References

1. Llach F, Arieff AI, Massry SG. Renal vein thrombosis and nephrotic syndrome: A prospective study of 36 adult patients. Ann Intern Med 1975;83:8-14.
2. Markowitz GS, Brignol F, Burns ER, Koenigsberg M, Folkert Renal VW. Renal vein thrombosis treated with thrombolytic therapy: Case report and brief review. Am J Kidney Dis 1995;25:801-6.
3. Clark RA, Wyatt GM, Colley DP. Renal vein thrombosis: An underdiagnosed complication of multiple renal abnormalities. Radiology 1979;132:43-50.
4. Mitchell DG, Friedman C, Druy EM, Swanberg LE, Phillips M. Xanthogranulomatous perinephritis: Unusual cause of renal vein and vena caval thrombosis. Urol Radiol 1985;7:35-8.
5. McHugh K, Stringer DA, Hebert D, Babiak CA. Simple renal cysts in children: Diagnosis and follow-up with US. Radiology 1991;178:383-5.
6. de Lichtenberg MH, Nielsen OS. Infected renal cyst simulating acute abdomen. Case report. Acta Chir Scand 1989;155:135.
7. Toprak U Erdoğan A Akar E Karademir AM. Infected renal cyst: Unusual cause of renal vein thrombosis. Eur J Rad Extra 2005;55:97-9.
8. Takeuchi N, Fujimura M, Sekita N, Suzuki H, Mikami K. An infected renal cyst communicating with the urinary tract: A case report (Article in Japanese). Hinyokika Kiyo 2014;60:485-8.
9. Suwabe T. Cyst infection in autosomal dominant polycystic kidney disease: Our experience at Toranomon Hospital and future issues. Clin Exp Nephrol 2020;24:748-61.
10. Libby P, Simon DI. Inflammation and thrombosis: The clot thickens. Circulation 2001;103:1718-20.
11. Shimad Y, Nagaba Y, Hagaba H, Kamata M, Murano J, *et al*. Edoxaban was effective for treating renal vein thrombosis in a patient with nephritic syndrome. Intern Med 2017;56:2307-10.

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An Abysmal Quadrad: A Rare Case of Ankylosing Spondylitis, Ulcerative Colitis, and Takayasu Aortoarteritis Complicated by Secondary Renal Amyloidosis

Abstract

Ankylosing Spondylitis (AS) is a chronic inflammatory arthritis that typically manifests in young males and may present with extra-articular manifestations. Takayasu aortoarteritis (TA) is a large vessel vasculitis that predominantly affects young and middle-aged females. Despite the limited number of studies examining the potential association between these two diseases, we report a unique case of an individual with ankylosing spondylitis and ulcerative colitis who subsequently developed Takayasu aortoarteritis. This progression ultimately led to the development of secondary renal amyloidosis, attributed to a combination of inflammatory pathologies.

Keywords: *Ankylosing spondylitis, amyloidosis, Takayasu, vasculitis, aortoarteritis*

Introduction

Ankylosing spondylitis (AS) is a chronic inflammatory disorder primarily affecting the axial skeleton. It is strongly associated with human leukocyte antigen (HLA) B27 and related conditions such as inflammatory bowel disease (IBD), reactive arthritis, and uveitis. Takayasu aortoarteritis (TA) is a large vessel vasculitis that primarily affects the aorta and its major branches. In this report, we present a rare case of a young male with AS and ulcerative colitis (UC), who later developed TA, the combination of which culminated into renal amyloidosis (RA).

Case Report

An 18-year-old male patient presented with a chronic history of low back pain and stiffness in the hips. On examination, the dorsolumbar spine showed limited ante- and lateral flexion [see Table 1 for detailed investigations]. He was diagnosed with AS and initiated on nonsteroidal anti-inflammatory drugs (NSAIDs) and physiotherapy. After 3 months, he developed frequent episodes of loose stools and fever. Colonoscopy revealed ulcers in the sigmoid colon, along with loss of vascular pattern in the transverse and ascending colon, with colonic biopsy showing cryptitis, crypt abscess, and crypt loss, consistent with UC. Treatment with sulfasalazine and prednisolone (1 mg/kg/d) was initiated, which was tapered after achieving disease remission.

Two years later, the patient presented with sudden-onset transient blurring of vision in the left eye with headache. Investigations showed raised inflammatory markers. Eye examination showed no signs of uveitis. A thorough physical examination revealed a blood pressure (BP) difference between the left arm (124/78 mmHg) and the right arm (150/90 mmHg), with low pulse volume in the left arm and a bruit over the left common carotid artery (CCA). Magnetic resonance (MR) aortography [Figure 1] showed wall thickening and enhancement in the aortic arch (AoA), descending thoracic aorta (DTA), and upper abdominal

Table 1: All the important investigations at the time of diagnosis of AS, UC, TA, and RA

Investigations	At the time of AS diagnosis	At the time of UC diagnosis	At the time of TA diagnosis	At the time of AKI diagnosis	At the time of RA diagnosis	On treatment (current)
Hb (g/dl)	10.4	10.4	12.4	9.8	8.3	9.5
WBC (per mm^3)	11,860	18,500	12,900	13,420	14,300	8900
CRP (mg/dl)	181.9	7	95.6	63.9	17.7	s
ESR (mm/h)	95	70	76	56	55	
S. ALP (IU/l)	174	106	157	94		
Rheumatoid factor	Negative					
Anti-CCP (IU/ml)	Negative (<25)					
HLA B27	Positive					
ANA			Negative		Negative	
C3, C4					Negative	
PR3-ELISA					Negative	
MPO-ELISA					Negative	
Anti-ds-DNA					Negative	
Creatinine (mg/dl)	0.8	0.78	0.6	9.8	2.6	1.6
Urea (mg/dl)	25		14	154	57	43
Serum albumin (g/dl)	4.2	4.4	3.7	2.2	2.6	2.9
Urine albumin	Nil	Nil	Nil	3+	3+	3+
Urine RBC	Nil	Nil	Nil	Nil	Nil	Nil
Urine WBC	Nil	Nil	Nil	Nil	Nil	Nil
24-h urine protein (g/24 h)					2.6	1.2
			Radiologic investigations			

X ray bilateral hip uniform reduction of bilateral hip joint space with subchondral sclerosis. MRI both hips Reduced joint space along bilateral hip joint with mild join effusion, synovitis, and subchondral marrow edema along bilateral femoral head and acetabulum (L>>R). AKI=acute kidney injury, S.ALP=serum alkaline phosphatase, MPO- myeloperoxidase, ANA=anti-nuclear antigen, Anti-CCP=anti-cyclic citrullinated peptide, anti-ds-DNA=anti-double-stranded DNA antibody, AS=ankylosing spondylitis, C3, C4=complement factor 3, complement factor 4, CRP=C-reactive protein, ELISA=enzyme-linked immunosorbent assay, ESR=erythrocyte sedimentation rate, Hb=hemoglobin, HLA=human leukocyte antigen, RA=renal amyloidosis, RA factor=rheumatoid arthritis factor, RBC=red blood cells, TA=Takayasu arteritis, UC=ulcerative colitis, WBC=white blood cells

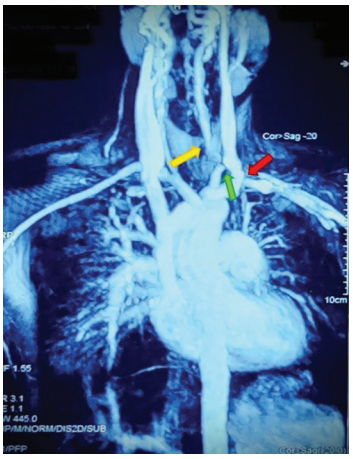

Figure 1: MR angiography of aorta and its branches shows wall thickening with enhancement of AoA, DTA, and upper AA with nearly complete occlusion of left common carotid artery (green arrow) and luminal narrowing of proximal subclavian (red arrow) and vertebral arteries (yellow arrow). AA = abdominal aorta, AoA = aortic arch, DTA = descending thoracic aorta, MR = magnetic resonance.

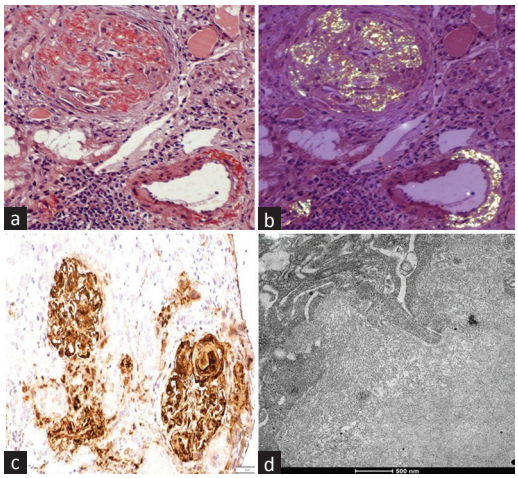

Figure 2: Histopathologic examination of kidney biopsy. (a) Congo red stain 200× showing amyloid deposits in the mesangium and focally in peripheral capillaries. (b) Congo red stain 200× showing apple green birefringence on polarized light. (c) Serum amyloid A (SAA) stain 200× showing positive deposits of amyloid in the mesangium and focally in peripheral capillary walls. (d) Electron microscopy 17,500 × shows basement membrane showing randomly arranged fibrils with a mean cross diameter of 9.97 mm.

aorta (AA), with nearly complete occlusion of left CCA and luminal narrowing of bilateral proximal subclavian (L > R) and vertebral arteries. A diagnosis of TA was made in light of these findings [Table 1] and prednisolone (1 mg/kg/d) was started, which led to a symptomatic improvement. However, a follow-up positron emission tomography (PET) scan at 6 months revealed increased metabolic uptake along the walls of ascending aorta, AoA, DTA, left CCA, and left subclavian arteries, despite therapy. He was advised infliximab, but he could not afford the same and was lost to follow-up.

Six months later, he presented with acute gastroenteritis and oliguria. Investigations showed elevated blood urea, serum creatinine, and albuminuria. With fluids and antibiotics, stool consistency and urine output improved. However, a follow-up after 3 weeks revealed persistent hypoalbuminemia, proteinuria, and increased serum creatinine compared to baseline [Table 1]. A kidney biopsy was done, which was suggestive of AA amyloidosis [Figure 2]. The patient was diagnosed with AS-associated UC and TA with secondary RA. Telmisartan and dapagliflozin were started, after which proteinuria improved. The necessity of biologic therapy was discussed with the patient, with infliximab planned once his financial constraints are resolved.

Discussion

AS often exhibits extra-articular manifestations. Cardiac manifestations include ascending aortitis, valvular regurgitation, and cardiac conduction abnormalities, but association with TA is rare.[1] Our case showed arteritis involving the carotid arteries, DTA, and AoA, suggesting TA rather than spondyloarthropathy-related aortitis. The largest case series of TA with spondyloarthropathies (n = 14) primarily describes middle-aged women with a high rate of HLA B27 negativity. In most cases, spondyloarthropathy precedes TA, with a median time gap of 4.5 years between the two conditions.[1] However, our patient was a young male, was HLA B27 positive, and had a short duration of 2 years between diagnosis of AS and TA (despite having previous exposure to steroids, which could have delayed the symptomatology of TA).

Renal involvement is often observed in both AS and TA. In AS, the most common cause of renal involvement is IgA nephropathy.[2] The incidence of RA in AS has been reported as 6.1%.[3] On the other hand, TA primarily involves renal vessels with resulting renovascular hypertension and ischemic nephropathy. RA is rare in TA.[4,5] A single retrospective cohort from Russia reports amyloidosis in TA with an incidence of 3.9%, speculating genetic susceptibility versus other ethnicities.[6]

Our case presented with amyloidosis 6 years after AS diagnosis and 2 years after TA diagnosis. Previous studies have shown a median time gap of 12 years between AS and RA[3] and 13 years between TA and RA.[6] The coexistence of multiple inflammatory disorders likely contributed to accelerated development of secondary RA in this case. While infliximab was recommended, financial constraints prevented its initiation. Currently, the patient receives the best possible supportive care and is under monitoring to assess the response to therapy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Conflicts of interest

There are no conflicts of interest.

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References

1. Rivière E, Arnaud L, Ebbo M, Allanore Y, Claudepierre P, Dernis E, *et al*. Takayasu arteritis and spondyloarthritis: Coincidence or association? A study of 14 cases. J Rheumatol 2017;44:1011-7.
2. He D, Wang R, Liang S, Liang D, Xu F, Zeng C, *et al*. Spectrums and prognosis of kidney disease in patients with ankylosing spondylitis. Kidney Dis (Basel) 2020;6:444-52.
3. Barbouch S, Hajji M, Jaziri F, Aoudia R, Fellah E, Hedri H, *et al*. Renal amyloidosis in ankylosing spondylitis: A monocentric study and review of literature. Saudi J Kidney Dis Transpl 2018;29:386-91.
4. Wada Y, Nishida H, Kohno K, Tamai O, Fujisawa M, Katoh S, *et al*. AA amyloidosis in Takayasu's arteritis--long-term survival on maintenance haemodialysis. Nephrol Dial Transplant 1999;14:2478-81.
5. Kos I, Stilgenbauer S, Bewarder M. Renal AA Amyloidosis leading to early diagnosis and treatment of takayasu arteritis: A case report and review of the literature. Clin Res Cardiol 2020;109:1438-41.
6. Mukhin N, Smitienko I, Novikov P, Moiseev S, Shevtsova T. AA Amyloidosis in a cohort of 128 patients with Takayasu's arteritis. J Vasc 2017;3:120.

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