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Neonatal Extra-Renal Wilm's Tumour

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Introduction

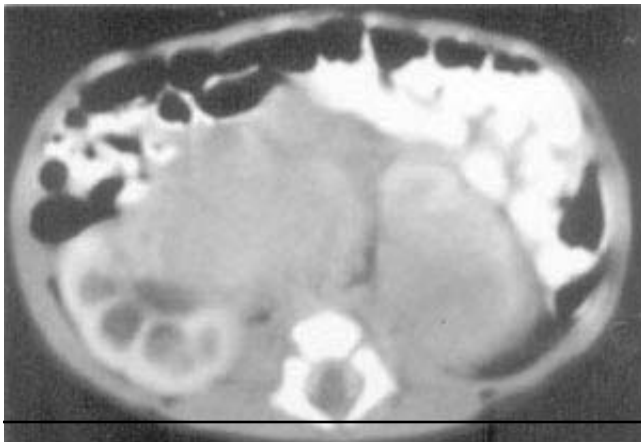
Wilms tumour is one of the most common solid neoplasia of the childhood and the most common tumour of genito-urinary tract in that age group.¹ Its origin in location other than kidney is rare. Only 57 such cases could be found in international literature mostly in infants and older children.² Wilms tumours are the malignancies with a characteristic histology seen typically during childhood and almost exclusively arising from the Kidney.³ In United States the incidence remained constant at 7.8 per million populations per year.⁴

Extra renal sites for origin of Wilm's tumour are extremely rare. The diagnosis of extra renal Wilms tumour is made only after a primary tumour of the kidneys has been ruled out with a secondary extra renal metastasis.⁵

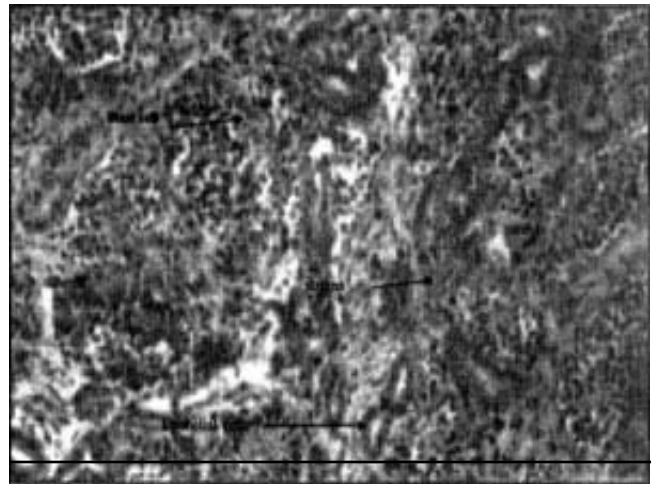
Below we report a case of extra-renal Wilm's tumour in a neonate with intra spinal extension through vertebral column.

Case Report

An eight days old male neonate presented with three days history of Jaundice, Abdominal distension and one episode of haematemesis. Clinical examination revealed a 3.0 kg lethargic and icteric neonate. His abdomen was distended with visible veins. A well defined abdominal mass was palpable in right Iliac fossa. Haematological and biochemical examination were normal except, total bilirubin 13 mg/dl, alpha-fetoprotein 6138 IU/ml; LDH 1463. Ultrasound scan and CT abdomen (Figure 1) revealed a 8x9cm heterogeneous mass in lower abdomen which was extending across the midline and also entering into spinal canal. Right kidney was hydronephrotic but both kidneys were free of any tumour and were well away from the mass.



On exploratory laparotomy a lobulated mass was present in lower abdomen retroperitoneally. It was extending from bifurcation of aorta up to coccyx and extending in both the flanks. On the right side mass was over ileo-psoas and on the left side it was extending behind the ileopsoas and was tethered to the vertebral column. The right iliac vessel and ureter was stretched over the tumour. Mass was excised completely but spinal canal was not opened. Histology showed mass to be well encapsulated, yellow and soft in consistency. Microscopically features were those of Wilms's tumour (Figure 2). In view of possible stage 3 disease with residual tumour chemotherapy was commenced. The follow up MRI, eight months after surgery, revealed persistence of soft tissue mass in spinal canal which was explored, mass removed but did not show any viable tumour. Two years following surgery, the child is doing well with no clinical or radiological recurrence.



Discussion

There are two types of extra renal Wilm's tumour.⁶ One arises in the line from the renal bed to the scrotum, supposedly from residual embryonic renal tissue.⁷ The second consists of teratome with nephroblastic tissue.⁸ The exact embryological origin is not clear. Multiple hypotheses have been proposed including a more primitive mesonephric or pronephric origin. Normally the mesonephros degenerates after fourth month of gestation. However, juxta-gonadal mesonephric glomeruli have been identified as late as 21st week of gestation.⁹ Thus the Wilm's tumour that is adjacent to the gonads may have a mesonephric origin. Likewise the pronephros, first and embryologically most primitive of the excretory organs,

arise in a cranial position, adjacent to the somites 9 to 12, which are lower thoracic in position.¹ Malignant transformation of aberrantly located cells derived from this tissue may account for chest wall tumours. The transformation of primitive embryological tissue into nephroblastome tissue is unknown. One theory is that the mesonephric conversion to Wilm's tumour requires two cellular events.¹⁰ The first results in the development of an aberrant rest of mesonephric remnant tissue. The second is malignant transformation of the mesonephric rest into Wilm's tumour. Twenty five percent of extra-renal Wilm's show the Wilm's tumour gene.³ Thus mutation of this gene may be the aetiology of extra-renal Wilm's tumour.

Most reported cases of extra renal Wilm's are in first four years of life. Only one has been reported in a neonate and oldest was a forty one year old.¹¹ The retro peritoneum being the most frequent location followed by inguinoscrotal area, sacrococcygeal teratoma, the mediastinum, and the lower chest wall.¹¹

The intra spinal extension has rarely been reported in literature. There is one case report of a four year old child, presenting with sudden onset of paraplegia and sacral tumour, in association with spina bifida. Imaging studies confirmed a sacral tumour with extradural extension up to T10 and spinal dysraphism. The histological features of sacral and intraspinal components of the mass was consistent with Wilm's tumour.¹²

Presently there is no staging for extra-renal Wilm's tumour. Coppe's series and the cases reported since show

that the prognosis and clinical course of extra renal Wilm's tumour is parallel to those of intra renal Wilm's tumour.¹³ Extra renal Wilms tumour should be treated according to NWTs protocol.

The unique features in our patient were a neonate with the origin of Wilms tumour at extrarenal sites from embryological elements and having an intraspinal component.

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