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## Medanta@Work

# Primary Intrarenal Teratoma in an Infant Masquerading as Multicystic Dysplastic Kidney

Primary intrarenal teratomas are incredibly rare tumours, with only about 20 cases reported globally. The sacrococcygeal region and the gonads are the most common sites of origin. Teratomas contain identifiable mature and immature cells or tissues derived from one or more of the three primordial germ layers.

In infancy, common flank masses include hydronephrosis, Wilms' tumour, cystic kidney diseases, and retroperitoneal malignancies such as neuroblastomas. Diagnosing a renal teratoma is challenging due to its rarity, the subtlety of its early symptoms, and its ability to mimic other tumours by containing tissues from multiple cell lines.

## Case Study

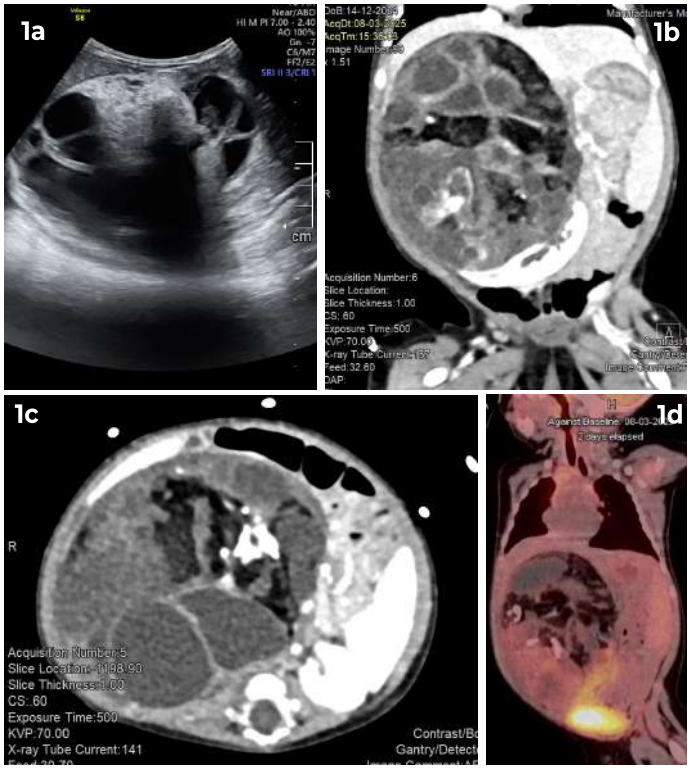
A 2-month-old female infant, with no anomalies detected on antenatal scans, presented with sudden-onset, rapidly progressing swelling in the right flank, poor feeding, and vomiting. She was referred to Medanta - Lucknow as a suspected case of multicystic dysplastic kidney (MCDK) based on ultrasonography at a local facility.

At presentation to our OPD, the mass had grown significantly, occasionally causing respiratory difficulty due to its size, along with reduced appetite. Despite this, the infant was alert and playful. On examination, a solid mass was palpable, extending from the inferior liver margin to the pelvic brim, crossing the midline medially.

Although initially presumed benign, the mass appeared more concerning on clinical evaluation. Infantile malignancies like neuroblastoma can present similarly and progress rapidly, necessitating urgent and accurate diagnosis.

A series of investigations followed: abdominal ultrasonography, contrast-enhanced CT (CECT KUB), 18F-FDG PET CT, and USG-guided biopsy with immunohistochemistry (IHC). Tumour markers (AFP, beta-HCG, LDH) and 24-hour urinary catecholamines were also assessed.

Imaging revealed a markedly enlarged right kidney measuring 11.8 × 7.3 cm, compressing the right lobe of the liver, extending medially to the midline and inferiorly to the iliac fossa. USG suggested MCDK with multiple non-communicating cortical cysts and loss of renal architecture.




1a: USG imaging of mass showing multiple non communicating cysts. 1b,1c: CECT KUB showing large encapsulated right renal mass with multiple cystic spaces and nodular calcification. Functioning renal parenchyma pushed down. 1d: PET CT imaging ruling out metastasis

CECT showed a well-encapsulated, oval, heterogeneous right renal mass (9.0 × 8.5 × 11 cm), with cystic areas, coarse calcifications, and macroscopic fat. Residual kidney tissue was displaced to the right iliac fossa. There was notable mass effect with displacement of bowel loops, scalloping of the liver, and compression or involvement of the aorta and IVC.

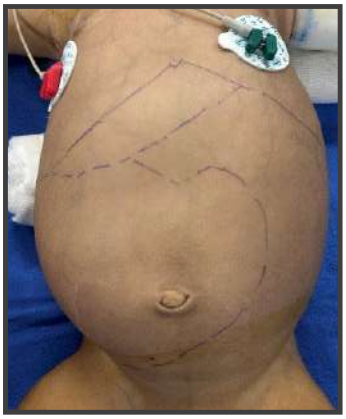
USG-guided biopsy of the solid areas revealed small undifferentiated blastemal cells in cords and nests, without significant anaplasia or atypical mitoses. WT1 and vimentin were patchily positive, suggesting a possible triphasic Wilms' tumour (favourable histology).

PET-CT showed faint metabolic activity in the mass, with no other areas of uptake. AFP was elevated at 1,755 ng/ml, while beta-HCG and LDH were within normal limits.

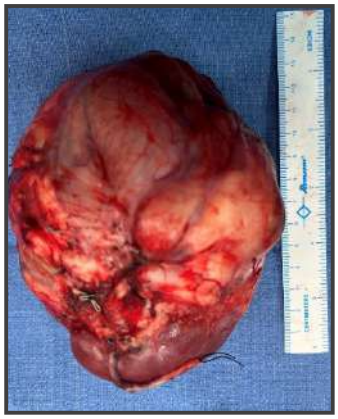
Following preoperative optimisation and with the cardiothoracic and vascular surgery team on standby, a high-risk radical right nephroureterectomy with lymph

node sampling was performed via a right transverse supraumbilical incision. A large, encapsulated mass was identified, displacing the bowel and adherent to the liver. Three major feeding vessels arose from the renal pedicle. The residual kidney was displaced to the iliac fossa.

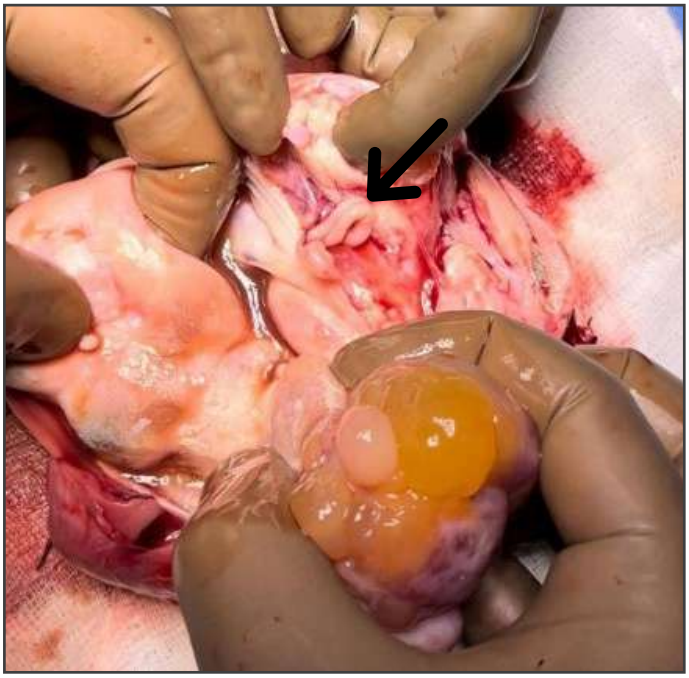
No lymphadenopathy or tumour thrombus was noted, and the IVC was freed from compression. The tumour was excised without spillage.



Preoperative surface marking of mass



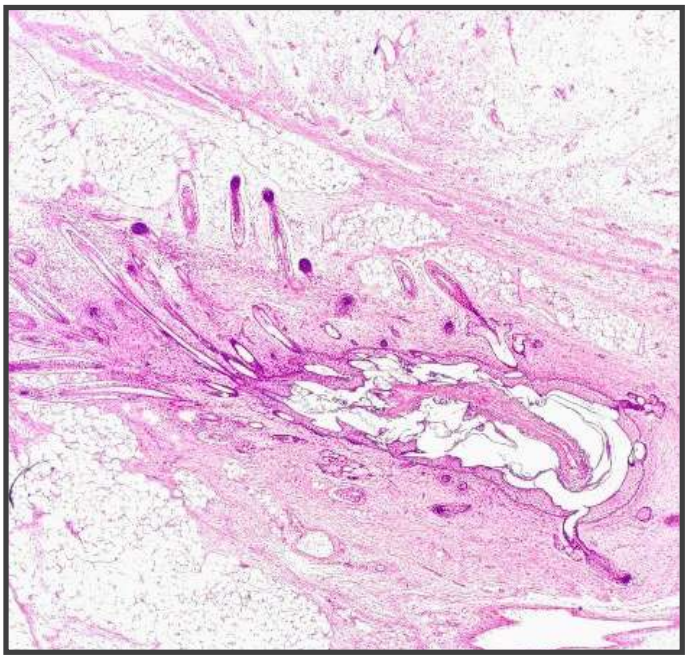
Gross specimen



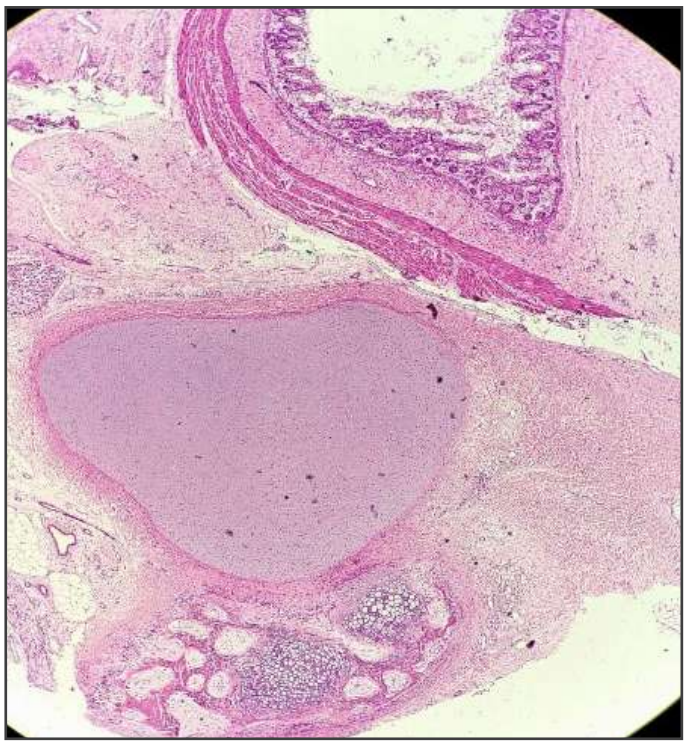
Specimen cut open revealing lanugo hair (left inferior), bowel with mesentery (arrow) and cystic areas

Blood loss was approximately 250 ml. The mass weighed 490 grams. Grossly, the specimen revealed structures from all germ layers, including lanugo hair, intestine with lumen, cartilage, bone, and Wharton's jelly.

Postoperatively, the infant developed a chyle leak, managed conservatively with a low-fat diet, medium-chain triglycerides, and octreotide. The abdominal drain was removed on day 5, and the child was discharged on day 7.



Cyst with squamous lining and hair follicles. Surrounding adipose tissue seen, H&E x40



Intestinal wall, cartilage and bone , H&E x40

Histopathology confirmed a teratoma with mature tissues (skin with hair follicles, bowel, respiratory epithelium, glial tissue, bone, cartilage, adrenal tissue, and more) along with immature neuroepithelial elements. Immature renal tissue composed of blastemal cells and tubules was also noted.

IHC showed WT1 positivity in blastemal and tubular cells, and negativity for synaptophysin. Focal yolk sac elements were confirmed by AFP immunostaining. The final diagnosis was intrarenal immature fetiform teratoma, Grade III, with focal yolk sac and nephroblastoma-like elements. No anaplasia, necrosis, rhabdoid or sarcomatoid features were seen. Margins, sampled lymph nodes, and peritoneal fluid were tumour-free.

The child has now been under follow-up for four months. AFP levels normalised by two months post-surgery, and USG abdomen remains normal. Chemotherapy has been kept as a backup, to be initiated only in case of recurrence.

## Discussion

Teratomas can imitate a wide range of conditions due to the presence of tissues from all germ layers. In this case, the fluid-filled spaces mimicked multicystic dysplastic kidney. With only around 20 reported renal teratomas globally, diagnosis requires high clinical suspicion.

To classify a lesion as a primary renal teratoma, two criteria must be met:

1. The tumour must originate entirely within the kidney.

2. It must show evidence of heterotopic organ formation.

Grading of immature teratomas is based on the number of low-power fields showing neuroepithelium. Grade III is defined as more than three fields with immature elements.

In this case, the clinical challenge lay in determining whether the mass was benign or malignant, and whether it was best treated surgically or with chemotherapy. Biopsy results can vary depending on site of sampling, and the decision must be based on a combination of histology, imaging, and clinical behaviour.

In infants, large masses can displace major vessels, making dissection and blood loss difficult to manage. Paediatric anaesthesia, intraoperative support (including CTVS backup), and meticulous dissection were key to the successful outcome.

As treatment guidelines for such rare tumours are not well defined, a conservative approach is often followed when histology does not show aggressive features. Regular monitoring with tumour markers and imaging is essential.

## Conclusion

Although rare, renal teratomas should be considered in the differential diagnosis of cystic renal lesions. Differentiating them from Wilms' tumour is essential to avoid unnecessary chemotherapy. Complete surgical excision remains the mainstay of treatment, with close follow-up and chemotherapy reserved for recurrence.

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