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CASE REPORT

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Sudden onset of symptoms in concurrent Takayasu arteritis and Ulcerative colitis: A case report with neurological complications

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Key Clinical Message

The rare co-occurrence of takayasu arteritis (TAK) and ulcerative colitis (UC), presenting with asymptomatic onset and neurological complications, highlights the importance of an integrated diagnostic approach for overlapping autoimmune conditions.

Abstract

We present a rare case of a 44-year-old female diagnosed with both UC and TAK, characterized by an unusual acute asymptomatic onset accompanied by neurological manifestations. The patient exhibited symptoms of acute ischemic stroke along with vascular abnormalities, as well as colon inflammation associated with UC. The patient's asymptomatic presentation at the onset differs from previously reported cases. The presence of additional complications, such as hepatocellular adenoma and primary sclerosing cholangitis, further complicated the diagnostic challenges. The patient's treatment involved a combination of methylprednisolone, azathioprine, and prednisolone leading to improved clinical outcomes. This case emphasizes the complexity involved in diagnosing overlapping conditions and highlights the significance of TAK in presenting atypical manifestations in relation to UC. Furthermore, this case contributes to the limited literature, underscoring the need for early detection and comprehensive treatment approaches.

KEYWORDS

inflammatory bowel disease, takayasu arteritis, ulcerative colitis, Vasculitis

1 | INTRODUCTION

Autoimmune and rheumatologic diseases have the potential to overlap, co-occur and progress to another autoimmune disease in a single subject, and should be considered by the medical teams for a wider range of differential diagnoses in cases with atypical symptoms. Inflammatory bowel disease (IBD) may be presented by extraintestinal symptoms that affect other organs. Diagnosing between IBD-associated conditions or new

primary disease is challenging in these patients. Most studies reported cases of Crohn's disease or ulcerative colitis (UC) developing takayasu arteritis (TAK) or aortitis over time.[1]

UC is a colon inflammatory disease that often relapses.[2] UC impacts 6.4% of TAK subjects, and both diseases share the same genes, including HLA–B*52:01 and IL12B.[3] Despite accurate incidence and prevalence of UC cases developed TAK has not been determined precisely; occasionally, UC patients are complicated by vasculitis like TAK.[4] Hence, we describe a unique case of TAK accompanied by UC.

2 | CASE HISTORY/EXAMINATION

A 44-year-old female with no known family and medical history was brought to the emergency department due to the sudden onset of visual impairment and left hemiparesis. She was admitted to the neurology department with a suspected diagnosis of acute ischemic stroke. Upon physical examination, a notable blood pressure difference of 25 mm Hg was observed between the right and left arms, along with the absence of right ulnar and radial pulses.

3 | METHODS (DIFFERENTIAL DIAGNOSIS, INVESTIGATIONS, AND TREATMENT)

A non-contrast computed tomography (CT) scan of the brain revealed a hypodense area in the right posterior temporal and right occipital lobe, indicative of ischemia in the territories supplied by the posterior cerebral arteries. The patient underwent magnetic resonance venography (MRV) and magnetic resonance arthrography (MRA) of the brain to investigate the case further based on the presenting symptoms and imaging findings. MRA revealed a filling defect in the basilar artery, indicating occlusion and occlusion of the right posterior cerebral artery. However, no pathological findings were reported in the MRV examination. Additionally, cervical MRA exhibited lesions at the origin of the right vertebral artery.

Following a rheumatology consultation, CT angiography of the neck, chest, abdominal aorta, and upper extremities was performed which showed left-sided carotid artery edema, wall thickening of the abdominal aorta, celiac trunk, and proximal renal arteries, indicating TAK. Also, the increased levels of inflammatory markers shown in Table 1 are consistent with this diagnosis and confirm it. Then, as a result of malignancy suspicions, abdominal and pelvic CTs were conducted, which revealed wall edema of the ascending colon and lymphadenopathy, with the largest lymph node measuring 15 mm.

TABLE 1 Laboratory parameters of the patient.

Laboratory parameters	Patient's values	Normal ranges
Leukocyte count, per μL	13,000 (67% Neut, 33% Lymph)	$4–10\times10^3$
Hemoglobin, g/dL	10.7	12.3–15.3
MCV	89	80–100
Platelet count per μL	291,000	150,000–450,000
Albumin		3.5–5.2
ESR, mm/h	105	0–30
CRP, mg/L	31	<6
AST g/dL	153	8–35
ALT, g/dL	213	8–35
Total Bilirubin	2.4	0.2–1.2
Direct Bilirubin	1.3	0–0.4
ALP IU/L	3696	64–306
LDH, IU/L	215	100–300
BUN, mg/dL	11.6	7–20
Creatinine, mg/dL	0.9	0.5–1.1
Gama GT	1244	8–42
ANA, IU/mL	Negative	<12 Negative
		12–18 Equivocal
		>18 Positive
Anti-dsDNA	Negative	–
C3, mg/dL	120	90–180
C4, mg/dL	25	10–40
APS Antibody	Negative	–
P-ANCA	Negative	–
C-ANCA	Negative	–

Following abdominopelvic CT findings and elevated liver function tests (LFTs), a colonoscopy was performed. The colonoscopy revealed friable, erythematous, and congested mucosae in the splenic flexure, descending colon, sigmoid, and rectum. Additionally, cryptitis and crypt abscesses were observed without vasculitis, indicative of UC (refer to Figures 1 and 2). Based on the elevated levels of LFTs, alkaline phosphatase (Alk. p), and gamma-glutamyltransferase (GT), Primary sclerosing cholangitis was considered in the context of IBD.

A mass was detected during hepatic MRI, which was later confirmed as hepatocellular adenoma through biopsy. Subsequently, the patient's biliary ducts were reported as normal in Magnetic Resonance Cholangiopancreatography. Based on the clinical, laboratory, and imaging findings, the patient was diagnosed concurrently with IBD, specifically UC and TAK.

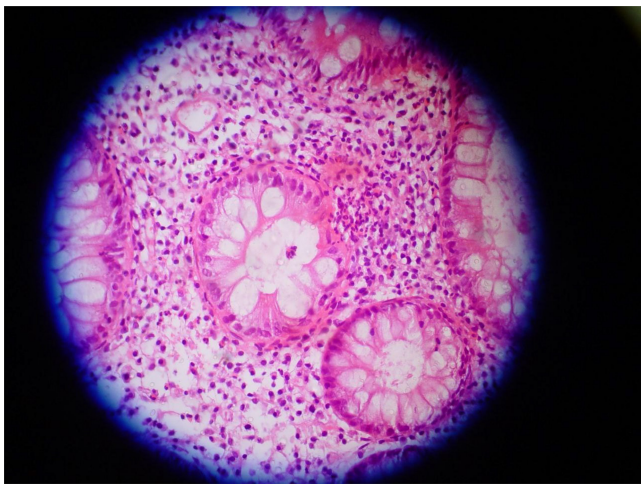

FIGURE 1 Cryptitis and crypt abscess.

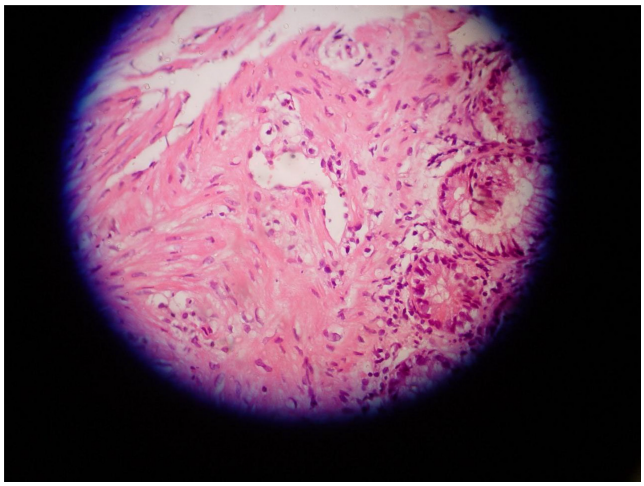

FIGURE 2 Normal vessel without features of vasculitis.

4 | CONCLUSION AND RESULTS (OUTCOME AND FOLLOW-UP)

The initial management approach began with intravenous pulse methylprednisolone 1000 mg daily for 3 days and was followed by planned treatment with 100 mg of Azathioprine and 60 mg of prednisolone daily. During the follow-up visit, her muscle forces improved, and LFTs and inflammatory markers decreased to normal values. Furthermore, right radial and ulnar pulses have been improved. However, Alk. p was as high as 600–700 IU/L and differences in systolic blood pressure.

5 | DISCUSSION

We present a complex and uncommon case of a 44-year-old female patient with concurrent TAK and UC. The patient presented with atypical symptoms, including neurological complications, vascular abnormalities, and liver lesions. The patient exhibited atypical symptoms, including neurological complications, vascular abnormalities, and liver lesions. A thorough evaluation led to the diagnosis, and the patient is currently receiving appropriate treatment. This case emphasizes the difficulties in diagnosing and managing overlapping conditions manifesting atypical presentations.

TAK is a form of large-vessel vasculitis that predominantly impacts the aorta and the main aorta branches, resulting in dilatation, occlusion, and stenosis of the affected vessels.[5] TAK is known to coexist with other autoimmune diseases occasionally and predominantly affects young females.[6–8] One of the most prevalent diseases that are TAK complicated is UC.[3] UC is a large intestine chronic disease sometimes correlated with rectal inflammation and usually advances proximally to affect more colon areas.[9] While the onset of UC typically occurs between the ages of 15 and 30, there are no apparent gender differences. Despite differences in the epidemiology of UC and TAK, they exhibit shared genes.[10]

TAK is defined by the vasa vasorum inflammation, which results in inflammatory cell infiltration and preceding arterial dilatation or stenosis. The roles of myeloid cells, T cells, and recently recognized novel autoantibodies, such as those detected in UC are explored. The association between TAK and UC may result from the same etiology.[11]

The number of studies reporting TAK and UC together is increasing; they share common genetic predispositions and pathophysiological mechanisms. When treating these subjects, if they fail to respond to immunosuppressant management and conventional corticosteroids, targeted synthetic and biological disease-modifying antirheumatic drugs can be utilized to treat them.[12]

UC was diagnosed in 6.4% of TAK cases found by a large case series from Japan.[13] Furthermore, TAK developed earlier in cases with UC compared to cases without UC. Sy et al.,[14] in a case series from North America, presented 12 IBD cases diagnosed with TAK, too. Also, Akiyama et al.[15] found that 9.2% of TAK cases in his study developed IBD. 69% of IBD patients preceded TAK, although just 31% of TAK patients developed IBD. The median time between detection, when TAK preceded IBD and when IBD preceded TAK, were 7.5 and four, respectively. For TAK cases for colonoscopy, indications were positive for fecal occult blood test (8%), diarrhea (15%), and melena or hematochezia (69%).

A predominantly persistent cell-mediated inflammatory response may significantly contribute to the pathophysiology of TAK in individuals with susceptible genetic factors. The inflammatory cascade is initiated by a triggering antigen

that acts similarly to normal cellular antigens, subsequently activating adaptive immunity and triggering the release of cytokines. This process ultimately leads to tissue fibrosis and the formation of granulomas, which are hallmark features of TAK pathology.[16] In the pathogenesis of TAK, several cytokines, including IL-1, IL-6, interferon-gamma, and TNF-alpha, can have vital functions. Similar to the IBD causation, these cytokines are considered to have roles.[17] The antigen that activates the TAK inflammatory process will be well exposed because of the bacterial translocation or tissue damage seen in IBD.[18]

A study showed that a 32-year-old Caucasian woman presented with atypical symptoms of TAK, including carotidynia, visual changes, and upper extremity paresthesia. Imaging studies revealed 50%–69% right carotid artery stenosis, but the patient did not meet the full criteria for a diagnosis of TAK. She was treated with corticosteroids but developed bradycardia, and the medication was discontinued. The patient subsequently developed UC, suggesting a possible link between the two conditions.[19] Also, our case closely resembled theirs with some variations in the initial presentation, and both two cases underscore the importance of considering TAK in patients with atypical symptoms, particularly those who also have IBD. Early diagnosis and treatment can help improve outcomes and prevent serious complications. Furthermore, Pyo JY and colleagues[20] reported a case of a 38-year-old female with a history of UC and optic neuritis presenting with neck pain, headache, and dizziness. Significant physical examination findings included absent brachial and radial pulses. Laboratory tests revealed an elevated CRP, ESR, platelet count, and a positive anti-proteinase 3 despite standard urine, renal, and LFTs. Additionally, tests for anti-myeloperoxidase (MPO), anti-glycoprotein, anticardiolipin, and antineutrophil cytoplasmic antibodies were negative. The patient exhibited significant Carotid artery stenosis, which was managed effectively with azathioprine and prednisolone, consequently diminishing inflammation markers and ameliorating symptoms.

Watanabe S et al.[21] showed a case involving a 27-year-old man with a history of UC who developed acute ischemic colitis associated with mild stenosis of the superior mesenteric artery due to TAK. The patient's condition was managed conservatively, and the ischemic colitis improved with follow-up, revealing UC activity limited to the rectum. Unlike the Watanabe S et al.[21] case, our case involves additional complications such as hepatocellular adenoma, Primary Sclerosing Cholangitis, and lymphadenopathy, requiring a multidisciplinary approach to diagnosis and management. Both cases underscore the importance of considering vasculitis in patients with UC and the necessity for thorough evaluations to discern overlapping conditions, ensuring comprehensive and effective care. Additionally, another study revealed a case of an 18-year-old girl who initially presented with colitis characterized by occasional high fever, ultimately diagnosed as TAK with IBD unclassified. The colonic inflammation exhibited atypical features, including a discontinuous distribution with greater intensity in the proximal colon, deviating from the typical UC pattern. However, over a 10-year observation period, the colonic inflammation displayed variations and evolved to resemble UC. This long-term observation provides valuable insights into the changing nature of TA-related colonic inflammation, shedding light on this condition's complex and dynamic aspects over time.[22]

6 | CONCLUSION

The emergence of TAK concurrent with UC presents a clinical challenge due to the atypical presentation and the rarity of concurrent diseases. Our case report underscores the complex nature of diagnosis when two overlapping conditions manifest together, as in the sudden onset of neurologically complicated TAK in a patient with UC. This dual presentation emphasizes the need for elevated clinical knowledge and a multidisciplinary approach to diagnose rare and atypical disease manifestations effectively. The overlapping symptomatology can confuse the clinical picture, making an early and accurate diagnosis critical for initiating appropriate treatment strategies. Improving clinical outcomes for such patients hinges upon early detection and comprehensive treatment regimens, as illustrated by the successful management of our case with immunosuppressive therapy. Our report contributes to the limited literature and reinforces the significance of a thorough investigation into atypical presentations of concurrent autoimmune diseases.

AUTHOR CONTRIBUTIONS

Amirreza Khalaji: Writing – original draft; writing – review and editing. **Taha Mahdian:** Data curation; investigation. **Amir Taher Eftekharsadat:** Resources; supervision; validation. **Mehdi Jafarpour:** Conceptualization; investigation; methodology; project administration; resources; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

None.

DATA AVAILABILITY STATEMENT

The data supporting the findings of this research are available upon reasonable request from the corresponding author.

ETHICS STATEMENT

This study was performed according to the principles outlined by the World Medical Association's Declaration of Helsinki on experimentation involving human subjects, as revised in 2000, and has been approved by the ethics committee of the Tabriz University of Medical Sciences.

CONSENT

Written informed consent was obtained from the patient to publish this report and clinical images. Consent has been signed and collected following the journal's patient consent policy.

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