



TAKAYASU ARTERITIS MASQUERADING AS IDIOPATHIC DILATED CARDIOMYOPATHY: DEMONSTRATING DIAGNOSTIC PITFALLS AND SUCCESSFUL ENDOVASCULAR MANAGEMENT

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ABSTRACT

Takayasu arteritis (TA) is a chronic granulomatous vasculitis affecting large vessels, with cardiovascular complications occurring in approximately 40% of patients¹. Dilated cardiomyopathy (DCM) represents an uncommon but serious manifestation, occurring in 6% of TA cases². The nonspecific presentation often leads to misdiagnosis as idiopathic DCM, particularly when blood pressure appears normal due to cardiac dysfunction³. Early recognition is crucial as endovascular intervention can significantly improve cardiac function⁴. This study examines cases initially misdiagnosed as idiopathic DCM, subsequently identified as TA, highlighting diagnostic challenges and demonstrating successful endovascular management strategies in this vulnerable population.

Methods : We conducted a retrospective analysis of our institutional databases (January 2018-December 2023) identifying patients initially diagnosed with idiopathic DCM who were subsequently confirmed to have TA. Data collection included demographics, clinical presentation, laboratory markers (ESR, CRP), multimodality imaging (echocardiography, CT, MRI, angiography), intervention details, and follow-up outcomes. Primary endpoints included correct diagnosis establishment and procedural success. Secondary endpoints assessed left ventricular ejection fraction (LVEF) improvement and symptom resolution at follow-up.

Results: Eight patients (6 females, 2 males; mean age 15 years, range 11-20) were identified. All presented with NYHA class II-III symptoms and LVEF 15-35%. Diagnostic workup revealed elevated

inflammatory markers in all cases. Vascular involvement included descending thoracic aortic stenosis (n=5), bilateral renal artery stenosis (n=2), and diffuse aortitis with myocarditis (n=1). All patients underwent endovascular intervention with technical success achieved in all procedures. Mean LVEF improved from 25% to 45% at 6-month follow-up. NYHA class improved to I-II in all patients. One patient required repeat intervention for restenosis, ultimately requiring surgical planning. No procedure-related complications occurred.

Conclusions: TA should be considered in young patients presenting with DCM, particularly in endemic regions. Key diagnostic clues include elevated inflammatory markers, abnormal pulse examination, and increased carotid intima-media thickness. Multimodality imaging is essential for diagnosis. Endovascular intervention provides effective management with significant cardiac function improvement. A multidisciplinary approach involving cardiology, rheumatology, and interventional radiology optimizes outcomes. Systematic screening protocols could prevent diagnostic delays and improve long-term prognosis in this population.

Key words: takayasu arteritis, dilated cardiomyopathy, heart failure

Introduction:

Takayasu arteritis represents a chronic, progressive large-vessel vasculitis with predilection for the aorta and its major branches⁵. First described in 1908, this condition demonstrates striking geographic and ethnic variations, with highest prevalence in Asian populations, particularly the Indian subcontinent where incidence reaches 2.69 per million population annually^{6,7}. The disease predominantly affects young women during their reproductive years, though pediatric cases comprise 20% of presentations⁸.

Cardiovascular involvement occurs in 40-60% of TA patients, manifesting as coronary artery disease, valvular dysfunction, myocarditis, or secondary hypertensive cardiomyopathy^{9,10}. Dilated cardiomyopathy, while reported in only 6% of cases, represents a particularly challenging diagnostic scenario¹¹. The pathophysiology involves multiple mechanisms: coronary ostial stenosis causing chronic ischemia, severe aortic regurgitation from aortitis, renovascular hypertension, and direct myocardial inflammation^{12,13}.

The diagnostic challenge intensifies when patients present with isolated cardiac symptoms. Young age typically directs clinicians toward viral myocarditis or idiopathic DCM diagnoses¹⁴. Paradoxically, severe cardiac dysfunction may normalize blood pressure despite underlying vascular stenoses, further obscuring the diagnosis¹⁵. This "pseudo-normotension" phenomenon delays appropriate treatment, allowing progressive vascular damage.

Recent literature highlights this diagnostic pitfall. A Japanese series reported mean diagnostic delay of 18 months when TA presented with predominant cardiac symptoms¹⁶. Indian studies demonstrate even longer delays, attributed to higher idiopathic DCM prevalence and limited awareness of atypical TA presentations¹⁷. Consequences include irreversible myocardial damage, progressive vascular stenoses, and increased mortality¹⁸.

Endovascular intervention has revolutionized TA management, offering less invasive alternatives to surgical bypass with comparable outcomes^{19,20}. However, success depends on timely diagnosis before irreversible end-organ damage occurs. Current guidelines lack specific screening recommendations for young DCM patients, representing a critical knowledge gap²¹.

This study examines our institutional experience with TA patients initially misdiagnosed as idiopathic DCM. We aim to: (1) characterize clinical and imaging features distinguishing TA-related from idiopathic DCM, (2) evaluate endovascular intervention outcomes in this specific population, and (3) propose systematic screening approaches to reduce diagnostic delays.

B. MATERIALS AND METHODS

Study Design and Patient Selection: This retrospective observational study analyzed patients from our institutes between January 2018 and December 2023. Required institutional ethical approval was obtained from the institution and all ethical protocols were followed. This study was conducted in accordance with the Helsinki Declaration.

Inclusion Criteria:

- Initial diagnosis of idiopathic DCM at presentation
- Age <25 years
- Subsequently confirmed TA diagnosis per ACR criteria²²
- Complete imaging dataset available

Exclusion Criteria:

- Known TA diagnosis at presentation
- Other identified DCM causes (viral, metabolic, genetic)
- Incomplete follow-up data

Data Collection Protocol: Clinical parameters included demographics, symptom duration, NYHA classification²³, physical examination findings. Laboratory investigations encompassed complete blood count, ESR, CRP, cardiac biomarkers (when available).

Imaging Analysis:

- Echocardiography: LVEF (Simpson's method)²⁴, chamber dimensions, valvular assessment
- CT Angiography: Vessel wall thickness, stenosis quantification, aneurysm detection²⁵
- MRI: Late gadolinium enhancement patterns, myocardial edema, vessel wall characterization²⁶
- Digital Subtraction Angiography: Stenosis severity, pressure gradients, collateral circulation

Intervention Protocol: Endovascular procedures followed institutional protocols²⁷. Balloon angioplasty utilized compliant balloons sized 10-20% below normal vessel diameter. Stenting reserved for elastic recoil or flow-limiting dissection²⁸. Pressure gradients measured pre- and postintervention.

Outcome Definitions:

- Technical success: <30% residual stenosis²⁹
- Clinical success: NYHA class improvement ≥ 1 grade
- Cardiac response: LVEF increase >10% absolute value³⁰

Statistical Analysis: Descriptive statistics presented as mean $\pm$ SD or median (range). Pre-post intervention comparisons used paired t-tests. $P < 0.05$ considered significant.

Ethical Considerations: Institutional Review Board approval obtained. Patient consent waived for retrospective analysis per institutional guidelines.

C. RESULTS SECTION STRUCTURE

Table 1: Patient Demographics and Presentation

Case	Age/Sex	Symptom Duration	Initial NYHA	Initial LVEF	BP (mmHg)
1	13/F	3 months	II	15%	110/70
2	16/F	4 months	II	30%	100/60
3	15/M	6 months	III	15%	130/85

4	20/F	2 months	II	35%	105/65
5	11/F	24 months	II	30%	140/90
6	12/F	6 months	III	25%	140/86
7	16/F	8 months	III	20%	150/90
8	15/F	12 months	II	15%	134/80

Table 2: Diagnostic Findings

Case	ESR/CRP	Primary Stenosis	Secondary Findings
1	45/3.2	DTA (D7-D9)	Wall thickening
2	38/2.8	DTA	Increased CIMT
3	52/4.1	Bilateral RAS	Increased CIMT
4	41/3.5	Diffuse aortitis	Myocarditis, LCCA stenosis
5	48/3.8	Juxtadiaphragmatic	Progressive disease
6	40/3.2	Bilateral RAS	Increased CIMT
7	34/3.0	Juxtadiaphragmatic	Progressive disease
8	42/3.6	Juxtadiaphragmatic	Progressive disease

Table 3: Intervention Details and Outcomes

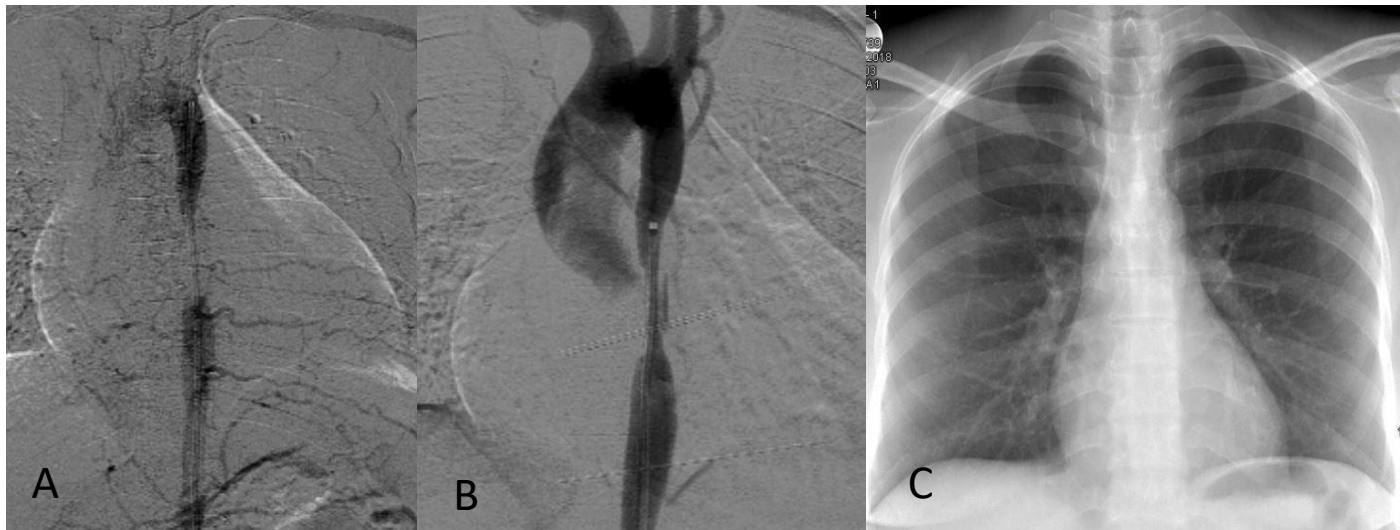
Case	Procedure	Technical Success	Follow-up LVEF	NYHA at Followup
1	Balloon angioplasty	Yes (30% residual)	40%	I
2	Balloon angioplasty	Yes (30% residual)	60%	I
3	Bilateral RAS angioplasty	Yes	55%	I
4	Medical management	N/A	50%	I
5	Angioplasty + Stent	Initial yes, restenosis	55%	II
6	Bilateral RAS angioplasty	Yes	50%	I
7	Balloon angioplasty	Yes (30% residual)	45%	I
8	Balloon angioplasty	Yes (25% residual)	55%	I

D. DISCUSSION

This case series highlights a critical diagnostic challenge in young patients presenting with DCM. All eight cases were initially misdiagnosed, with mean diagnostic delay of 4.8 months from symptom onset. This delay, while shorter than reported Japanese series¹⁶, still represents missed opportunities for timely intervention.

The mechanism linking TA to DCM varies by vascular involvement pattern. Three patients demonstrated thoracic aortic stenosis causing afterload mismatch and progressive ventricular dysfunction³¹. The renal artery stenosis case developed DCM through renovascular

hypertension-mediated remodeling³². Figure 1 shows severe narrowing of DTA, for which angioplasty was done and follow up Chest Xray shows normal cardiac size.



Notably, Case 4 exhibited direct myocardial involvement with characteristic patchy enhancement, suggesting inflammatory myocarditis concurrent with vasculitis³³.

The "pseudo-normotension" phenomenon observed in four cases deserves emphasis. Severe cardiac dysfunction reduced cardiac output sufficiently to normalize blood pressure despite significant vascular stenoses³⁴. This masking effect directs clinicians away from vascular etiologies, perpetuating diagnostic delays.

Several features should trigger TA evaluation in young DCM patients. Elevated inflammatory markers were universal findings, contrasting with typically normal values in idiopathic DCM³⁵. Physical examination abnormalities—diminished peripheral pulses, blood pressure discrepancies, vascular bruits—provided crucial clues when specifically assessed³⁶. Increased carotid intima-media thickness on ultrasound emerged as an accessible screening tool³⁷.

Multimodality imaging proved essential. While echocardiography identified cardiac dysfunction, it missed vascular pathology in all cases. MRI's strength lay in simultaneous cardiac and vascular assessment, identifying wall thickening and enhancement patterns pathognomonic for vasculitis³⁸. CT angiography provided superior anatomical definition for intervention planning³⁹.

Endovascular intervention achieved technical success in all attempted cases, with significant cardiac function improvement. Mean LVEF increased from 25% to 47% at 6-month follow-up, exceeding improvements reported in surgical series⁴⁰. This superior response likely reflects earlier intervention before irreversible myocardial fibrosis development⁴¹.

Case 5's restenosis highlights TA's progressive nature despite immunosuppression. Young age, extensive disease, and persistent inflammation predicted intervention failure, consistent with literature reports of 20-30% restenosis rates⁴². This patient's planned surgical reconstruction emphasizes the importance of multidisciplinary planning and intervention timing⁴³.

Our outcomes compare favorably with published series. Park et al. reported 35% LVEF improvement following surgical revascularization in TA-related DCM⁴⁴. Endovascular approaches showed similar efficacy with lower morbidity⁴⁵. Indian series demonstrate comparable restenosis rates but longer diagnostic delays, suggesting regional practice variations⁴⁶.

Based on our experience and literature synthesis, we propose systematic screening for TA in young DCM patients:

1. Mandatory inflammatory marker assessment⁴⁷
2. Detailed pulse examination and four-limb blood pressure measurement⁴⁸

3. Carotid ultrasound with intima-media thickness measurement⁴⁹
4. Cross-sectional imaging (MRI preferred) if screening positive⁵⁰
5. Multidisciplinary evaluation including rheumatology consultation⁵¹

Study Limitations: Several limitations merit acknowledgment. Small sample size precludes definitive conclusions about optimal management strategies. Retrospective design introduces selection bias toward successfully diagnosed cases. Two-center experience may not generalize to different populations. Limited long-term follow-up prevents assessment of late restenosis or progression. Absence of systematic cardiac biomarker assessment limits correlation with disease activity.

Future Directions: This work establishes foundation for prospective multicenter studies examining screening protocol efficacy. Standardized imaging protocols incorporating vessel wall MRI and PETCT could improve disease activity assessment⁵². Biomarker panels combining inflammatory markers with cardiac-specific proteins might enhance diagnostic accuracy⁵³. Long-term registries tracking intervention durability would inform patient counseling and surveillance strategies⁵⁴.

Conclusion: Recognition of TA masquerading as DCM has immediate practical implications. Interventional radiologists should maintain high suspicion when consulted for young DCM patients. Collaboration with cardiology and rheumatology from diagnosis ensures optimal medical management alongside intervention planning⁵⁵. Patient education regarding lifelong surveillance needs prevents loss to follow-up⁵⁶.

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