Case Report:

Coexisting Rheumatoid Arthritis and Takayasu's Arteritis: A Case Report

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Abstract:

The coexistence of Rheumatoid Arthritis (RA) and Takayasu's Arteritis (TA) is a rare combination and described only in case reports in the literature. Although concurrent presence of RA and TA has been described only in a few literatures to date and the number of reports is increasing, the association between RA and TA remains to be clarified. We present a case of a female patient with both RA and TA, presenting with polyarthritis.

Introduction:

Takayasu's arteritis (TA) is a rare, idiopathic, chronic inflammatory disease characterized by necrotizing and obliterative segmental pan arteritis of unknown aetiology, which attacks the aorta and its major branches [(1)]. Autoimmunity is thought to play an important role, and several cases have been reported an association between TA and various autoimmune disorders. Rheumatoid arthritis (RA) is a chronic inflammatory disease characterized by symmetric, multiple arthritis and extra-articular manifestations (EAM), including the skin, eyes, lungs and blood vessels. Rheumatoid vasculitis is an unusual complication of longstanding, severe RA, which involves the small and medium sized arteries in the body [(2)]. The controversy exists about the concurrent presence of both diseases, and it is also unclear whether this disease occurs incidentally or overlaps like other connective tissue diseases. However, the reports of coexistence of RA and TA are increasing in spite of very rare description only in case reports in the literature [(4-14)]. More reports, experience and case control studies will be needed to define the relationship between RA and TA. We here described a case of a young woman with both RA and TA and reviewed the clinical features of the reported cases with coexistence of RA and TA.

Case report:

A 51-year-old female patient presented with joint pain, breathlessness, easy fatiguability since 6 months. Joint pain involving 4-10 small joints, which is associated with morning stiffness of more than 1 hour. Breathlessness since 6 months which progressed from grade 3 to grade 4 NYHA. No significant past history, family history, personal history.

On examination she was found to have feeble pulses in both upper limbs, left > right (axillary, brachial & radial), well felt in the lower limbs. She has reverse arm- leg systolic blood pressure discrepancy of 50 mmhg (84/60 mm hg in right arm, 80/58mm Hg in left arm,140/80 in right leg,140/80 in left leg). There was bruit over right subclavian artery. On examination, there was palpable purpura over bilateral medial malleolus & swan neck deformity of index finger of left hand. Cardiovascular examination showed features suggestive of severe

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pulmonary arterial hypertension with tricuspid regurgitation. Respiratory system, Central nervous system & per abdomen examination was normal.

On admission, laboratory test revealed white blood cell (WBC) of 9400 cells/cumm with neutrophilia and haemoglobin 9.9 g/dL, platelet 299000/mL, erythrocyte sedimentation rate (ESR) 120 mm/hr, C-reactive protein (CRP) 56 mg/L. Rheumatoid factor (RF) level was 71.81 IU/mL (normal ＜14 IU/mL) and anti-CCP antibody level was 4.6 IU/mL (normal ＜5 IU/mL).

EULAR CRITERIA:

1) **JOINT DISTRIBUTION:** 4 – 10 Small joints - 3 points

2) **SEROLOGY:** High positive RF- 3 points

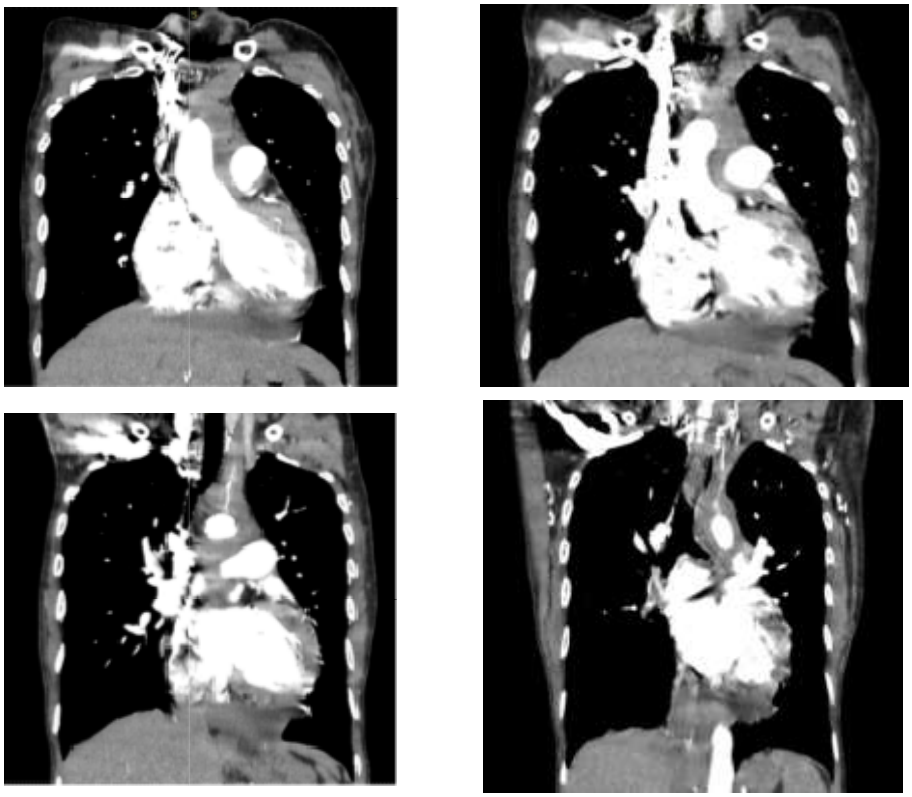
3) **SYMPTOM DURATION:** 6 Months - 1 point

4) **ACUTE PHASE REACTANTS:** Both CRP and ESR are high -1 point

According to the criteria, the total points are **8** which is highly suggestive of Rheumatoid Arthritis.

HRCT and CT Angio Thorax:

- Diffuse wall thickening of aorta and its branches, right and left pulmonary arteritis are suggestive of **Takayasu Arteritis** (type II a).



Discussion:

Takayasu's arteritis (TA) is a nonspecific large vasculitis, which induces occlusive or dilative lesions in the aorta, its main vessels, and pulmonary or coronary arteries [(1)]. TA most frequently affects young Asian woman, with an age at onset usually between 10 and 40 years and median age at diagnosis of 28 years. The pathogenesis

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remains to be clarified but has been suggested that the vasculitis may deteriorate via autoimmune mechanism with infiltration of cytotoxic T lymphocyte and release of proinflammatory cytokines [(2)]. This is the reason that TA is a rare pathology associated with other autoimmune disease, such as systemic lupus erythematosus and systemic sclerosis [(2)]. Despite a similar pathogenesis with a prominent role of TNF, the association between RA and TA is rare [(3)]. The relationship between TA and RA remains unknown. Despite its predominant articular manifestations, RA is a systemic disease often associated with organ-specific Extra articular manifestations [(2)]. Rheumatoid vasculitis affects from ＜1% to 5% of RA patients and can occur in any organ, although about 90% of cases have cutaneous lesions [(2)]. Commonly inflammation of the small and medium sized arteritis in the extremities may be seen, but aortic root changes and aortitis were reported in RA. RA associated aortitis is almost accompanied by severe extra-articular vasculitic manifestations with positive rheumatoid factor (RF) and subcutaneous nodules [(8)]. Although aortitis is rarely reported in patients with RA, necropsy data suggested that may be seen in up to 15% of cases [(8)]. In the previous reports of this disease, age at diagnosis of RA or TA, mucocutaneous, articular manifestations and rheumatoid factor were described (Table 1). In addition, results of genotype of human lymphocyte antigen (HLA) and type of involvement of large blood vessels in TA were described [(4-14)]. According to the research on TA in 129 Korean patients, the most common clinical symptom of TA was headache (60%) and the second most common symptom is exertional dyspnoea (42%) [(15)]. But in this case, patient present with nonspecific respiratory symptoms. The concurrent presence of Takayasu arteritis and rheumatoid arthritis (RA) is described in only few cases in the literature to date, these were often in female (18 females and 6 males), and the mean age at the time of RA diagnosis was 46.5±14.3 years (range 16～74). The mean age at the time of TA diagnosis was 52.1±15.7 years (range 20～82), and the age at diagnosis of TA in patients with RA have older than patients with isolated TA. Only 3 studies have reported case developed TA before RA [(11,13)], TA diagnosed after arthritis signs and symptoms in most cases. The average time elapsing between the onset of RA and the development of TA was 5.6 year. Most of the patients had positive RF and subcutaneous nodules were detected. These studies found that clinical difference regarding arthritis and arthralgias in the course of TA may be attributable to a genetic background, as polymorphic HLA genes and their combinations may have had a role in modulating the clinical findings. Despite the fact that there is still a debate on the exact nature of the coexistence of RA with TA, we suggest that it can be a very rare rheumatic disease entity. Clinical characteristics of our case and previous case reports of coexisting RA and TA (Table 1)[9]

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Table 1. Clinical characteristics of our case and previous case reports of coexisting RA and TA

References	Age/Sex	Age at diagnosis (years)		RF	Nodule	Erosion	HLA types	Involvement type	
		RA	TA					Angiography	Necropsy
Sanding et al. (5)	63/F	40	63	+	NR	NR	NR	I	NR
Sanding et al. (5)	65/F	55	65	+	+	NR	NR	I	NR
Falicov et al. (6)	16/F	16	20	NR		+	NR	I	NR
Reimer et al. (7)	49/F	44	49	+	+	NR	NR	NR	III
Rush et al. (8)	37/F	37	26	+	+	+	DR4	III	NR
Sketchler et al. (9)	53/F	50	53	NR	NR	NR	NR	I	NR
Mimura et al. (10)	50/F	48	50	+	NR	NR	NR	NR	III
Gravellese et al. (11)	61/F	50	61	+	+	NR	NR	NR	I
Gravellese et al. (11)	82/F	74	82	NR	NR	NR	NR	NR	I
Gravellese et al. (11)	68/M	47	68	+	+	NR	NR	NR	III
Gravellese et al. (11)	61/F	58	61	+	+	NR	NR	NR	I
Gravellese et al. (11)	52/F	26	52	+	+	NR	NR	NR	I
Gravellese et al. (11)	69/M	68	69	+	+	NR	NR	NR	Aortic root
Gravellese et al. (11)	60/M	48	60	+	+	NR	NR	NR	NR
Gravellese et al. (11)	46/M	37	46	+	+	NR	NR	NR	II
Gravellese et al. (11)	67/M	65	67	+	+	NR	NR	NR	III
Gravellese et al. (11)	64/F	61	64	+	+	NR	NR	NR	III
Towned et al. (12)	44/M	NR	44	+	+	+	NR	Aortic root	NR
Nakabayashi et al. (13)	64/F	60	64	+	NR	+	DR2, 12	III	NR
Jung et al. (14)	30/F	28	24	+	+	+	NR	NR	NR
Korkmaz et al. (15)	36/F	34	36	+	NR	+	DR4, 1	I	NR
Yokoe et al. (16)	50/F	46	39	+	NR	NR	NR	I	NR
Verweij et al. (17)	49/F	48	49	-	NR	NR	NR	I	NR

RA-Rheumatoid arthritis TA-Takayasu Arteritis RF- Rheumatoid Factor HLA-Human leucocyte antigen NR-not reported

Of these two diseases in our patient or other previously reported cases might be a complication of the chronic autoimmune inflammatory disease and a nonspecific vascular response to a generalized inflammatory process in association with genetic background predisposing to both RA and TA. The genetic relationship between RA and TA is not yet clearly established. however, it has been shown that a considerable number of reported cases. Thus, physicians should consider that RA patients with extra-articular manifestations should be monitored carefully for the development of an occlusive arterial involvement such as TA. Although coexistence of RA and TA were very rarely reported, the inter-relationship between these diseases as an EAM has controversies. The clinical characteristics of coexisting RA and TA were not well known. We found 23 other cases of RA overlapped with TA or aortitis in the English literature [(4-14)].

We report the case of an Asian woman who was diagnosed with Rheumatoid arthritis and Takayasu's arteritis. As mentioned above, there is still a debate on the coexistence of RA with TA, it can be a very rare rheumatic disease entity. Physicians should consider that RA patients with extra-articular manifestations should be monitored carefully for the development of an occlusive arterial involvement such as TA.

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