

## CASE REPORT

# A teratoid cyst containing nephrogenic tissue in a woman with a horseshoe kidney

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A 21 year old woman presenting with flank pain and vomiting was found to have a cystic lesion associated with a horseshoe kidney. The inner aspect of the cyst wall consisted of connective tissue intermingled with colonic-type epithelium. Within the cyst wall there were multiple foci of immature and incompletely differentiated renal elements, together with fragments of urothelium, smooth muscle, bone, and neuroendocrine tissue. No immature renal blastema were found. This lesion is unique and labelled as a teratoid cyst containing nephrogenic tissue.

Horseshoe kidneys are often associated with other congenital abnormalities that may be incompatible with life.<sup>1</sup> The possession of a horseshoe kidney alone does not itself result in shorter lifespan, although there does seem to be a higher incidence of tumours in horseshoe kidneys than in normal kidneys.<sup>2</sup> A surprisingly high number of tumours occur in the isthmus of the horseshoe kidney. This is thought to result from teratogenic factors responsible for abnormal migration of nephrogenic cells to form the isthmus, leading on to horseshoe development and increased potential for carcinoma development.<sup>3</sup>

“The possession of a horseshoe kidney alone does not itself result in shorter lifespan, although there does seem to be a higher incidence of tumours in horseshoe kidneys than in normal kidneys”

We report the first adult case of a teratoid cyst containing nephrogenic tissue, occurring in a patient with a horseshoe kidney and no other anatomical abnormalities.

### CASE REPORT

A 21 year old woman presented with right flank pain associated with vomiting and fever. An abdominal ultrasound showed a solid calcified mass arising from the anterior lower pole of the right kidney. Abdominal computed tomography revealed a horseshoe kidney with a 5.5 × 6.3 × 7 cm calcified mass lesion lying anterior to the right limb of the horseshoe; it was not clear if the mass originated from the gut or the kidney. A magnetic resonance imaging scan provided no further information. A biopsy of the mass was non-contributory with mucin obtained.

The patient continued to have severe pain, and requested surgical resection of the lesion. At surgery, the right limb of the horseshoe kidney was identified and a well circumscribed mass was identified adherent to the isthmus of the kidney, which was dissected off the isthmus of the kidney intact.

Pathological examination of the lesion revealed a 6 × 5 × 4 cm cyst (fig 1) with a thin focally calcified wall 0.4 cm thick and filled with yellow/green gelatinous material,

which was shown to be mucin histologically. Microscopically (fig 2A), the cyst had a fibromuscular capsule, which separated the cyst from the adjacent adherent renal parenchyma of the isthmus of the horseshoe kidney. The inner aspect of the cyst wall consisted of connective tissue intermingled with a variety of differentiated tissue types, of which colonic-type mucin secreting glandular epithelium predominated. The tissue external to the fibrous capsule comprised elements of mature renal cortex derived from the isthmus of the kidney. Within the cyst wall there were multiple foci of immature and incompletely differentiated renal elements (fig 2B), together with fragments of urothelium, smooth muscle, bone, and a small focus of neuroendocrine tissue resembling a carcinoid tumour. All the tissue was fully differentiated, or immature, including immature nephrogenic tubules and glomeruloid bodies. Renal tubular microcysts containing proteinaceous material were present, but no immature renal blastema were identified despite extensive tissue sampling.

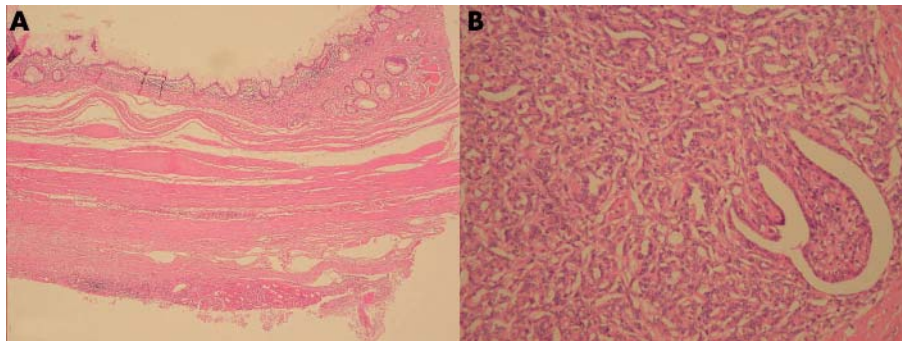
### DISCUSSION

Our patient presented a diagnostic dilemma. Despite numerous preoperative investigations, resection of the lesion was ultimately required for symptom relief and definitive diagnosis. The differential diagnosis was: benign differentiated teratoma, extrarenal cystic partially differentiated nephroblastoma, or a teratoid cyst containing nephrogenic tissue.

There were no epidermal structures present, so the lesion was not characteristic of an ovarian dermoid. The cyst was thought not to be a true teratoma, which is neoplastic, because renal elements are not a recognised feature of teratomas, and because elements from all three germ layers



**Figure 1** Bivalved surgical specimen: 6 × 5 × 4 cm cyst with a calcified wall, containing mucoid material.



**Figure 2** (A) Photomicrograph of the teratoid cyst. The cyst lining is at the top of the picture with colonic-type mucosa. There are primitive renal elements at the top right hand side. Normal renal parenchyma from the adjacent kidney is at the bottom of the picture, and the kidney is separated from the cyst by a dense capsule. Magnification objective,  $\times 10$ ; haematoxylin and eosin (H&E). (B) Photomicrograph of primitive renal structures including primitive tubules and a glomerulus. Magnification objective,  $\times 20$ ; H&E.

were not identified. This is in contrast to a similar case reported by Otani *et al*, where intrarenal cysts were lined by keratinising squamous epithelium continuous with transitional epithelium.<sup>4</sup>

Cystic partially differentiated nephroblastomas characteristically occur in paediatric kidneys, although they can occur in adults. However, the lesion did not have the typical features of a cystic partially differentiated nephroblastoma; in particular, the absence of immature renal blastema suggests that the diagnosis of nephroblastoma is unlikely.

“The location of this tumour in the midline of a horseshoe kidney supports the theory that abnormal development of the ureteric bud may have occurred”

This lesion is best regarded as a teratoid cyst containing nephrogenic tissue. The cyst contained mesodermal (renal) and endodermal (gut mucosa) components and was assessed as being a benign cyst. The term “teratoid” is justified to account for the variety of different differentiated tissues present. If this lesion had been intrarenal the nephrogenic rests would have been interpreted as nephroblastomatosis. Nephroblastomatosis is a developmental abnormality whereby persistent immature nephron elements exist after birth, and are often associated with abnormal ureteric bud growth. Ureteric bud epithelium in abnormal circumstances frequently undergoes metaplastic transformation to colonic-type epithelium. The location of this tumour in the midline of a horseshoe kidney supports the theory that abnormal development of the ureteric bud may have occurred. Emerson *et al* have demonstrated a genetic predisposition to nephroblastomatous development of germ cell tumours; however there are several differences between that case report and ours.<sup>5</sup>

### Take home messages

- We report a unique case of teratoid cyst containing nephrogenic tissue in a 21 year old woman with a horseshoe kidney
- Although no further active treatment is indicated longterm follow up was advised, but because this case is unique, it is impossible to be sure of the best follow up regimen

This lesion is similar to a previously reported case of a 6 month old infant presenting with an abdominal mass.<sup>6</sup> Histological examination of the resected tumour in that case revealed differentiated intermediate grade histology with primitive glomeruli and tubuli, and less than 10% blastema. Most of the resected tumour comprised cystic structures with colonic and urothelial epithelium as teratoid components. This lesion was described as a Wilms’ tumour with teratomatous cysts. There were several differences between this case and ours. Our patient was in her third decade of life, long after one would expect presentation with a congenital lesion. There were no blastemal elements found at all in the specimen, so that it is not a Wilms’ tumour. In the van der Poel *et al* case report<sup>6</sup> the horseshoe kidney had to be surgically divided, with the lower pole of the left kidney being sacrificed; in our case, the lesion was entirely extrarenal and the renal capsule was left in situ.

Variend *et al* described a teratoid Wilms’ tumour with the usual constituents of a nephroblastoma but with a large variety of epithelia.<sup>7</sup> Teratoid Wilms’ tumour has been described several times in the literature but is a different entity to the pathology described in our case.

Nephroblastomatosis and nephrogenic rests may be precursor’s to nephroblastoma; reassuringly for this patient, the development of Wilms’ tumour usually occurs before adulthood.<sup>8,9</sup> However, as an extrarenal mass, this lesion is one from which an extrarenal Wilms’ tumour may have arisen.<sup>9,10</sup> Although no further active treatment is indicated longterm follow up was advised. This will take the form of history, clinical examination, and repeated magnetic resonance imaging scans. As a unique case it is impossible to be sure of the best follow up regimen.

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The patient gave full consent for the publication of this case report.

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