CASE REPORT

Renal cell carcinoma with sarcomatoid transformation, presenting as skin rashes

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SUMMARY

A 68-year-old man presented to our outpatient department, with chief complaints of rashes all over the body. On physical examination, purpuric rashes were present over all four limbs and a lump was palpable in right flank. Contrast-enhanced CT scan showed enhancing large right renal mass. He underwent right radical nephrectomy, and histopathological analysis showed features of renal cell carcinoma. Skin lesions disappeared after surgery.

BACKGROUND

The classic triad of presentation in renal cell carcinoma (RCC), palpable mass, haematuria and flank pain is now called too late triad and is seen in <10% cases at presentation.¹ Most of the RCC is being diagnosed incidentally. RCC has been called as the great masquerader as it can have various clinical presentations involving almost any organ system.² Apart from constitutional symptoms, RCC can present with a variety of paraneoplastic manifestations. There are only a few cases reported in literature showing association of RCC with cutaneous vasculitis. We report a case of RCC, with focal papillary areas and sarcomatoid transformation presenting as purpuric skin rashes, which was managed with radical nephrectomy and cutaneous lesions subsided after the surgery. To the best of our knowledge, this is the second case in literature showing association of cutaneous vasculitis with sarcomatoid transformation of RCC.

CASE PRESENTATION

A 68-year-old man presented to our outpatient department, with chief complaints of rashes all over the body. There was no history of fever, itching or recent travel. There was no history of abdominal pain or haematuria. There was no history of any drug intake. On physical examination, there were no signs of pallor or icterus. Pulse was 74/min, blood pressure 140/80 mm Hg and chest was clear on auscultation. There was no lymphadenopathy. Purpuric rashes were present over all four limbs, more over lower limbs (figure 1A) which had multiple purpuric rashes, mostly discrete and 3–4 mm in size. The skin lesions were non-tender and were not palpable. On inspection there was fullness noted in the right flank region. A lump was palpable in right flank.

INVESTIGATIONS

An ultrasound scan of abdomen revealed right renal mass. Further evaluation with a contrast-enhanced CT scan showed enhancing large (12 cm × 10 cm) right renal mass with areas of necrosis within (figure 2). Patient was also evaluated for the skin lesions. Laboratory findings were haemoglobin 12 g/dL, total leucocytic counts 10 500/mm³, platelet count 243 × 10⁹/L, erythrocyte sedimentation rate was 15 mm/hour, anti-nuclear antibody (ANA) and laboratory evaluation for HIV, hepatitis C virus (HCV) and hepatitis B virus surface antigen (HBsAg) and a peripheral blood smear were all negative. Urine routine and microscopy showed no red blood cells and no protein.

DIFFERENTIAL DIAGNOSIS

The CT scan was suggestive of a malignant mass. The skin lesions were not thought of associated with the mass and a battery of investigations were done to find out the likely cause, which included complete blood count, peripheral blood smear, prothrombin time/partial thromboplastin time and bleeding time, anti-neutrophil cytoplasmic antibody and ANA for autoimmune and connect tissue disorders, rheumatoid factor, complement levels (C3, C4), laboratory evaluation for HIV, HCV and HBsAg were done and all were within normal limits. A skin biopsy was planned along with the planned nephrectomy for a definitive diagnosis.

TREATMENT

Patient underwent open right radical nephrectomy. It was a large tumour involving the mid and the lower pole and showed greyish white tumour, 12 cm in greatest diameter, with large areas of necrosis on bivalving. A skin biopsy was also done.

OUTCOME AND FOLLOW-UP

Histopathological analysis showed features of RCC with focal papillary areas and sarcomatoid transformation. Biopsy from skin showed perivascular mixed inflammatory infiltrate with few eosinophils, but definite fibrinoid necrosis was not seen. Skin lesions started resolving from postoperative day 2. At a 2-week follow-up, skin lesions had disappeared completely (figure 1B).

DISCUSSION

RCC has been called 'the great masquerader', for it can present with involvement of almost any organ system. The varied paraneoplastic syndromes

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Figure 1 (A) Skin rash at the time of presentation over lower limbs. (B)–Same site (lower limb) after nephrectomy.

can be divided into endocrine and non-endocrine syndromes.³ Constitutional symptoms like fever and weight loss are seen in 20%–30% of the patients and can be the sole presenting complaint in approximately 2% of cases.⁴ Although uncommon, RCC can present with various dermatological manifestations. Cutaneous metastasis is seen in 3%–7% of patients.⁵ Cutaneous paraneoplastic syndromes have also been reported with RCC and include bullous dermatosis, dermatomyositis, Henoch-Schönlein purpura, cutaneous sarcoidosis and cutaneous vasculitis.⁶ Association of cutaneous vasculitis with RCC has been rarely reported in literature. Hoag *et al* reported two cases of vasculitis associated with RCC.⁷ One patient had cutaneous leucocytoclastic vasculitis and the other had giant cell arteritis. In these two cases, the RCC was recognised at autopsy and cause and effect association could not be established. Another case of RCC reported by Mautner *et al* showed leucocytoclastic vasculitis in skin lesions, but this case was also associated with cryoglobulinemia.⁸ Lacour *et al* have reported two cases of clear cell RCC associated with leucocytoclastic vasculitis which resolved after nephrectomy.⁹ Andrasch *et al* reported a case of a 63-year-old woman who presented with cold-induced ischaemia and gangrene of digits along with hypergammaglobulinaemia and a renal mass. Skin biopsy showed a mild leucocytoclastic vasculitis, and histopathology of renal mass showed sarcomatoid adenocarcinoma.¹⁰ Her symptoms improved after nephrectomy.

There are no conclusive data in literature comparing the outcomes in patients with cutaneous paraneoplastic manifestations and those without it. Although early detection and treatment may improve the outcomes, the prognosis depends on

the underlying malignancy. To the best of our knowledge, this is the second case reported in literature showing association of sarcomatoid RCC with cutaneous vasculitis. In our case, the histopathology from renal mass showed focal papillary areas and sarcomatoid transformation, while biopsy from skin lesion showed a mixed perivascular inflammatory infiltrate but no definite fibrinoid necrosis was seen. The skin lesion disappeared completely after nephrectomy.

Learning points

- ▶ The classic triad of presentation in renal cell carcinoma (RCC), palpable mass, haematuria and flank pain is now called too late triad and is seen in <10% cases at presentation.
- ▶ RCC can present with a variety of paraneoplastic manifestations.
- ▶ These could be endocrine or non-endocrine in origin.
- ▶ Hypercalcaemia, hypertension, polycythaemia, non-metastatic hepatic dysfunction, Cushing's syndrome and altered glucose metabolism are the common endocrine paraneoplastic manifestations, hypercalcaemia being the most common.
- ▶ Amyloidosis, neuromyopathy, vasculopathy, cutaneous vasculitis and nephropathy are non-endocrine paraneoplastic manifestations.
- ▶ RCC has been truly called as the internists tumour, and although rare, cutaneous paraneoplastic manifestations must be kept in mind while evaluating a patient presenting primarily with skin lesions or with renal lesion associated with skin manifestations.

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REFERENCES

- 1 Motzer RJ, Bander NH, Nanus DM. Renal-cell carcinoma. *N Engl J Med* 1996;335:865–75.
- 2 Marshall FF, Walsh PC. Extrarenal manifestations of renal cell carcinoma. *J Urol* 1977;117:439–40.
- 3 Palapattu GS, Kristo B, Rajfer J. Paraneoplastic syndromes in urologic malignancy: the many faces of renal cell carcinoma. *Rev Urol* 2002;4:163–70.

Figure 2 Preoperative contrast-enhanced CT scan of abdomen showing right renal mass.

- 4 Laski ME, Vugrin D. Paraneoplastic syndromes in hypernephroma. *Semin Nephrol* 1987;7:123–30.
- 5 Lumpkin LR, Tschen JA. Renal cell carcinoma metastatic to skin. *Cutis* 1984;34:143–4.
- 6 Amin A, Burgess EF. Skin manifestations associated with kidney cancer. *Semin Oncol* 2016;43:408–12.
- 7 Hoag GN. Renal cell carcinoma and vasculitis: report of two cases. *J Surg Oncol* 1987;35:35–8.
- 8 Mautner G, Roth JS, Grossman ME. Leukocytoclastic vasculitis in association with cryoglobulinemia and renal cell carcinoma. *Nephron* 1993;63:356–7.
- 9 Lacour JP, Castanet J, Perrin C, *et al.* Cutaneous leukocytoclastic vasculitis and renal cancer: two cases. *Am J Med* 1993;94:104–8.
- 10 Andrasch RH, Bardana EJ, Porter JM, *et al.* Digital ischemia and gangrene preceding renal neoplasm. An association with sarcomatoid adenocarcinoma of the kidney. *Arch Intern Med* 1976;136:486–8.

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