CASE REPORT AND LITERATURE REVIEW

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SYSTEMIC SCLEROSIS PRESENTING AS A PARANEOPLASTIC SYNDROME IN RENAL CELL CARCINOMA: A RARE CASE REPORT

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Abstract

Systemic Sclerosis (SSc) is a chronic autoimmune disease of unknown aetiology characterized by vasculopathy and organ fibrosis. Renal cell carcinoma (RCC) is associated with paraneoplastic syndromes. RCC has been reported in association with paraneoplastic SSc, and eight cases have been reported since 1992 to date. In this case, we present a patient with SSc who showed rapid systemic progression within a period of 6 months. A mass was detected in the left kidney on imaging two months after the diagnosis of SSc, and the patient underwent a left partial nephrectomy. The histopathology of the mass was consistent with RCC. Despite intensive treatment, the patient developed progressive SSc involvement in the skin, lungs, and gastrointestinal tract within 6 months after the diagnosis of SSc. This case report emphasizes that SSc, which progresses with rapid and widespread systemic organ involvement, can occur as a paraneoplastic syndrome and that there should be a high suspicion for underlying malignant diseases in such cases.

Keywords: Paraneoplastic syndrome, renal cell carcinoma, systemic sclerosis

Introduction

Systemic Sclerosis (SSc) is a chronic autoimmune disease of unknown aetiology, characterized by vasculopathy and organ fibrosis. Multiple factors, such as genetic, environmental, infectious, and hormonal factors, are responsible for the disease's development. Previous studies have reported the association of paraneoplastic SSc with various neoplasms, especially breast, lung, and skin malignancies (1). However, the incidence of paraneoplastic SSc together with renal cell carcinoma (RCC) is quite rare, and only eight cases have been reported to date (1-8).

Case Report

We present a 68-year-old female patient who showed rapid SSc systemic involvement within 6 months under treatment with 15 mg/week oral methotrexate, 4 mg/day prednisone, 200 mg/day hydroxychloroquine, 100 mg/day acetylsalicylic acid, and 60 mg/day nifedipine. At the time of diagnosis, the patient had sclerodactyly, skin hardness, avascular areas, and dilated capillaries on capillaroscopy. There was no dysphagia, exertional dyspnea, or orthopnea. Laboratory values showed an anti-nuclear antibody nucleolar staining pattern of 1/3200 (++++) titer and positivity for an anti-scl70 (++++) titer. The patient developed abdominal pain 2 months after the diagnosis of SSc. Abdominal computed tomography (CT) (Figure 1), and subsequent positron emission tomography revealed a 32x14 mm mass with high fluorodeoxyglucose uptake in the left kidney. The pathology of the patient who underwent left partial nephrectomy was consistent with clear cell RCC. In histopathological examination, no tumour tissue or

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lymphovascular invasion was detected in the surgical margins of the tissue.

SSc systemic involvement developed in the skin, renal, pulmonary, and gastrointestinal (GI) systems over the specified periods. Digital ulcers increased, skin hardness extended to the elbow, effort dyspnea progressed, and uncontrolled hypertension and dysphagia developed. High-resolution CT showed lung involvement in a non-specific interstitial pneumonia pattern and dilated oesophagus. Despite 2 g/day mycophenolate mofetil treatment, the patient's GI and pulmonary system involvement progressed, and the patient reached a stage where she could no longer tolerate oral feeding. Despite 2 g/day mycophenolate mofetil treatment, the patient's GI and pulmonary system involvement progressed, and they became unable to tolerate oral feeding. Percutaneous endoscopic gastrostomy was performed; nutritional needs were met; and monthly cyclophosphamide treatment was started. The patient is being followed up during the third month of a monthly 1000 mg cyclophosphamide treatment course without progression of SSc complications.

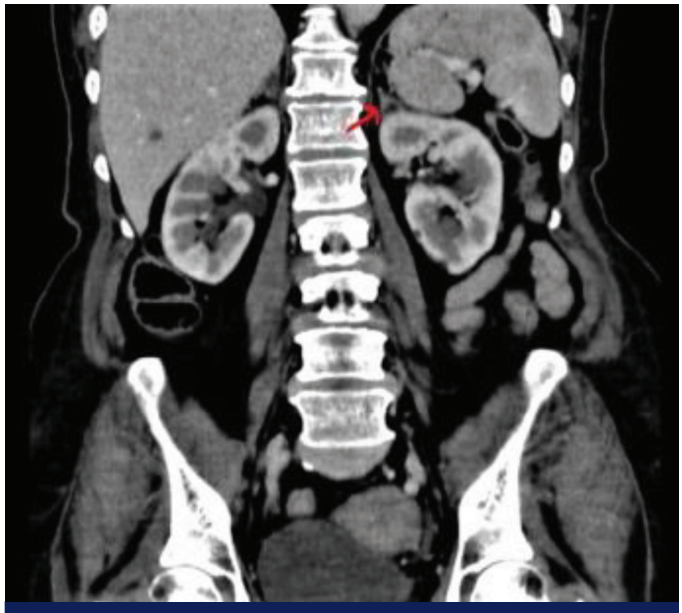

Figure 1. Contrast-enhanced abdominal computed tomography shows a 32x14 mm heterogeneously enhancing nodular lesion with exophytic extension in the upper pole of the left kidney (RCC)

RCC: Renal cell carcinoma

Discussion

This case report was prepared to emphasize that it may present as a paraneoplastic syndrome in RCC. In our case, shortly after the diagnosis of SSc, a complaint of abdominal pain developed, and RCC was subsequently detected on radiological imaging. In this case, SSc was considered a paraneoplastic syndrome of RCC. The patient underwent successful tumour resection. However, despite aggressive treatment, rapid progression occurred in the systemic organ involvement of SSc. Similar to our results, rapid progression occurred in the systemic involvement of SSc after nephrectomy, as reported in the literature, in some cases (1,2,6). At the same time, improvement in the clinical symptoms of SSc was observed after nephrectomy in some cases (3-5). Some reports indicated that RCC developed after a more extended period (approximately 2 years) after the diagnosis of SSc compared to our case (9). Table 1 shows literature studies reporting paraneoplastic SSc in RCC.

In this report, we emphasized the presence of underlying malignancies, especially in treatment-resistant cases, where the time between SSc diagnosis and systemic involvement is short. However, more research is needed on the importance of the close temporal relationship between RCC and the clinical

Table 1. Literature studies reporting paraneoplastic SSc seen in RCC

Age	Gender	RCC subtype	Timing of RCC to SSc onset	SSc outcome after nephrectomy	Reference*
68	Female	Clear cell	After 2 months	Progress	Our case
75	Male	Papillary	After 1 months	Progress	Patel et al. (1)
49	Female	Clear cell	After 7 months	Progress	Rutherford et al. (2)
55	Male	Unknown	After 14 months	Improvement	Nunez et al. (5)
75	Female	Unclassified type	3 months after progression of existing SSc	Improvement	Abrich et al. (4)
69	Male	Unknown	Unknown	Improvement	Angulo et al. (3)
31	Female	Unknown	After progression of existing SSc	Progress	Puett and Fuchs (6)
33	Female	Conventional type	After progression of existing SSc	Unknown	Eisenberg et al. (8)
37	Female	Conventional type	After progression of existing SSc	Unknown	Eisenberg et al. (8)

*Exponents indicate the reference number. RCC: Renal cell carcinoma, SSc: Systemic sclerosis

onset of SSc. Further studies in this area may provide insights into the pathogenesis of paraneoplastic SSc, which is especially prevalent in RCC. Furthermore, additional studies should clarify whether SSc is a paraneoplastic syndrome in RCC or a disease predisposing to RCC.

Conclusion

This case report emphasizes the need to investigate underlying malignant diseases in the presence of SSc, that are resistant to treatment and progress with organ involvement. Good recognition of paraneoplastic SSc that develops based on malignancy may provide early and practical approaches for diagnosing and treating severe systemic involvements such as pulmonary hypertension, malignant hypertension, interstitial lung disease, and GI involvement that may occur during the disease.

Ethics

Informed Consent: The Declaration of Helsinki was adhered to during the patient's treatment. The patient gave consent for all treatments and other procedures. The patient permitted publication as a case report.

Footnotes

Financial Disclosure: The authors declared that this study received no financial support.

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