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Polyarteritis nodosa with ureteric stenosis

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Abstract

Polyarteritis nodosa (PAN) is a primary systemic medium-vessel vasculitides. In systemic PAN, the most frequent skin lesions are purpura, livedo, and nodules. We report a case of PAN with stenosis of the ureter. A 60-year-old Japanese woman had a 5-year history of subcutaneous nodules, purpura, livedo-like dark erythema on the bilateral lower legs, with histopathology of necrotizing vasculitis of medium-sized arteries. Radiologically, hydronephrosis of both kidneys was found with stenosis of the ureters. She received radical nephrectomy of the right kidney and ureter and retention of ureteral stent for the left one. The removed ureter showed stenosis with prominent fibrosis of the lamina propria and muscular layer. We diagnosed the condition as systemic PAN with ureteric stenosis. Oral prednisolone exerted a good therapeutic effect. There have been reported 16 cases of systemic PAN with ureteric stenosis, including our case. Ureteric stenosis is one of the important manifestations of systemic PAN.

KEYWORDS

hydronephrosis, polyarteritis nodosa, purpura, ureteric stenosis, vasculitis

1 | INTRODUCTION

Polyarteritis nodosa (PAN) is a primary systemic medium-vessel vasculitides with heterogeneous presentations and disease course. The skin is a frequently affected site, and when the condition is single organ vasculitis, it is called cutaneous PAN.[1] In systemic PAN, the most frequent skin lesions are purpura, livedo, and nodules in the order of frequency, while cutaneous PAN mainly features nodules, livedo racemosa, and ulcerations.[2] PAN should be differentiated from microscopic polyangiitis (MPA) and other antineutrophil cytoplasmic antibody (ANCA)-related vasculitides.

We report a case of PAN with stenosis of the ureter. We first tentatively diagnosed the patient as having cutaneous PAN, but complication of hydronephrosis led to the diagnosis of systemic PAN.

2 | CASE REPORT

A 60-year-old Japanese woman was referred to us for evaluation of a 5-year history of subcutaneous nodules on the bilateral lower legs. She first developed a tender nodule on her left knee. During the following 2 years, the nodules increased in number and occurred on the right leg. The lower legs became edematous with pain. On examination, the patients had edema on the bilateral lower extremities with multiple subcutaneous nodules (Figure 1A). Most intriguingly, there were subcutaneous nodules with erythema and purpura on the left knee (Figure 1B). Livedo-like dark erythema was also noted. There was a painful nodule on the right leg (Figure 1C). A biopsy specimen taken from the nodule revealed necrotizing vasculitis of medium-sized arteries (Figure 1D). Fibrinoid degeneration with nuclear debris was observed within the lumen. Elastica van Gieson staining showed partial destruction of internal elastic lamina (Figure 1E). Laboratory examinations showed normal complete blood counts, liver enzyme levels, CK, BUN, and creatinine. However, albumin was low (3.4 g/dL; normal, 3.4-4.9 g/dL) and CRP was high (2.5 mg/dL; normal, <0.3 g/dL). IgA was elevated to 438 mg/dL (normal, 110-410 mg/dL), while IgG, IgM, C3, C4, and CH50 were within normal values. The following values were negative: anti-nuclear antibody, anti-cardiolipin antibody, PR3-ANCA, MPO-ANCA, and cryoglobulin. We tentatively diagnosed the patient for having cutaneous PAN.

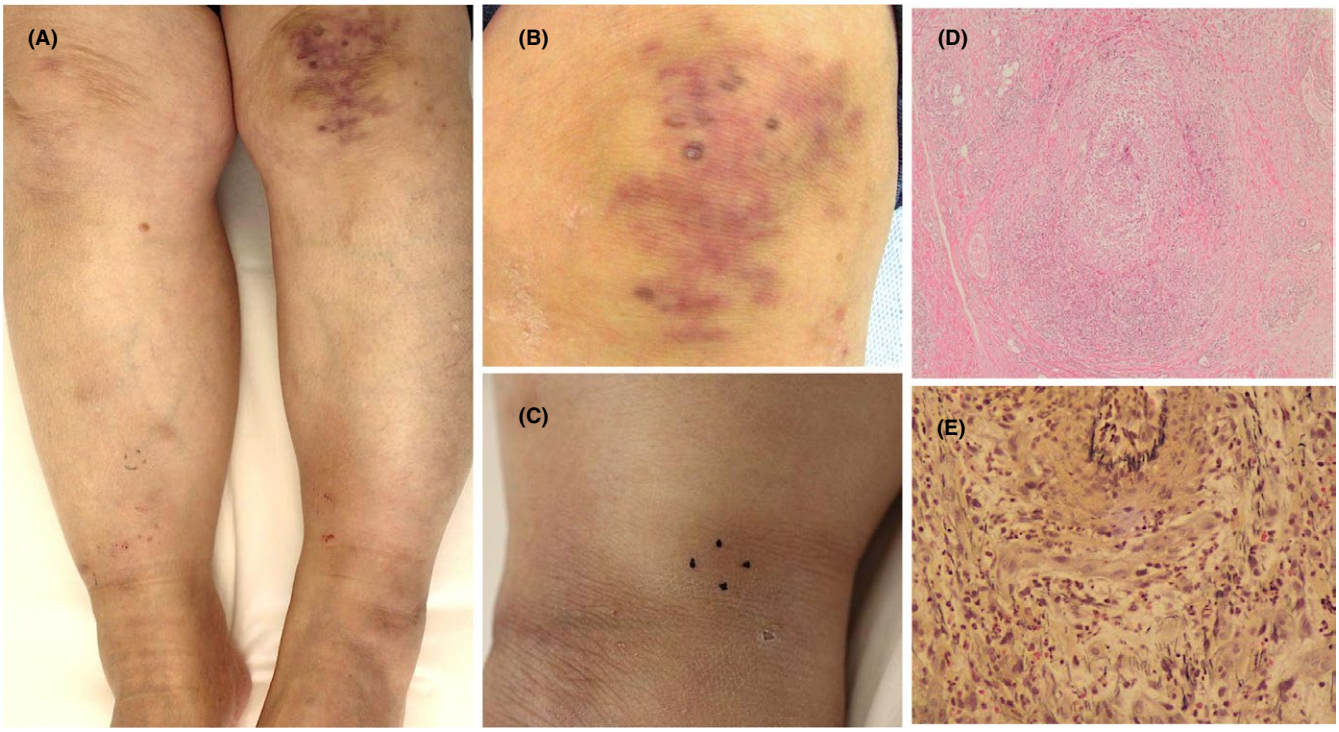

FIGURE 1 Clinical appearance and histopathological findings. A, Multiple subcutaneous nodules and edema on the lower legs. B, Multiple nodules, erythema, and livedo-like erythema and purpura on the left knee. C, Subcutaneous nodule on the right lower leg. D, Histopathology, showing necrotizing vasculitis of a medium-sized artery (hematoxylin & eosin). E, Elastica van Gieson staining, showing partial destruction of internal elastic lamina

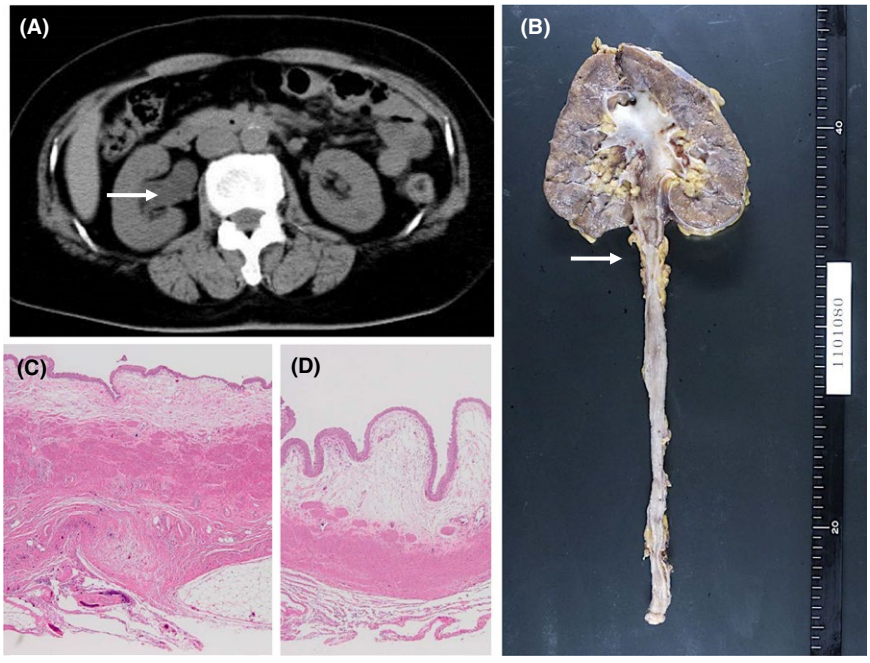

FIGURE 2 Examinations for kidneys and ureters. A, Computed tomography, showing hydronephrosis of both kidneys with stenosis of the ureters. Arrow: Right ureter. B, Removed kidney and ureter. Arrow: stenosis of ureter. C, Histopathology, stenotic part of the ureter. And D, non-stenotic part (hematoxylin & eosin)

To further evaluate edematous and painful lower limbs with hematuria (3+) and proteinuria (1+), urological examinations were performed. Computed tomography exhibited hydronephrosis of both kidneys with stenosis of the ureters (Figure 2A). The right kidney and ureter (arrow) were more severely affected than the left ones. She received radical nephrectomy of the right kidney and ureter and retention of ureteral stent for the left one. The removed ureter showed stenosis (Figure 2B, arrow). Histopathologically, there was more prominent fibrosis of the lamina propria and muscular layer in the stenotic part of the ureter (Figure 2C) than in the non-stenotic part (Figure 1D). We diagnosed the condition as systemic PAN with ureteric stenosis.

TABLE 1 Case reports of PAN with ureteric stenosis

Case	Age/Sex	Ureteric stenosis	Main symptoms	Operation	Treatment	Outcome	Reference
1	13/F	Both sides	Skin lesions, Arthralgias			Death	3
2	21/F	One side	Skin lesions, Neuritis	+		Free from symptoms	4
3	52/M	One side	Myalgias, Arthralgias	+		(Not described)	5
4	19/M	One side	Skin lesions		Steroids	Stenosis improvement	6
	6/M	Both sides	Skin lesions, Myalgias		Steroids	Stenosis improvement	
5	19/M	Both sides	Arthralgias		Steroids	Stenosis improvement	7
6	33/M	One side	Skin lesions, Myalgias, Neuritis	+	Steroids, Cyclophosphamide	(Not described)	8
7	30/F	Both sides	Myalgias	+	Steroids	Stenosis improvement	9
8	63/F	One side	SLE (suspected)	+	Steroids, Cyclophosphamide	Stenosis improvement	10
9	41/M	Both sides	Raynaud's phenomenon, Neuropathy	+	Steroids, Cyclophosphamide	(Not described)	11
10	34/M	Both sides	Skin lesions, Arthralgias		Steroids, Cyclophosphamide	Stenosis improvement	12
11	56/F	Both sides	Myalgias		Steroids	Stenosis improvement	13
12	55/M	Both sides	Skin lesions, Myalgias		Steroids, Azathioprine	Stenosis improvement	14
13	41/F	Both sides	Hepatitis B, Neuropathy		Steroids, Cyclophosphamide Plasma exchange	Stenosis improved	15
14	40/M	One side	Skin lesions	+	Steroids	(Not described)	16
15	20/F	Both sides	Skin lesions		Steroids, Azathioprine, Cyclophosphamide,	Stenosis improvement	17
16	60/F	Both sides	Skin lesions, Myalgias, Arthralgias	+	Steroids	Stenosis improvement	Our case

Following the operation, oral prednisolone at 20 mg daily was initiated. Serum creatinine and CRP were decreased 1 month later with alleviated hematuria. Currently, 2 years after operation, her skin lesions and renal condition were improved and prednisolone was tapered to 4 mg daily.

3 | DISCUSSION

We first diagnosed our patient's condition as cutaneous PAN; however, development of urological disorder and investigation of removed ureter clearly showed she had systemic PAN with ureter involvement. Corticosteroid therapy was effective for the renal condition.

To our knowledge, there have been reported 16 cases of systemic PAN with ureteric stenosis, including our case (Table 1).[3–17] The age ranged from 6 to 63 years. In 10 out of 16 cases, ureteric stenosis was also associated with skin involvement.[3,4,6,8,12,14,16,17] In at least 11 cases, corticosteroid improved stenosis of ureter,[6,7,9,10,12–15,17] indicating that efficacious treatment of PAN exerts a beneficial effect on ureteric stenosis.

It should be kept in mind that ureteric stenosis is one of the important manifestations of systemic PAN. Dermatological assessment may lead to the accurate diagnosis of PAN in such complicated cases.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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