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CASE REPORT

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A challenging case of Takayasu's arteritis in a young male with various manifestations and poor outcome

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Abstract

A 17-year-old boy complaining of progressive dyspnea, fever, palpitations, a 22 mm Hg blood pressure difference between the 2 arms, and arm claudication. He had a history of psoriasis-like skin lesions and bronchiectasis. Echocardiography revealed a reduced left ventricular ejection fraction, severe eccentric aortic insufficiency, circumferential aortic wall thickening, and a dilated ascending aorta with severe atherosclerotic changes. Based on imaging findings, a TA diagnosis was suggested. During his follow-up, the patient developed strabismus, blurred vision, and right sixth cranial nerve paralysis and went into a deep coma. Unfortunately, after 6 months of treatment, he expired due to COVID-19 infection.

Keywords Case report, Takayasu, Cardiovascular disease, Vasculitis

Introduction

Takayasu's arteritis (TA) is classified as a rare granulomatous panarteritis affecting the aorta and its major branches. It is predominant in women and has a peak incidence between ages 20 and 40 [1].

The most frequent cardiac manifestations of TA are valvular abnormalities, found in more than 60% of patients [2], and aortic regurgitation is the most common type of valvular heart disease [3]. Other non-common cardiac presentations are acute myocardial infarction, myocarditis, pericarditis, and pulmonary hypertension [4].

Noninvasive imaging techniques, such as magnetic resonance angiography (MRA) and computed tomography angiography (CTA), are playing important role for diagnosing TA and extension of vascular involvement.

Case description

A 17-year-old boy presented to the emergency department complaining of progressive dyspnea with New York Heart Association functional class III. Over the prior decade, he had visited medical centers on numerous occasions. At 6 years of age, he was taken to a pediatrician due to dyspnea, urticaria, and food allergies. Two weeks after treatment initiation for allergies, his symptoms changed to skin erythematous plaques with thick scales that covered his elbows and thighs, suspicious of psoriasis. But he discontinued all treatments without further evaluation of skin lesions. Nevertheless, his dyspnea progressed to the extent that after 2 years, he was unable to participate in football games with his peers. He was taken to a medical center, where he received treatment for bronchiectasis and asthma.

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On arrival at our facility, at age 17, with progressive dyspnea and right-arm claudication, a 22 mm Hg blood pressure difference between the arms, symmetrical and bounding peripheral pulses, tachycardia (103 bpm), oral temperature of 38 ℃, and oxygen saturation of 98% in room air. He was eupneic at rest and had mild basal crackles on pulmonary auscultation and early diastolic heart murmurs (III/VI) on cardiac auscultation.

Electrocardiography demonstrated sinus tachycardia. A chest X-ray showed cardiomegaly and bilateral infiltration. Laboratory evaluations revealed a white blood cell count of 14,200 cells/mm^3 (neutrophil seg = 72%, lymphocyte = 7%, and eosinophil = 15%), a C-reactive protein level of 46 mg/L, an erythrocyte sedimentation rate of 23 and a B-type natriuretic peptide (BNP) level of 4360 pg/mL, with no other alterations.

Transthoracic echocardiography revealed severe left ventricular (LV) dilation with severe LV systolic dysfunction (global ejection fraction = 20%) and global hypokinesia; severe eccentric aortic insufficiency; circumferential aortic wall thickening; a dilated ascending aorta (5.2 cm) with severe atherosclerotic changes; holodiastolic flow reversal in the descending aorta; a narrowing just distal to the origin of the subclavian artery with systolic turbulence and without a diastolic tail; circumferential thickening of the abdominal aorta with atherosclerotic changes and moderate pulmonary hypertension (Fig. 1).

The patient was admitted to the cardiac care unit. A CTA of the total aorta illustrated dilation of the ascending aorta, calcification of the aortic wall from the ascending to the mid-descending thoracic aorta, and irregularity of the aortic wall (Fig. 2).

MRA also revealed a dilated ascending aorta (max at 54 mm at the right pulmonary artery level), a thickened and irregular aortic wall, atherosclerotic or intruding lesions from the vessel wall into the arch and descending aorta causing narrowing in the narrowest part of the arch (11 mm), and multiple aneurysmal regions in the ascending and descending aorta. No evidence of myocardial inflammation or edema was detected in short tau inversion recovery (STIR)-T2. In the gadolinium study, no thrombus was evident in the LV and right ventricle in the early phase, while the late phase revealed mid-wall enhancement in the mid-septal wall (Fig. 3).

The patient's clinical and imaging findings, as well as rheumatology consultations, confirmed the diagnosis of TA.

Afterward, due to the patient's symptoms, guideline-directed medical therapy (GDMT) for heart failure, namely angiotensin-converting enzyme inhibitor, mineralocorticoid receptor antagonist, Low dose Beta Blocker, SGLT2 inhibitor, was started with acceptable tolerance. At discharge, the patient was clinically stable with no signs of heart failure, normal C-reactive protein, a progressive reduction in BNP levels without changing in LVEF in last echocardiography. Disease-modifying drugs for heart failure were maintained at discharge. Because of a new TA flare-up and severe aortic wall calcification, surgical options for treatment were not chosen in this session of hospitalization.

After discharge, 2weaks later, the patient was referred to a rheumatology center. He received Methylprednisolone (500 mg daily for four days) therapy. Laboratory studies also revealed negative antineutrophil cytoplasmic antibodies (ANCA), rheumatoid factor (RF), antinuclear antibodies (ANA), and HLA-B27, as well as normal C3 and C4. After an acceptable response to corticosteroids, he was discharged to continue treatment at home via monotherapy with oral prednisolone (30 mg daily, tapered to 10 mg daily).

Two months later, the patient experienced changes in his symptoms; lethargy, ataxia, blurred vision, strabismus, right sixth cranial nerve paralysis, hallucination, and muscle weakness with a decreased muscle force (4/5, symmetrically, both upper and lower limbs) were recorded. Based on ophthalmology evaluation, ischemic ocular TA was mentioned. Moreover, in neurologic evaluation, the brain CT scan showed hypodensity in the left basal ganglia and right parietal. A brain MRI revealed some degrees of vasculitis in the major branches of the cerebral arteries. Methylprednisolone pulse therapy was started, then mycophenolate mofetil (500 mg BID) and infliximab (anti-TNF recombinant antibody) were added to the basic treatment with oral prednisolone. Subsequently, the patient experienced one episode of deep

(See figure on next page.)

Fig. 1 Transthoracic echocardiogram obtained at admission. **A** Parasternal long axis view showing dilation of ascending aorta (5.2 cm). **B** Parasternal long axis view showing AI vena contracta: 0.62cm in favor of severe AI. **C** Four chamber view showing LV end diastolic volume index: 114.9 ml/m2. **D** Five chamber view showing AI Doppler. **E** Diastolic flow reversal in descending thoracic aorta by pulse-wave Doppler and end-diastolic velocity is greater than 20 cm/s. **F** A narrowing is seen just distal to the origin of the subclavian artery. **G** The image shows circumferential thickening of the abdominal aorta with atherosclerotic changes and the normal size of the abdominal aorta in the proximal part (2.24 cm). **H** A narrowing is seen just distal to the origin of the subclavian artery with systolic turbulence (the peak pressure gradient = 20 mm Hg) and without a diastolic tail

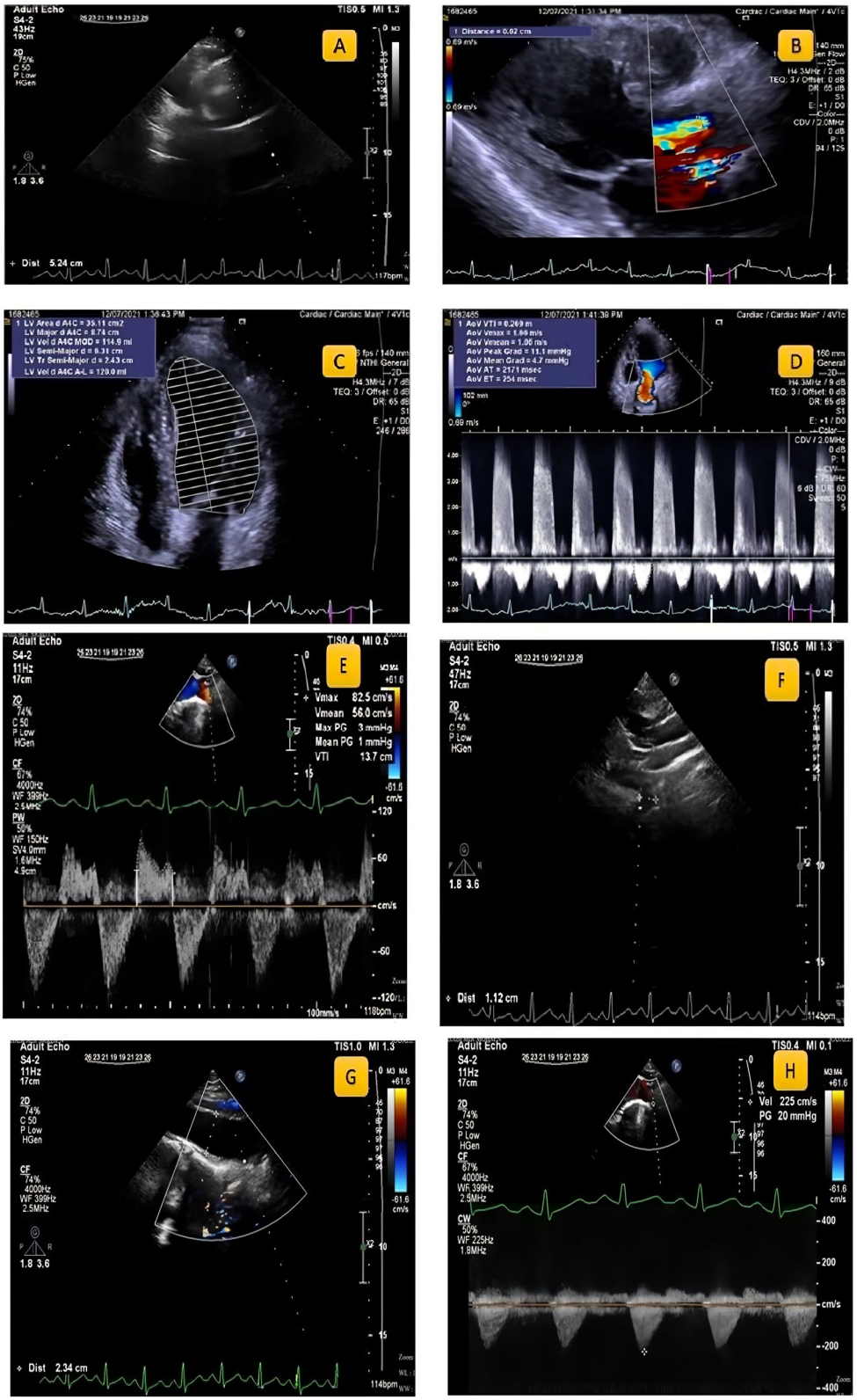

Fig. 1 (See legend on previous page.)

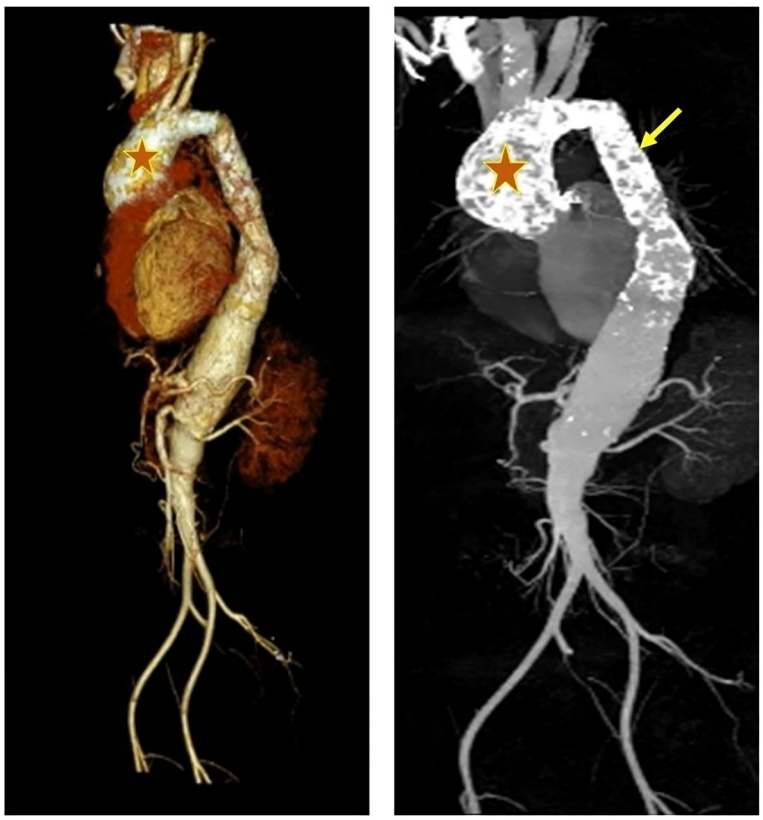

Fig. 2 The aortic computed tomography angiography with 2 different volume-rendering technique (VRT) reconstructions reveals dilation of the ascending aorta (the asterisk), calcification of the aortic wall from the ascending to the mid-descending thoracic aorta (the yellow arrow), and irregularity of the aortic wall. Abbreviations: VRT: (Volume Rendering Technique)

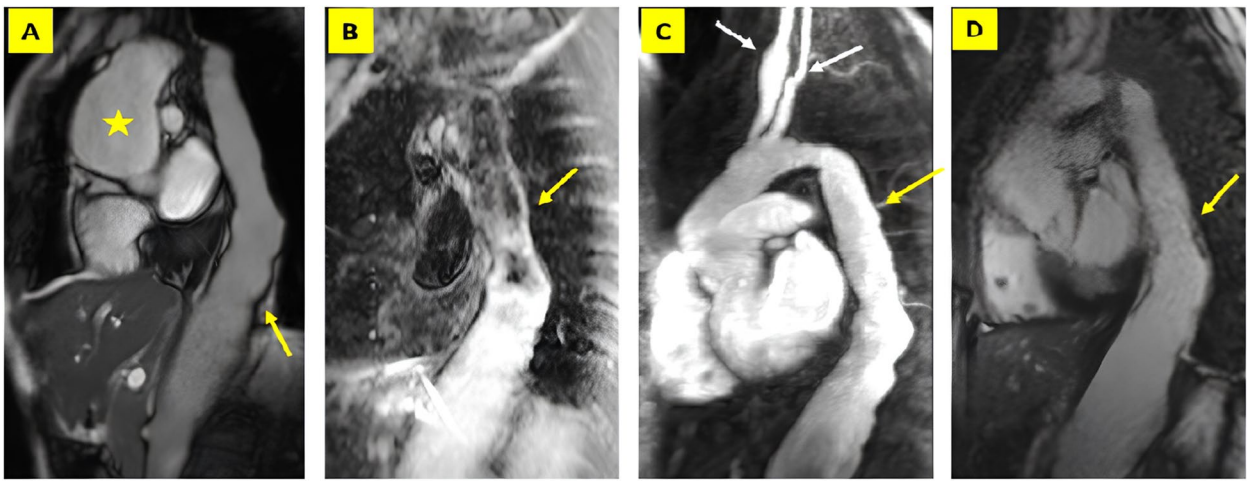

Fig. 3 Takayasu arteritis is displayed in different cardiac magnetic resonance sequences. **A** The aortic double-oblique candy-cane view demonstrates irregularity of the aortic wall (the yellow arrow) and dilation of the ascending aorta (the asterisk). **B** The short tau inversion recovery (STIR) image shows edema of the aortic wall (the yellow arrow). **C** The magnetic resonance angiography depicts irregularity, localized dilation, and mild narrowing of the aortic arch branches (the white arrows) and irregularity of the aortic wall (the yellow arrow). **D** The late gadolinium enhancement (LGE) sequence illustrates aortic wall fibrosis (the yellow arrow)

coma for a period of four days. Following the administration of methylprednisolone pulse therapy (500 mg daily) for a duration of four days, the patient's level of consciousness returned to a normal state.

Eventually, 2 months later, Ipsilateral swelling of the face, dyspnea, multiple mouth aphthous ulcers, epistaxis and loss of consciousness occurred. The initial evaluation showed a positive COVID-19 polymerase chain reaction test and mucormycosis with a perforated septum and a saddle nose deformity. Additionally, cytomegalovirus and Klebsiella pneumonia were detected in bronchoscopic specimens. Broad-spectrum antibiotics and ganciclovir, dexamethasone, and remdesivir comprised the principal treatments. However, after 14 days, the patient died in the setting of bradycardia and asystole.

Discussion

In general, we encountered a young boy with Takayasu's arteritis, presenting with atypical features at first (urticaria, bronchiectasis, psoriasis), which resulted in a near-decade-long misdiagnosis. Thereafter, the condition progressed to heart failure, severe aortic regurgitation, aortic-cerebral-ocular vasculitis without coronary involvement, porcelain aorta and multiple flare-ups. It is regrettable to report that he died from complications of coronavirus disease 2019 (Covid-19).

Based on the American College of Rheumatology (ACR)/the European League against Rheumatism (EULAR) Classification Criteria for TA (2022), the most common features include claudication, angina, vascular bruits, and reduced pulses in the upper extremities, carotid artery abnormalities, and systolic blood pressure differences between arms [5]. Our patient exhibited imaging criteria, claudication, and systolic blood pressure differences, leading to the proposal of Takayasu arteritis as the primary diagnosis.

While TA is much more frequent in women aged between 20 and 40, our patient was a young man who became symptomatic at age 17.

Unfortunately, the patient never had a meticulous examination of his skin lesions; still, case reports indicate that the concomitance of TA and psoriasis in a patient is, albeit rare, probably not coincidental, suggesting shared pathogenetic mechanisms [6, 7].

TA have also some atypical features. Bronchiectasis is a case in point. It is quite rare for Takayasu arteritis to

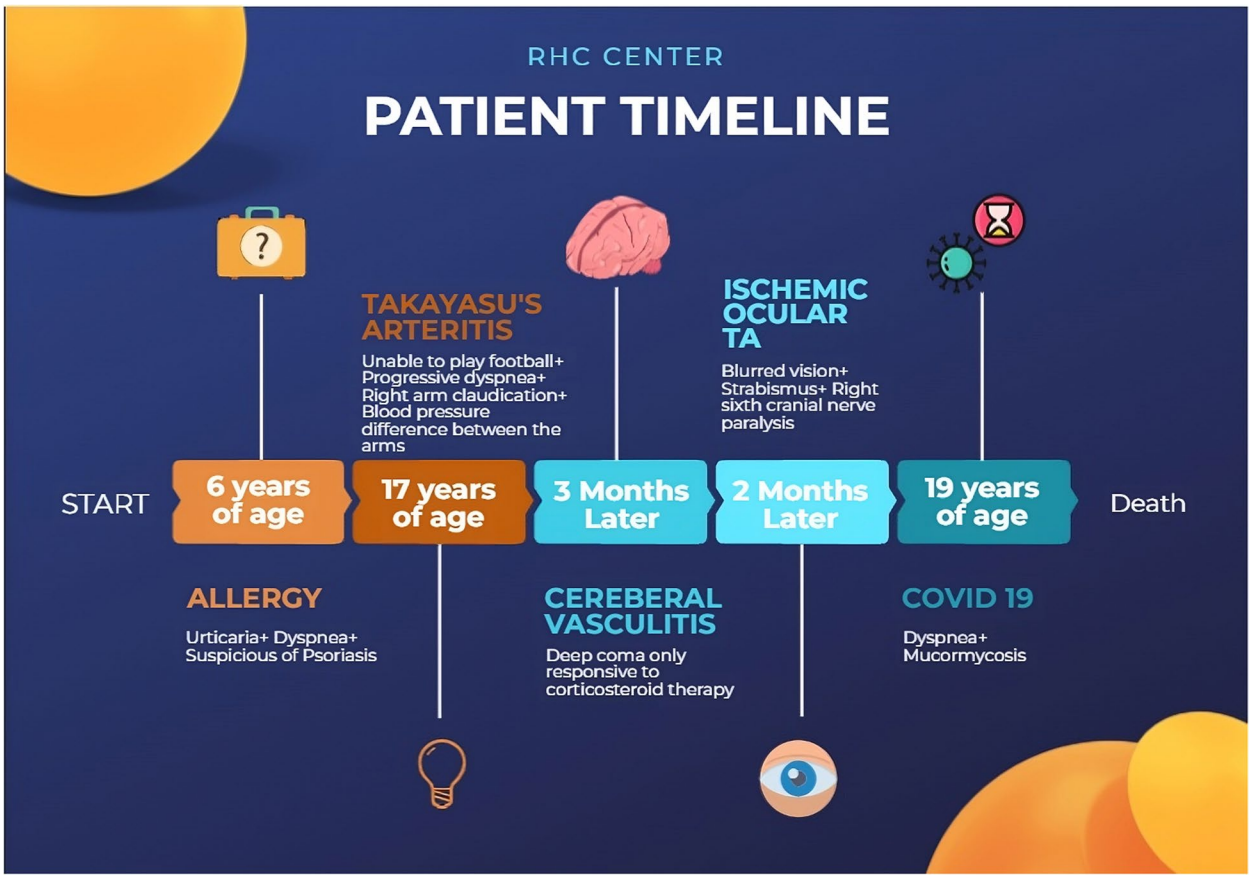

Fig. 4 Patient timeline

manifest with bronchiectasis in the absence of vascular symptoms. A review of the literature revealed a paucity of cases in which Takayasu arteritis and bronchiectasis occur concurrently [8]. All cases exhibited a long systemic phase before the onset of definite vascular symptoms. Other tragic events were cerebral and ocular vasculitis, which manifested as coma and strabismus and highlight the importance of early diagnosis and effective immunosuppressive treatment.

Moreover, there are no suitable surgical or endovascular procedures for patients with TA and severe aortic insufficiency, severe calcification of the aorta (defined as a porcelain aorta). Indeed, manipulate friable and inflamed tissue increase the risk of valve detachment and anastomotic aneurysms, as well as manipulation of a porcelain aorta, which increases the risk of stroke and embolization.

Despite treatment with corticosteroids, infliximab, and mycophenolate mofetil, our patient's rheumatologic symptoms continued to worsen. Fortunately, recent years have witnessed the emergence of new targeted biological agents for TA treatment [9]. Nonetheless, the increased infection risk precluded us from utilizing them.

The last, but by no means the least, tragic event was our patient's contracting COVID-19. Immunosuppressive agents, as the integral component of TA control, can increase susceptibility to infections. We commenced the latest universal treatment for COVID-19 promptly. Our patient, however, failed to respond to initial treatments, which indicates the lifesaving role of vaccination in patients with TA.

Conclusions

Here, we reported a rare and intriguing case of TA in a patient who presented bronchiectasis and psoriasis prior to the onset of vascular symptoms. These symptoms progressed to severe aortic insufficiency, porcelain aorta, heart failure, aortic-cerebral-ocular vasculitis over a decade. Indeed, our patient exemplifies the significance of prompt diagnosis, efficacious immunosuppressive therapy, novel endovascular or surgical approaches, and close monitoring (Fig. 4).

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Not applicable.

Patient permission
Although our manuscript does not reveal the patient identity in any part, we have consent form include permission of patient legal guardian (his father). Undoubtedly, we would provide the copy if specifically requested by the journal.

Declarations of interest
None.

Declaration of generative AI and AI-assisted technologies in the writing process
None.

Clinical trial number
Not applicable.

Authors' contributions
Corresponding author: Saba Simiyari. Shabnam Boudagh, Marzieh Mirtajaddini, Simin Almasi and Saba Simiyari wrote the main manuscript text. Shabnam Boudagh, Nahid Rezayean and Saba Simiyari prepared figures 1-3. All authors reviewed the manuscript.

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Data availability
All data generated or analyzed during this study are included in this published article [and its supplementary information files].

Declarations

Ethics approval and consent to participate
The patient's father, acting as the patient's legal guardian, provided informed consent for the participation and publication of the patient's information in the journal. The special consent form for Rajaie Hospital, as well as the special BMC journal consent form, can be accessed in Persian and English as a PDF file in the "related files" section.

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Competing interests
The authors declare no competing interests.

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