.4 MIU/ml (5-15 mIu/ml), 86.0 MIU/ml (5-15 mIu/ml) and 10.7 MIU/ml (0-9 mIu/ml), respectively.

Turner's syndrome was first described by Turner in 1938(1) and is caused by complete or partial monosomy of the short arm of 'X' chromosome, the incidence being 1 : 2500 female births. Almost 95% of all Turner's conceptuses are aborted. Fifty-seven per cent of all Turner's syndrome have 45 XO. Other chromosomal anomalies are--mosaicism (45X/46XX, 45X/47XXX, 45X/46XY) and isochromosomes(2). Patients presenting during the neonatal period are usually 45XO, as mosaics present later in life.

Investigations include karyotyping, FSH levels in blood and in urine, diabetes