Case report analysis using the CARE case report guidelines

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CARE
case report guidelines

CARE Checklist of information to include when writing a case report

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Topic	Item	Checklist item description	Reported on Line
Title	1	The diagnosis or intervention of primary focus followed by the words "case report"	Page 1 line 1
Key Words	2	2 to 5 key words that identify diagnoses or interventions in this case report, including "case report"	Not reported
Abstract (no references)	3a	Introduction: What is unique about this case and what does it add to the scientific literature?	Page 1 lines 5
	3b	Main symptoms and/or important clinical findings	Page 1 line 5
	3c	The main diagnoses, therapeutic interventions, and outcomes	Not reported
	3d	Conclusion—What is the main "take-away" lesson(s) from this case?	Page 1 lines 7
Introduction	4	One or two paragraphs summarizing why this case is unique (**may include references**)	Page 1 lines 15
Patient Information	5a	De-identified patient specific information.	Page 1 lines 21
	5b	Primary concerns and symptoms of the patient.	Not reported
	5c	Medical, family, and psycho-social history including relevant genetic information	Not reported
	5d	Relevant past interventions with outcomes	Not reported
Clinical Findings	6	Describe significant physical examination (PE) and important clinical findings.	Not reported
Timeline	7	Historical and current information from this episode of care organized as a timeline	Not reported
Diagnostic Assessment	8a	Diagnostic testing (such as PE, laboratory testing, imaging, surveys).	Page 2 lines 23
	8b	Diagnostic challenges (such as access to testing, financial, or cultural)	Not reported
	8c	Diagnosis (including other diagnoses considered)	Page 2 lines 24
	8d	Prognosis (such as staging in oncology) where applicable	Not reported
Therapeutic Intervention	9a	Types of therapeutic intervention (such as pharmacologic, surgical, preventive, self-care)	Page 2 lines 27
	9b	Administration of therapeutic intervention (such as dosage, strength, duration)	Page 2 lines 27
	9c	Changes in therapeutic intervention (with rationale)	Page 3 lines
Follow-up and Outcomes	10a	Clinician and patient-assessed outcomes (if available)	Page 3 lines 24
	10b	Important follow-up diagnostic and other test results	Page 3 lines 25
	10c	Intervention adherence and tolerability (How was this assessed?)	Not reported
	10d	Adverse and unanticipated events	Not reported
Discussion	11a	A scientific discussion of the strengths AND limitations associated with this case report	Page 3 lines 30
	11b	Discussion of the relevant medical literature **with references**.	Page 3 lines 30
	11c	The scientific rationale for any conclusions (including assessment of possible causes)	Page 3 lines 2
	11d	The primary "take-away" lessons of this case report (without references) in a one paragraph conclusion	Page 3 lines 14
Patient Perspective	12	The patient should share their perspective in one to two paragraphs on the treatment(s) they received	Not reported
Informed Consent	13	Did the patient give informed consent? Please provide if requested	Yes ☐ No ☐

title

- criteria 1:The diagnosis or intervention of primary focus followed by the words “case report”
- Comment: fulfilled

Case Report

A case of aortic dissection in familial Mediterranean fever ☆

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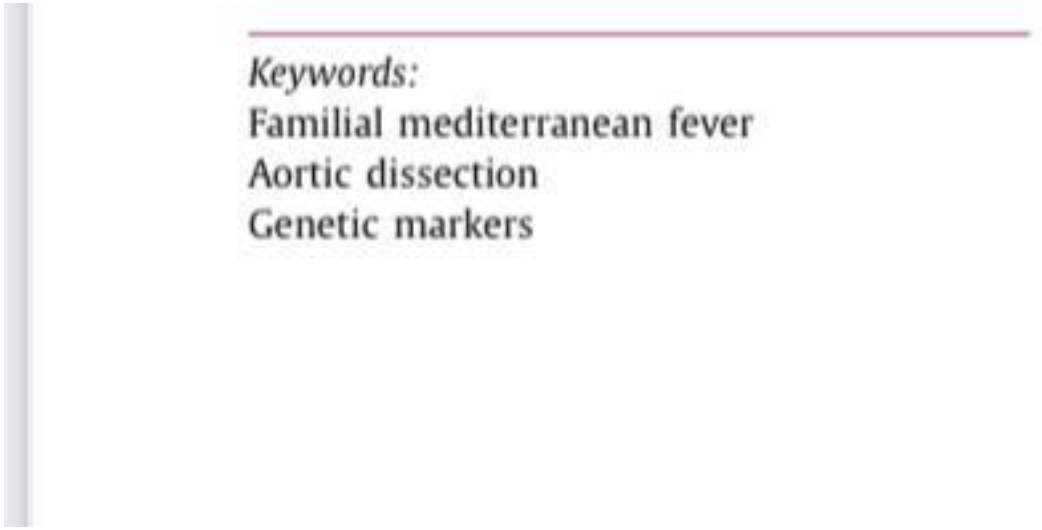
Key Words

- 2: 2 to 5 key words that identify diagnoses or interventions in this case report , including "case report"
- Comment: doesn’t include case report



Keywords:
Familial mediterranean fever
Aortic dissection
Genetic markers

Abstract

- 3a: Introduction: What is unique about this case and what does it add to the scientific literature?

  3b: Main symptoms and/or important clinical findings

  3c: The main diagnoses, therapeutic interventions, and outcomes

  3d Conclusion—What is the main "take-away" lesson(s) from this case?

- Comment: all are fulfilled except that the therapeutic interventions, and outcomes were not mentioned

ABSTRACT

Familial Mediterranean fever (FMF) is an ethnicity-specific autoinflammatory disease prevalent in the Arab world. It has various presentations including amyloidosis, small-vessel vasculitis, and pericarditis. Different mutations in the Mediterranean fever (MEFV) gene can cause this disorder and it is inherited in an autosomal recessive pattern. We present a case of a 23-year-old man of Iranian descent with history of periodic fever, diagnosed as Stanford type A aortic dissection secondary to an acute attack of FMF. To the best of our knowledge, this is the first case reported of such a complication.

Learning objectives:

1. Diagnosis of familial Mediterranean fever is a diagnosis of exclusion as many diseases involve large arteries and present with aortic involvement.
2. Familial Mediterranean fever usually involves small- and medium-sized arteries but unusual case presentations can occur in some patients involving the aorta.
3. Physicians in Pakistan must be made aware of the rapidly increasing prevalence of familial Mediterranean fever due to migration of susceptible ethnic groups in order to manage patients while they are still amenable to treatment.

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Introduction

- 4: One or two paragraphs summarizing why this case is unique (may include references)
- Comment: fulfilled and it includes references

Introduction

Familial Mediterranean fever (FMF) is an ethnicity-specific autoinflammatory disease of the Mediterranean region. It is highly prevalent in Turks, Arabs, Jews, and Armenians, although sporadic cases are reported in other regions as well. Different mutations in the Mediterranean fever (MEFV) gene can cause this disorder and it is inherited in an autosomal recessive pattern. The M694V mutation is the most common in various ethnic groups [1,2].

FMF is characterized by attacks of fever and polyserositis, including peritonitis, pleuritic, and pericarditis [3]. Case reports have suggested a broad spectrum of clinical presentations such as renal vasculitis to fatal myocardial infarctions (MI). Vasculitides usually manifest in small to medium arteries in the form of Henoch Schonlein purpura (HSP) or polyarteritis nodosa (PAN) [4]. Large

vessel arteritis and its associated complications is rarely reported with FMF and to our knowledge associated aortic dissection has never been reported.

Patient Information

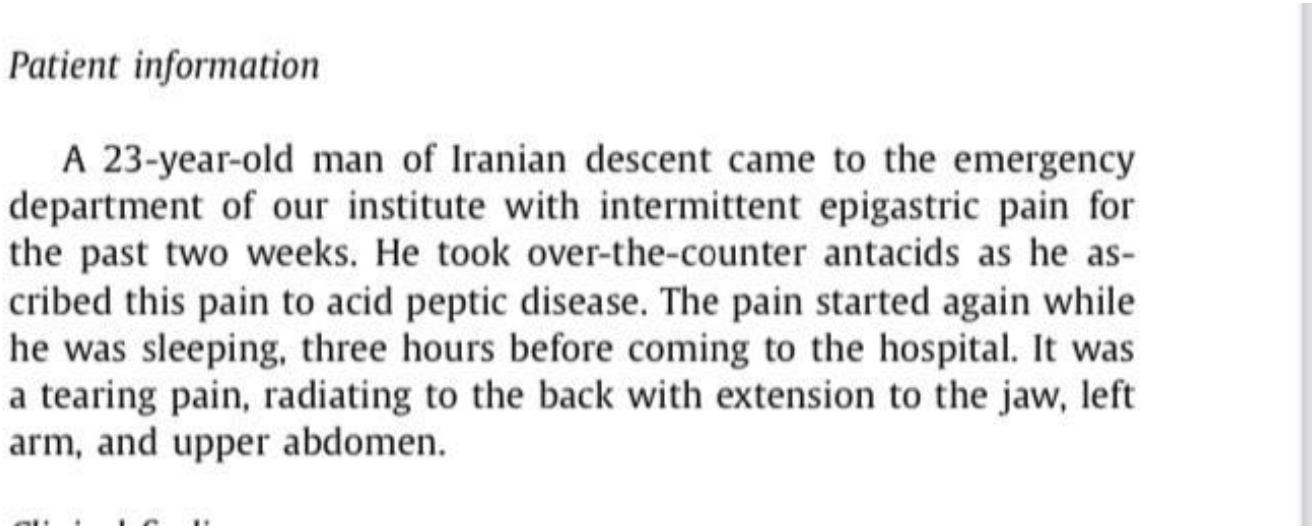
- 5a:De-identified patient specific information.

5b: Primary concerns and symptoms of the patient.

5c :Medical, family, and psycho-social history including relevant genetic information

5d:Relevant past interventions with outcomes

- Comment: regarding the personal history the occupation was not mentioned. the chief complaint of the patient was stated with good analysis of the complain, however the Medical, family, psycho-social history and Relevant past interventions with outcomes were not mentioned



Patient information

A 23-year-old man of Iranian descent came to the emergency department of our institute with intermittent epigastric pain for the past two weeks. He took over-the-counter antacids as he ascribed this pain to acid peptic disease. The pain started again while he was sleeping, three hours before coming to the hospital. It was a tearing pain, radiating to the back with extension to the jaw, left arm, and upper abdomen.

Clinical Findings

- 6 :Describe significant physical examination (PE) and important clinical findings
- Comment :not fulfilled as there were no (PE) findings weather general or local described, also the items of past history and previous interventions that were missing in the patient information section were mentioned here in the clinical findings

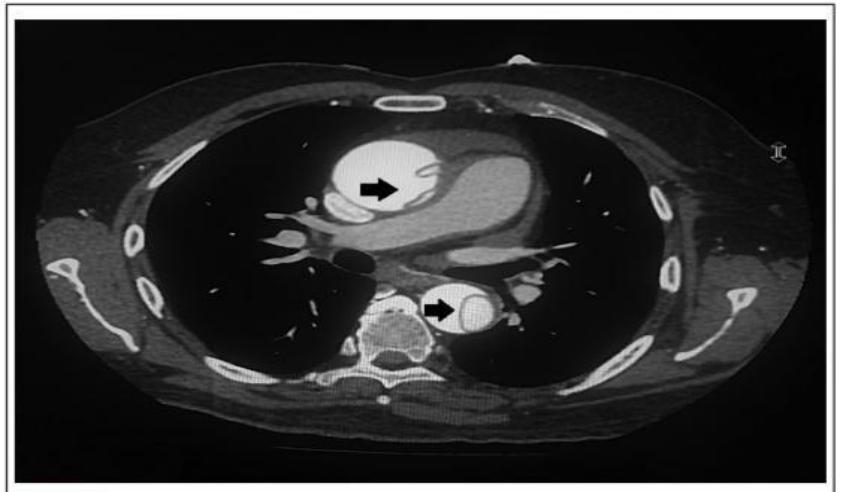


Fig. 1. Axial view on computed tomography showing aortic dissection starting from ascending aorta and continuing forward to the descending aorta.

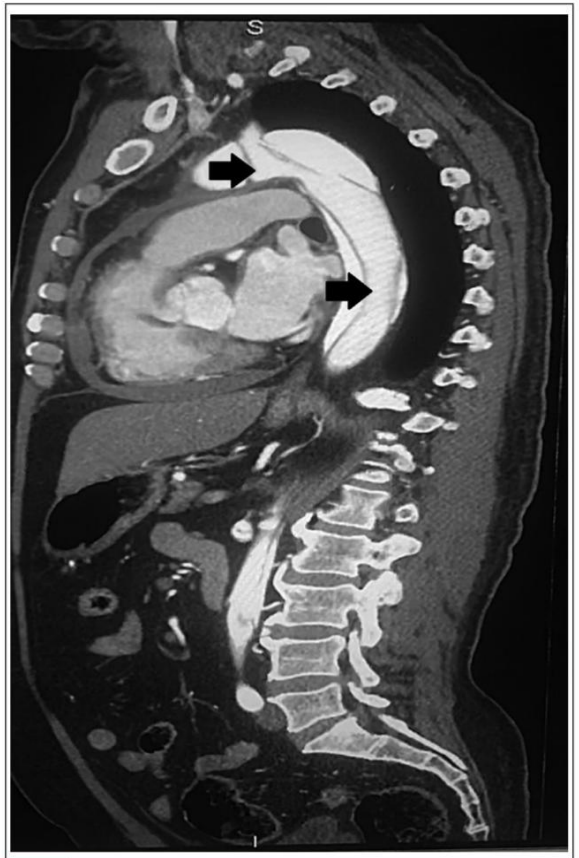


Fig. 2. Sagittal view on computed tomography. Black arrows show aortic dissection coursing through the abdominal aorta.

Clinical findings

Our patient gave no history of ischemic heart disease (IHD), diabetes, hypertension, dyslipidemias, chronic kidney disease (CKD), or smoking. There was a history of periodic fever since childhood which spiked spontaneously between 37.8 to 38.9 °C and then subsided within a week after taking antipyretics. This fever was associated with myalgias, arthritis of multiple joints, and backache. There was a history of laparotomy 6 years previously, when the patient was 13 years old, with suspicion of acute abdomen. It did not show any significant findings and his appendectomy was done subsequently. There was no history of mouth or genital ulcers, conjunctivitis, or skin rash. He was not taking any drugs and there was no history of exertion before chest pain.

Timeline

- 7 :Historical and current information from this episode of care organized as a timeline
- Comment: not fulfilled

Diagnostic Assessment

- 8a: Diagnostic testing (such as PE, laboratory testing, imaging, surveys)

8b: Diagnostic challenges (such as access to testing, financial, or cultural)

8c: Diagnosis (including other diagnoses considered)

8d: Prognosis (such as staging in oncology) where applicable

- Comment: diagnostic testing was mentioned and the diagnosis was stated after showing in the Computed tomography of the aorta with no other diagnosis considered ,however no diagnostic challenges or prognosis were mentioned

Diagnostic assessment

In the emergency department, his blood pressure was 176/110 mmHg, heart rate was 94 beats/min, and oxygen saturation was at 94% without oxygen. His-temperature was 39.4 °C. There was no radio-radial or radio-femoral delay but pulsation in his right popliteal artery and dorsalis pedis artery were weak. Lungs were clear to auscultate and there were no added sounds in auscultation of the heart. No neurological finding was elicited on examination. There were no marfanoid features.

Chest X-ray was unremarkable and the electrocardiogram (ECG) showed a sinus tachycardia of 110 beats/min. The echocardiogram had no regional wall motion abnormality and no valvular pathology was recognized. However, there was a suspicion of an aortic dissection flap. His-troponin and creatine kinase-MB levels were normal. Computed tomography of the aorta was planned which showed a Stanford type A dissection going through the length of the thoracic and abdominal aorta into the right common iliac branch of the aorta (Figs. 1 and 2).

Therapeutic Intervention

- 9a: Types of therapeutic intervention (such as pharmacologic, surgical, preventive, self-care)

9b: Administration of therapeutic intervention (such as dosage, strength, duration)

9c: Changes in therapeutic intervention (with rationale)

- Comment: all are fulfilled

Therapeutic intervention

He was started on labetalol infusion at 2 mg/min to control his heart rate and blood pressure. An immediate vascular surgery consultation was called. After a discussion with the heart team, it was decided to move the patient immediately for correction of the defect in the operating room. Cardiac surgeons undertook ascending aortic replacement with aortic arch and abdominal aortic replacement. Comprehensive post-operative treatment measures, including antipyretics, anti-infection, and nutritional support were given. After surgery, he remained febrile for two days despite antipyretics and tazobactam/piperacillin. An infectious disease consultation was done and various tests were performed.

Laboratory evaluation showed an erythrocyte sedimentation rate (ESR) of 102 mm/hour and C-reactive protein 14.54 mg/L. His-complete blood picture, liver, kidney, and thyroid function tests were all within normal limits. Serum electrolytes, amylase, uric acid, and proteins were normal. Serologic tests were negative for tuberculosis, human immunodeficiency virus, hepatitis C virus, hepatitis B virus, syphilis, and cytomegalovirus. Antinuclear antibodies, anti-double stranded DNA, anti-sjogren's syndrome antibodies-A and –B, rheumatoid factor, and anti-cyclic citrullinated were negative. Arterial tissue samples collected by surgeons exhibited macrophage infiltration, presence of monocytes, lymphocytes, and proliferation of blood vessels and connective tissue. There was intimal thickening present on histological examination indicating a chronic inflammatory process (Fig. 3).

Although the history of presentation was typical, the diagnosis of FMF was considered late as it is rarely seen in our region and large vessels are not usually involved in attacks of FMF. He was labeled according to the diagnostic criteria for FMF. His-genetic analysis was sent for known mutations in Marfan's syndrome (FBN-1), and Loeys-Dietz syndrome (TGFβR1, SMAD-3, TGFβ2). Giant cell arteritis was ruled out as all other branches of the aorta appeared normal on computed tomography scan. Genetic analysis showed a mutation in M680I, confirming the diagnosis of FMF. After a consultation from a rheumatologist, he was started on prednisolone 0.5 mg/kg/day eight hourly and colchicine 1.2 mg/day once daily.

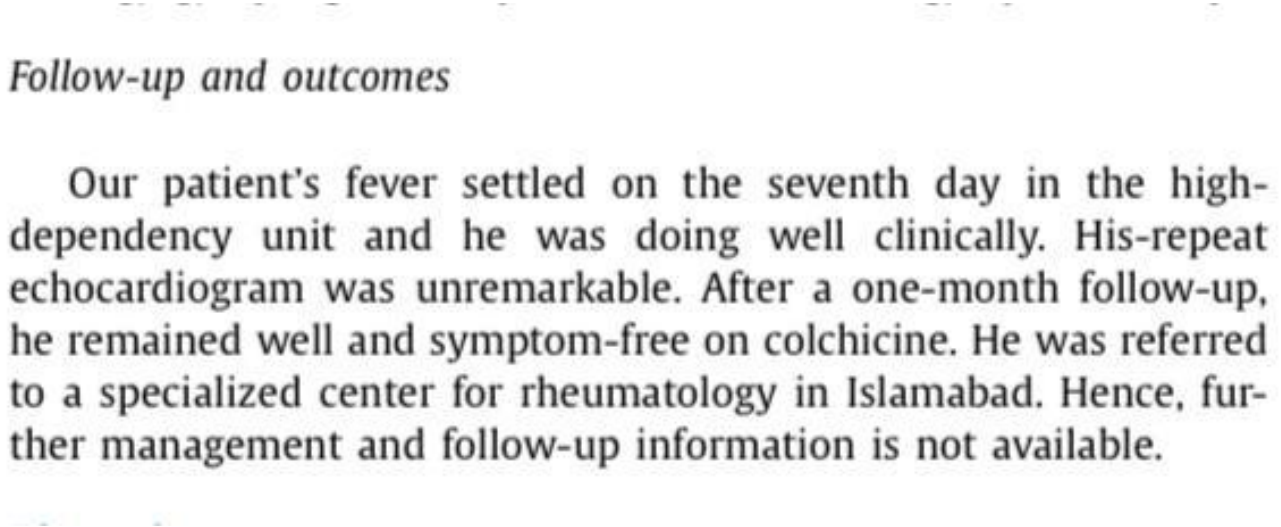
Follow-up and Outcomes

- 10a: Clinician and patient-assessed outcomes (if available)

  10b: Important follow-up diagnostic and other test results

  10c: Intervention adherence and tolerability (How was this assessed?)

  10d: Adverse and unanticipated events
- Comment: no new outcomes were found and the patient was followed up with echocardiogram with unremarkable results. No problems in adherence or tolerability to the intervention were stated or how it was assessed and no adverse or unanticipated events were mentioned



Follow-up and outcomes

Our patient's fever settled on the seventh day in the high-dependency unit and he was doing well clinically. His-repeat echocardiogram was unremarkable. After a one-month follow-up, he remained well and symptom-free on colchicine. He was referred to a specialized center for rheumatology in Islamabad. Hence, further management and follow-up information is not available.

Discussion

- 11a: A scientific discussion of the strengths AND limitations associated with this case report

11b: Discussion of the relevant medical literature with references

11c: The scientific rationale for any conclusions (including assessment of possible causes)

- Comment: fulfilled as ,the key features of the case were discussed also the basic mechanisms (e.g., pathophysiology). Limitations were mentioned as that the results from a single case may not be applicable to patients in general. There was reference to relevant medical literature throughout the discussion and in the last paragraph the rationale was explained

Discussion

Aortic dissection is an age-related life-threatening disease that is associated with high morbidity and mortality. It is characterized by the splitting of the layers within the aortic wall. This way, blood flow is diverted to the medial layers of the aortic wall through the intimal tear. Most widely used is the Stanford classification which distinguishes between type A and B according to the involvement of the ascending aorta [5]. A weakened medial layer of the aorta and a disorder of certain extracellular matrix proteins is the main pathology of dissection. Another risk factor is aging, which causes degeneration of the medial layer. The prevalence of aortic dissection increases rapidly with increasing age [5].

FMF creates an autoinflammatory dysfunction causing small- and medium-vessel vasculitis in various case reports and systematic reviews [4,5]. Periodic inflammatory attacks in FMF cause chronic persistent inflammatory state, leading to endothelial dysfunction, which is attributed to accelerated atherosclerosis. As a consequence, there is increased arterial stiffness, which is an independent risk factor for acute coronary syndromes causing morbidity and potential mortality [6]. Akdoban et al. suggested that there was an increased carotid intima-media thickness in patients presenting with periodic attacks when compared with healthy controls [7]. Similarly, a number of studies has demonstrated an increased aortic wall stiffness in FMF, providing evidence of a potential risk for aortic pathologies in this patient cohort [8].

Our patient had no symptoms of a small vessel or any multiorgan involvement and had only one of the more commonly reported symptoms, which was abdominal pain. Although this began while the patient was 13 years and fulfills a major pediatric criterion for diagnosis of FMF, he had none of the other symptoms required for diagnosis as an adult. This is consistent with a study conducted in Iran that found male gender, age less than 20 years, and abdominal pain as a presenting symptom, to be risk factors for the development of severe attack of FMF, although these patients also tended to respond well to treatment with colchicine [9].

Research by a large Iranian center has revealed that male members of their population can present with clinical manifestations of FMF Type 1 before the age of 20 years. This points to estimated prevalence rates of FMF in the region of Iran and Pakistan to be much higher, as concluded by a genetic epidemiological study that found genetic carriers for FMF to be located as far north in Pakistan as Lahore [10].

Discussion

- 11d: The primary “take-away” lessons of this case report (without references) in a one paragraph conclusion
- Comment: fulfilled

Conclusion

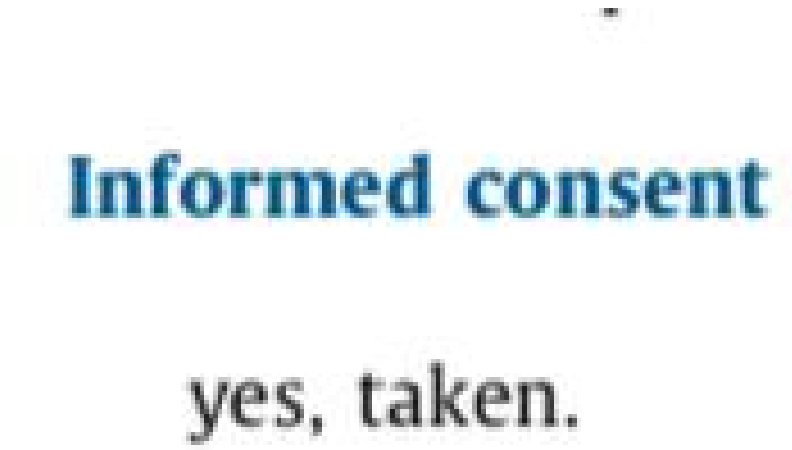
FMF is an autoinflammatory disease with various clinical presentations. It typically involves small- and medium-vessels causing vasculitis and aneurysm formation. However, we present a case with large-vessel involvement, and to the best of our knowledge, this is the first case ever reported of an aortic dissection in a patient with FMF. Clinical suspicion for an aortic dissection in a young patient must take into account underlying pathology and thorough history in order to reach a diagnosis promptly to initiate therapeutic interventions in a timely manner.

Patient Perspective

- 12: The patient should share their perspective in one to two paragraphs on the treatment(s) they received
- comment: not fulfilled

Informed Consent

- 13: Did the patient give informed consent? Please provide if requested
- Comment: fulfilled



suggestions

Patient information:

A 23-year-old man of Iranian descent came to the emergency department of our institute with intermittent epigastric pain.the pain started two weeks ago he took over-the-counter antacids as he ascribed this pain to acid peptic disease. The pain started again while he was sleeping, three hours before coming to the hospital. It was a tearing pain, radiating to the back with extension to the jaw, left arm, and upper abdomen.
The patient has a history of periodic fever since childhood which spiked spontaneously between 37.8 to 38.9 °C and then subsided within a week after taking antipyretics. This fever was associated with myalgias, arthritis of multiple joints, and backache. There was a history of laparotomy 6 years previously, when the patient was 13 years old, with suspicion of acute abdomen. Also there is history of appendectomy done subsequently .

suggestions

Clinical findings:

In the emergency department, his vital signs showed blood pressure was 176/110 mmHg, heart rate was 94 beats/min, and oxygen saturation was at 94% without oxygen. His-temperature was 39.4 °C.
There was no radio-radial or radio-femoral delay but pulsation in his right popliteal artery and dorsalis pedis artery were weak.
Lungs were clear to auscultate and there were no added sounds in auscultation of the heart.
No neurological finding was elicited on examination. There were no marfanoid features.