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Rare Systemic Vasculitides: Polyarteritis Nodosa and Takayasu Arteritis

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Case series
Patients: Female, 20-year-old • Female, 38-year-old
Final Diagnosis: Takayasu arteritis • vasculitis
Symptoms: Extensive involvement of the aorta and its branches • treatment-resistant hypertension • significant vascular symptoms
Clinical Procedure: —
Specialty: General and Internal Medicine • Radiology • Rheumatology

Objective: Rare disease

Background: Polyarteritis nodosa (PAN) and Takayasu arteritis (TA) are systemic medium- and large-vessel vasculitides associated with significant morbidity when diagnosis is delayed. Although described in the literature, reports directly demonstrating the impact of diagnostic timing on vascular outcomes remain limited. This report presents 2 rare and contrasting cases highlighting the importance of early recognition with timely vascular and immunosuppressive interventions, which can prevent irreversible ischemic complications, whereas delayed diagnosis can result in permanent structural damage, reinforcing the need for accurate differentiation between PAN and TA.

Case Reports: Case 1 was a young woman with refractory hypertension and significant vascular manifestations, in whom PAN was diagnosed through combined analysis of symptoms, laboratory tests, and angiographic findings. Early angioplasty and treatment with glucocorticoids and azathioprine resulted in a favorable response. Case 2 was a woman with TA who had extensive involvement of the aorta and its branches with irreversible sequelae, including aortic valve replacement. Diagnostic delay led to progression. Glucocorticoids and adalimumab achieved clinical stabilization without reversing established structural damage.

Conclusions: The presented cases reinforce the importance of early identification and appropriate differentiation of systemic vasculitides to prevent irreversible vascular lesions. This study compares a case of PAN diagnosed early, with vascular intervention preventing ischemic complications, and a case of TA diagnosed late, with permanent structural damage despite treatment. Early use of vascular imaging combined with prompt immunosuppressive therapy and multidisciplinary management contributes to a favorable prognosis, highlighting the need for greater clinical awareness and careful diagnostic strategies in rare vasculitides.

Keywords: Takayasu Arteritis • Polyarteritis Nodosa • Vascular Diseases • Early Diagnosis

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/951271

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Introduction

Primary vasculitis of medium- and large-vessel caliber, such as polyarteritis nodosa (PAN) and Takayasu arteritis (TA), represents significant clinical challenges due to their potentially severe course, heterogeneous presentation, and diagnostic difficulty. Both diseases are rare, with an origin that is still unclear, and are characterized by progressive vascular inflammation that can lead to stenosis, thrombosis, or aneurysm formation, resulting in multisystem involvement.

PAN is a necrotizing vasculitis of medium-sized arteries, with an estimated prevalence of 31 cases per million inhabitants. The cutaneous form accounts for about 4% of cases and shows a female predominance [1]. Clinical manifestations include nonspecific systemic symptoms such as fever, weight loss, hypertension, and neurological, cutaneous, and renal involvement, which often delays diagnosis. Despite therapeutic advances, morbidity and mortality remain significant, especially when the disease is recognized late.

TA is a chronic granulomatous vasculitis that affects the aorta and its major branches, causing transmural inflammation capable of generating stenosis, occlusions, and aneurysms. Diagnosis relies heavily on imaging methods, as early signs and symptoms are nonspecific. Silent progression and extensive arterial involvement make management particularly challenging.

Given the diagnostic and therapeutic complexity presented by both conditions, whether PAN or AT, case reports continue to play an important role in describing unusual, simultaneous, overlapping, or atypical presentations. Early identification, combined with a multidisciplinary approach, can reduce complications, reinforcing the importance of studying such cases.

Here, we report the case of a patient with PAN who achieved therapeutic success with endovascular therapy combined with immunosuppression, and the case of a patient with TA presenting extensive vascular involvement extending from the aortic valve to the abdominal aorta and its branches.

It is essential to emphasize that both patients had rare diseases with high severity indexes. However, Case 1 received an early diagnosis, which allowed for rapid intervention during disease recurrence. As soon as symptoms began, clinical suspicion was raised, followed by imaging tests (abdominal CT angiography), identification of stenosis, and subsequent angioplasty, thus preventing a severe case of mesenteric ischemia, which is the main cause of mortality in this select group. Case 2 received a late diagnosis, which progressed to irreversible vascular sequelae. This clearly highlights the difference between the outcome of early clinical diagnosis and that of late intervention.

Understanding these 2 differing scenarios may guide strategies and recommendations, with appropriate use of imaging tests, multidisciplinary management, and more effective protocols. In addition, we aim to expand opportunities for future research focused on identifying early markers of severity, improving imaging techniques, and developing more personalized therapeutic protocols.

This case study involves 2 rare systemic vasculitides—PAN and TA—and discusses their main clinical and pathophysiological manifestations and therapeutic strategies. It also highlights challenges encountered in diagnosis and management of these diseases, emphasizing the importance of early diagnosis for determining the most appropriate and effective treatment options.

Furthermore, this study aims to contribute to the dissemination of knowledge about PAN and TA, since these are rare conditions that require differentiated medical approaches, with treatment guided according to severity of involvement and associated symptoms. We emphasize that the combination of early endovascular therapy with immunosuppressive therapy may improve clinical outcomes compared with medical therapy alone.

Case Reports

Case 1

A 20-year-old woman with diabetes mellitus (DM) was seen at the rheumatology outpatient clinic of a tertiary hospital in the interior of Rio Grande do Sul state. In 2020, she developed systemic arterial hypertension that was refractory to treatment. She reported a previous diagnosis of systemic arterial hypertension that had been refractory to treatment. In 2020, she underwent computed tomography (CT) angiography, which showed 50% celiac trunk stenosis, greater than 90% left renal artery stenosis, and more than 50% superior mesenteric artery stenosis at its origin. In 2021, she underwent angioplasty for left renal artery stenosis, with resolution of the condition at that time.

In 2024, she presented with progressive weight loss associated with diffuse postprandial abdominal pain. Physical examination results were normal. Laboratory tests were performed for the differential diagnosis of systemic vasculitides, showing nonreactive antinuclear antibody (ANA), nonreactive antineutrophil cytoplasmic antibody (ANCA), negative hepatitis B serology, and normal inflammatory markers. A new follow-up CT angiography showed greater than 75% stenosis of the superior mesenteric artery and perivascular soft-tissue attenuation in this region, possibly corresponding to an inflammatory process (vasculitis).

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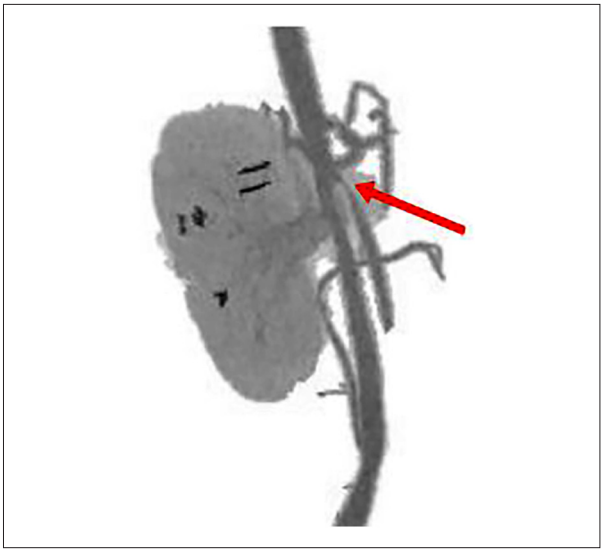

Figure 1. Abdominal angiotomography, Case 1. Demonstrating 75% occlusion of the superior mesenteric artery (arrow).

After anamnesis, physical examination, and evaluation of imaging studies, she was diagnosed with PAN, fulfilling the European Alliance of Associations for Rheumatology (EULAR) classification criteria: weight loss >4 kg, recent-onset hypertension, and characteristic arteriographic imaging.

Due to the unavailability of cyclophosphamide at the time, she promptly underwent emergency angioplasty of the superior mesenteric artery combined with treatment using prednisone 0.5 mg/kg/day and azathioprine 2 mg/kg/day, with good clinical evolution even after corticosteroid tapering. She is currently in disease remission, with adequate blood pressure control, weight stabilization, and reduction of inflammatory markers.

Case 2

A 38-year-old woman presented to the rheumatology service of a tertiary hospital in the interior of Rio Grande do Sul state, with a previous diagnosis of TA made by a rheumatologist in her hometown in 2019. Following the classification criteria of the American College of Rheumatology (ACR) – EULAR, she presented with: age <60 years, evidence of vasculitis on CT angiography, female sex, reduced pulse in the upper extremities, abnormal bilateral carotid arterial flow (bruit), blood pressure difference between upper limbs ≥20 mmHg, involvement of 3 or more arterial territories, symmetric arterial involvement, and involvement of the abdominal aorta with renal and mesenteric artery involvement. She has a history of aortic valve replacement in 2022 due to aortic insufficiency, probably secondary to her underlying disease.

On physical examination, she had unreadably low blood pressure in the left upper limb, diffuse systolic murmur in the precordium, more audible in the aortic focus, vascular murmur in the carotid and subclavian arteries (weaker in the left carotid), absent left radial pulse, filiform right radial pulse, abdominal aortic murmur, and filiform left peripheral pedicle pulse.

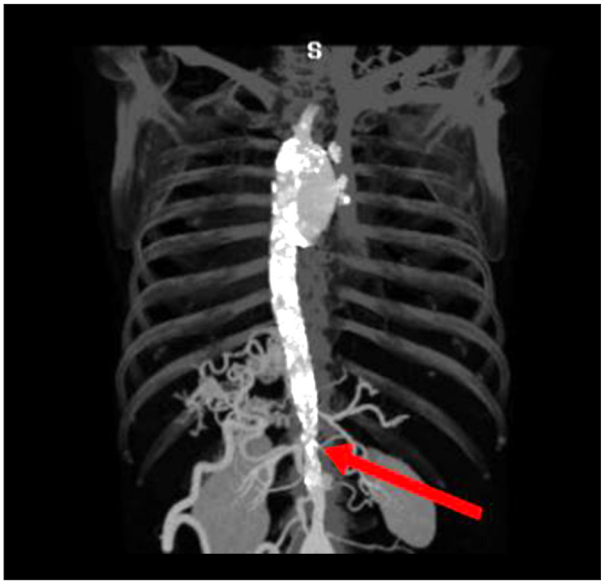

Figure 2. Abdominal angiotomography, Case 2. Reduced caliber of the abdominal aorta in the infrarenal segment (arrow).

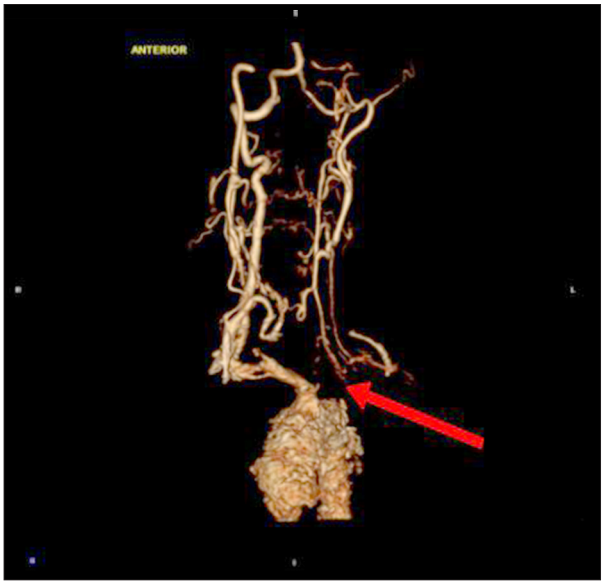

Figure 3. Cervical angiotomography, Case 2. Absence of contrast in the proximal portion of the left subclavian artery, indicating occlusion of this vessel (arrow). Distal filling is observed from the origin of the left vertebral artery.

Previous cervical angiotomography showed significant stenosis of the left subclavian artery (greater than 70%), sub-occlusive stenosis in the brachiocephalic trunk extending to the right

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common carotid artery, and occlusion in the left common carotid artery and the left internal carotid artery. Angiotomography of the thoracic, abdominal, and iliac aorta showed the thoracic aorta had diffuse parietal thickening measuring 6.0 mm (the previous study showed 8 mm) in the ascending segment. There was occlusion of the proximal portion of the left subclavian artery and the proximal portion of the left common carotid artery. The pulmonary artery trunk had an increased caliber measuring 3.6 cm, suggesting a degree of pulmonary hypertension and increased cardiac area. The abdominal aorta had diffuse parietal thickening measuring 3.5 mm, and there were areas of diffuse reduction in the caliber of the abdominal aorta, especially in the infrarenal segment (less than 50%) of its lumen. In addition, there was reduced caliber in the celiac trunk (75% to 90%), the origin of the superior mesenteric artery not identified and was possibly occluded, we found parietal thickening at the origin of the renal arteries with reduction in caliber of 50% to 75%, and the inferior mesenteric artery was significantly reduced in caliber (75% to 90%.

The patient had previously been treated with methotrexate and azathioprine, with lack of therapeutic response. According to the American College of Rheumatology (ACR) vasculitis recommendations, treatment was initiated with prednisone 0.5 mg/kg with gradual tapering and adalimumab 40 mg every 14 days. The choice of the biologic agent adalimumab was justified by her preserved cardiac function. She showed a good response and achieved complete asymptomatic clinical remission [2].

Figures 1 and 2 show the impact of vasculitides, especially in the aortic segment and its primary branches. **Figure 3** shows marked stenosis of the subclavian artery. Currently, the patient is undergoing corticosteroid tapering, with good tolerance to azathioprine and is clinically stable.

Discussion

Arterial vasculitides such as polyarteritis nodosa (PAN) and Takayasu arteritis (TA) pose a significant challenge in clinical practice, both for diagnosis and therapeutic decision-making, particularly because of their often-silent presentation and wide spectrum of manifestations. The 2 cases presented here illustrate how diagnostic timing and initial vascular inflammatory burden directly influence therapeutic decisions and clinical outcomes, in accordance with current international guidelines [3,4].

In Case 1 the diagnosis of PAN was supported by characteristic angiographic findings, including arterial irregularities and vascular involvement associated with refractory hypertension. This method remains a valid diagnostic tool, particularly when combined with compatible clinical manifestations [3,5]. The severity of hypertension and the imminent risk of ischemic complications justified emergency angioplasty aimed at restoring blood flow and preventing permanent damage. After intervention and treatment with glucocorticoids combined with azathioprine, marked improvement was observed in blood pressure control, reduction of systemic inflammatory markers, and stabilization of vascular abnormalities on follow-up imaging studies, confirming a favorable therapeutic response.

The choice of azathioprine was based on the patient's clinical profile and current recommendations, which indicate lower levels of toxic immunosuppressants in cases without visceral involvement posing an immediate risk to life or severe organ failure [2]. This approach is associated with lower risk of cumulative adverse effects while preserving efficacy in controlling inflammation and disease progression [6]. Furthermore, emerging evidence suggests that biologic therapies may be considered in refractory cases or in patients who are intolerant to conventional therapies, reinforcing the importance of individualized therapeutic strategies for systemic vasculitides [7,8].

In contrast, Case 2 had extensive involvement of the aorta and its branches at the time of TA diagnosis, including advanced structural lesions and prior need for valvular surgical intervention. The chosen treatment approach is consistent with contemporary recommendations, since nonspecific initial symptoms such as fever, fatigue, and weight loss contribute to delayed diagnosis, allowing silent progression of vascular inflammation [9]. It is further evident that imaging studies play a fundamental role in accurately characterizing the disease and determining its extent, as they reliably demonstrate structural abnormalities and thereby guide therapeutic decision-making.

Treatment with glucocorticoids combined with biologic therapy using adalimumab was considered after assessment of disease extent, persistence of inflammatory activity, and history of established vascular damage. After therapy initiation, clinical stabilization, reduction of inflammatory markers, and absence of progression of vascular lesions were observed, supporting the effectiveness of tumor necrosis factor inhibitors in refractory cases or in those at high risk of structural progression [10,11]. These cases demonstrate the relevance of biologic therapies as an effective alternative in patients with inadequate response to conventional therapy.

Both cases also highlight the importance of imaging modalities in diagnosis and follow-up of systemic vasculitides. Angiography was decisive for PAN diagnosis and early vascular intervention, whereas CT angiography and other imaging studies allowed assessment of disease extent in TA and monitoring of therapeutic response. Although FDG-PET is described as a valuable tool for evaluating vascular inflammatory activity and therapeutic follow-up, its unavailability in this study reflects limitations frequently encountered in clinical practice [5,12,13].

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This case study directly compares 2 distinct scenarios. In Case 2, with PAN, early diagnosis and immediate vascular intervention achieved effective clinical control and prevented irreversible damage. In contrast, Case 1, with TA, was diagnosed late, and although treatment stabilized disease activity, it did not allow reversal of pre-existing structural sequelae. These cases demonstrate that inflammatory burden and the degree of vascular involvement at diagnosis are critical prognostic factors that influence the reversibility of vascular damage.

Our study has certain limitations. The diagnosis of PAN was based on clinical and angiographic findings without histopathological confirmation, and FDG-PET was not available, which additionally limited direct assessment of vascular inflammatory activity. The follow-up periods were limited, preventing evaluation of long-term outcomes.

This study of 2 cases provides relevant clinical evidence, contributing to the characterization of rare conditions, and shows the need for awareness among healthcare professionals, reflected in the expanded use of imaging and the implementation of immunosuppressive and biological therapies. Detailed investigation and multidisciplinary follow-up are indispensable for optimizing the prognosis and quality of life, especially in patients with atypical or refractory vascular manifestations.

Conclusions

The cases presented here reinforce several highly important clinical lessons, such as the establishment of diagnosis and the appropriate management for each disease modality associated with medium- and large-vessel systemic vasculitides. With regard to refractory hypertension in young patients, especially when associated with vascular findings such as renal artery stenosis, it is important to emphasize how early investigation through vascular imaging studies contributes to ruling out vasculitis. Case 1 demonstrates that rapid performance of angiography, immediate initiation of immunosuppression, and early angioplasty can prevent progression to ischemia and irreversible damage, providing evidence of the impact of timely escalation of therapy and multidisciplinary care.

Delayed recognition of Takayasu arteritis can result in extensive and irreversible cardiovascular complications. In Case 2, diagnostic delay led to progression of inflammatory damage to the aorta and its branches, resulting in the need for aortic valve replacement. This outcome supports that delayed disease identification can allow the development of permanent structural sequelae that could potentially be avoided with early diagnosis and treatment.

Thus, it is reasonable to state that, based on the cases studied, it became evident how the integration of clinical suspicion, high-resolution vascular imaging methods, and multidisciplinary management can significantly modify the disease course. Even with certain limitations regarding available therapeutic decision-making tools—such as access to FDG-PET—timely diagnosis, individualized immunosuppressive and biologic therapy remain essential resources for preventing irreversible vascular damage.

Institution Where Work Was Done

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Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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