CASE REPORT

Giant kidney worms in a patient with renal cell carcinoma

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SUMMARY

Diectophyma renale (*D. renale*), or giant kidney worms, are the largest nematodes that infect mammals. Approximately 20 cases of human infection have been reported. We present a case of a 71-year-old man with a recent history of unintentional weight loss and painless haematuria, passing elongated erythematous tissue via his urethra. CT revealed a left renal mass with pulmonary nodules and hepatic lesions. On microscopy, the erythematous tissue passed was identified as *D. renale*. On subsequent renal biopsy, pathology was consistent with renal cell carcinoma. This is the first reported case of concomitant *D. renale* infection and renal cell carcinoma, and the second reported case of *D. renale* infection of the left kidney alone.

BACKGROUND

When painless haematuria is associated with a mass in the flank, severe anaemia and significant weight loss, renal cell carcinoma is highly suspected. In our case, this constellation of symptoms was due to both renal cell carcinoma and a parasitic infestation of *Diectophyma renale* (*D. renale*), known as giant kidney worms. *D. renale* is the largest nematode that infects fish-eating mammals including mink, dogs, wolves, coyotes and foxes. Mammals are the definitive host as the nematode prefers to complete its life cycle in the mammalian kidney, with a preference for the right kidney.¹ A review of the literature revealed 20 cases of human infestation reported to date.¹ *D. renale* has been found in subcutaneous nodules,^{2–3} the right kidney,^{4–6} the left kidney,⁷ bilateral kidneys,^{1–8} the retroperitoneal space^{1–9} and the liver.¹⁰ One case also described an adult man in whom *D. renale* infection occurred simultaneously with malignant fibrous histiocytoma of the ureter.¹¹ This case is unique as it is the first reported case of human *D. renale* infection with associated renal cell carcinoma and the second reported case of *D. renale* infection of the left kidney alone.

CASE PRESENTATION

A 71-year-old African American male nurse presented with lightheadedness, a 10-month history of painless haematuria and significant unintentional weight loss. He also reported passing elongated red tissue via his urethra for 3 months. He was treated, at a clinic, for presumed schistosomiasis due to a remote history of baptism in the River Jordan. He noted some improvement after treatment with praziquantel and doxycycline. However, he continued to pass blood and elongated erythematous

matter via his urethra. He denied recent fresh water swimming and travelling, but reported weekly ingestion of local fish, caught in a river by his family, which may have been undercooked. There were no ill contacts with similar symptoms, to his knowledge.

INVESTIGATIONS

On physical examination, the patient was pale and cachectic. He was tachycardic with otherwise normal vital signs. Physical examination revealed hepatomegaly, a left sided flank mass and gross haematuria. He was severely anaemic with haemoglobin of 4.7 g/dL. Further laboratory testing revealed low albumin, elevated platelets and haematuria, but otherwise normal renal and liver function. CT with and without intravenous contrast demonstrated a left inferior pole renal mass measuring 11.2×12.3×14.9 cm with central necrosis and punctate calcifications invading the renal pelvis. There was hydronephrosis with diffuse thickening and enhancement in the left ureter, and a bladder filling defect (figure 1). Multiple pulmonary nodules and hepatic lesions were also present.

The erythematous matter passed via the urethra was collected and identified by microscopy as *D. renale*. The largest of these worms measured 20 cm in length with width of 5 mm, as shown in figure 2. The worms were red-brown, and tapered at the anterior and posterior ends with a thin striated cuticle. Examination of urine sediment was positive for 396 eosinophils but negative for *D. renale* ova as well as for malignant cells.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses included infestation with *D. renale* with a collection of worms in the kidney and bladder, with collections of larvae in the liver and lungs; metastatic renal cell carcinoma with concomitant infestation; and infestation with secondary malignancy.

TREATMENT

After he was admitted and stabilised with blood transfusions, this patient refused recommended invasive diagnostic and therapeutic procedures during his first admission for personal reasons. However, he agreed to treatment with antiparasitic medication and was treated as an outpatient with a 5-day course of ivermectin. He additionally received steroids and antihistamines to prevent a potential life-threatening inflammatory response. He denied side effects from this treatment, and his haematuria resolved. Repeat CT demonstrated a

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Figure 1 CT with and without intravenous contrast showing a left inferior pole renal mass measuring 11.2×12.3×14.9 cm, with central necrosis and punctate calcifications invading the renal pelvis, and a bladder filling defect.

stable renal mass and hepatic lesions but progression in size and number of pulmonary nodules. The patient continued to refuse invasive procedures.

OUTCOME AND FOLLOW-UP

Six months later, the patient was admitted with severe weight loss of 20 kg in 6 months, weakness and constipation. Laboratory testing revealed hypercalcaemia and anaemia, with haemoglobin of 6.1 g/dL. Repeat CT revealed intussusception of the small bowel with a stable renal mass and hepatic lesions but further progression of pulmonary nodules. The patient was transfused and his intussusception resolved with conservative management. He agreed to renal biopsy and the pathology was consistent with renal cell carcinoma. His Eastern Cooperative Oncology Group performance status was 3. Given his rapidly declining health, palliative care was recommended. He was discharged home, developed a bowel obstruction and died in a local hospital 2 months later.

DISCUSSION

In this report, we present a rare case of infection with *D. renale*, which, to our knowledge, is the first report of human *D. renale* infection with associated renal cell carcinoma and the second report of *D. renale* infection of the left kidney alone. Although previously reported cases of *D. renale* have been initially concerning for malignancy,^{1 4 6 9} this is the first case where pathology confirmed a concomitant renal cell carcinoma.

D. renale was first reported in 1782 in the canine kidney,¹ and the first report of *D. renale* infestation in a human host appeared in 1845.¹⁰ *D. renale* is a red-coloured nematode with a thin striated cuticle. The female worm is longer (20–100 cm) than the male worm (14–20 cm). The key to understanding *D. renale* infection is its complex life cycle depicted on the Center

Figure 2 Photograph of giant kidney worm measuring 20 cm in length and 0.5 cm in diameter.

for Disease Control website (see figure 3).¹² Definitive hosts are usually carnivores that harbour the adult worms and shed unembryonated ova in their urine. The first stage (L1) larvae develop in the ova after about 1 month in an aqueous environment. The ova are then ingested by annelids (eg, earthworms). In the gastrointestinal tract of these intermediate hosts, the L1 larvae differentiate into L2 and then L3 larvae. If the intermediate host is ingested by a paratenic host such as a fish or one of various amphibians, the L3 larvae encyst in the musculature and cease further development. The definitive hosts become infected by ingestion of these paratenic (fish, amphibians) or intermediate (annelids) hosts. Although the precise route of larval migration in the definitive host has not been defined, the larvae are believed to migrate preferentially to the right kidney either directly through the duodenal wall,¹³ or indirectly through the wall of the stomach to the liver before renal invasion,¹ due to the anatomic proximity of the organs.

Figure 3 Life cycle of giant kidney worm as depicted on the Centers for Disease Control and Prevention website.

In the definitive carnivorous host, the larvae mature into the male and female adult worms about 6 months following infection, and the female sheds both embryonated and unembryonated ova into the urine, thus completing the life cycle. Adult worms can survive in the definitive mammalian host for up to 5 years.

As with other mammalian hosts, it is surmised that humans contract *D. renale* by eating undercooked fish or frogs, or by drinking water containing infected annelids.¹² Based on case reports, the course of *D. renale* infection in humans is unclear. In some cases, the human becomes the definitive host as larvae mature into adult worms in the right kidney and then produce ova, which are found in the urine. However, there have been cases of ectopic parasitism where larvae migrate and mature into worms producing ova in the retroperitoneum⁹ and the liver.¹⁰ In other cases, the human is an accidental host, with larvae that do not go on to complete the life cycle being found

in subcutaneous nodules. If worms of only one sex are found in the kidney, the life cycle is also interrupted.

There are a few known parasites that affect the genitourinary tract. *D. renale* causes destruction by consuming the renal parenchyma and sparing only the capsule,⁷ which differentiates it from other parasites. In schistosomiasis caused by *Schistosoma haematobium*, egg deposition induces chronic bladder infection, calcification and fibrosis, with extension to the ureters and subsequent squamous cell carcinoma of the bladder.¹⁴ In visceral leishmaniasis, *Leishmania donovani* induces immune-complex mediated nephritis. Finally, malaria caused by *Plasmodium falciparum* results in acute tubular necrosis and renal cortical necrosis due to hypovolaemia and haemolysis secondary to erythrocyte sequestration. There is increasing evidence that chronic inflammation of parasitism predisposes to malignancy of the urinary tract.¹⁵ Specifically, schistosomiasis is associated with an increased risk of bladder cancer. This is theoretically due to

chronic irritation causing metaplastic changes, dysplasia and carcinoma. In addition, inflammatory cells and eosinophils stimulate growth factors, cytokines and proteases, which enhance tumour progression.¹⁶ Given the chronicity of *D. renale* infections and the invasion into the parenchyma, it is possible a similar theory could be applied with renal cell carcinoma. However, there is no association as this is the first reported case.

Due to destruction of the renal parenchyma, *D. renale* usually presents with haematuria and a renal mass with or without flank pain. In cases of ectopic parasitism, pain in the affected region has been the dominant symptom without haematuria. Some present with painless subcutaneous nodules caused by larvae.² In the present case, the left renal mass and filling defect in the bladder were likely due to a combination of a collection of adult worms and renal cell carcinoma, with the pulmonary and hepatic lesions being nodules caused either by larvae or metastases from the renal cell carcinoma. In the absence of surgical intervention or biopsy of the pulmonary and hepatic lesions, we can only speculate. The absence of ova in the urine suggests that the nematodes were either dead or belonged to a single sex. As the migration route of the larvae is not clearly defined, it is also unclear why the left kidney instead of the right kidney was infected.

Learning points

- ▶ *D. renale* infection is a rare cause of painless haematuria and unintentional weight loss.
- ▶ *D. renale* can infect the left kidney although the literature supports a preference for right kidney infestation.
- ▶ After treatment with ivermectin and a short steroid course, the haematuria and passing of the giant kidney worms resolved.
- ▶ *D. renale* infection can be associated with renal cell carcinoma.

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Contributors JK and LL, the primary authors, contributed equally to preparation of the manuscript. JK, LL and CMPH evaluated the patient, diagnosed the condition and managed the treatment. WMJ is the microbiologist who identified the worm. JK and LL were involved in collecting data, searching the literature and writing the manuscript. CMPH and WMJ assisted in submission and approval of the final draft.

Competing interests None declared.

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