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ACUTE STANFORD TYPE A AORTIC DISSECTION PRESENTING WITH RESPIRATORY SYMPTOMS AS THE INITIAL MANIFESTATION OF SUSPECTED MARFAN SYNDROME: CASE REPORT

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Abstract

Introduction: Acute Stanford type A aortic dissection is a life-threatening emergency that requires immediate diagnosis and surgical intervention. While the classic presentation involves severe chest or back pain, recognition may be delayed in patients with atypical symptoms, particularly those with underlying connective tissue disorders such as Marfan syndrome.

Case Presentation: A 46-year-old man (height 190 cm) with no prior diagnosis of connective tissue disease presented with a three-day history of fever, cough, and mild chest discomfort, without severe chest, back, or abdominal pain. Physical examination revealed a Marfanoid habitus, and the family history was notable for aortic dissection in several first-degree relatives. Computed tomography angiography of the aorta identified an acute Stanford type A (DeBakey I) dissection originating at the aortic root and extending through the ascending aorta, aortic arch, and descending thoracic aorta, with propagation toward the iliac arteries. Transthoracic echocardiography confirmed the diagnosis by revealing a markedly dilated aortic root (66–68 mm), an intimal flap at the right coronary cusp, and severe aortic regurgitation. The patient underwent urgent surgical repair using a Bentall procedure. The postoperative course was complicated by significant bleeding, requiring surgical re-exploration. Following this intervention, recovery was uneventful. The overall clinical presentation and perioperative findings were most consistent with an underlying diagnosis of Marfan syndrome.

Conclusion: This case underscores the importance of considering aortic dissection in patients with a Marfanoid phenotype and positive family history, even when typical symptoms are absent. Early diagnosis and surgical intervention are critical for improving survival in this high-risk population.

Keywords: *Aortic dissection; Bentall procedure; Marfan syndrome.*

Introduction

Acute type A aortic dissection (ATAAD) is a life-threatening cardiovascular emergency involving the ascending aorta that requires prompt diagnosis and immediate surgical intervention (1,2). Without timely surgical treatment, early mortality increases by approximately 1% to 2% per hour during the first 24 hours, and nearly half of untreated patients die within one week (1). Diagnosis becomes particularly challenging when patients lack the classic symptoms of sudden, severe, tearing chest or back

pain and instead present with nonspecific complaints, which can delay definitive treatment (2–4).

In younger adults, the presence of marfanoid features combined with a strong family history of aortic dissection should raise suspicion for both ATAAD and an underlying heritable thoracic aortic disorder (1,5). However, Marfan syndrome cannot be diagnosed solely on the basis of these features; a comprehensive clinical evaluation using established criteria and, when appropriate, molecular genetic testing is required (5,6). This report describes an extensive ATAAD in a young adult with marfanoid features and a significant family history who presented with fever, cough, dyspnea, and only mild chest discomfort. This atypical, minimally painful presentation initially suggested a predominantly respiratory illness. This case highlights the necessity of considering acute aortic pathology in high-risk patients even when characteristic pain is absent, which can lead to earlier diagnosis and treatment.

Case Presentation

A 46-year-old male patient was admitted to the hospital with a three-day history of fever, cough, dyspnea, and mild chest discomfort. He denied any sudden or severe chest, back, or abdominal pain. His medical history included hypertension, hyperlipidemia, hyperthyroidism, and active tobacco use. Importantly, he had not previously been diagnosed with Marfan syndrome or other connective tissue disorders.

On admission, the patient was hemodynamically stable, with a blood pressure of 104/70 mmHg in the right arm and 115/80 mmHg in the left arm, and a heart rate of 90 beats per minute in sinus rhythm. He was 190 cm tall and weighed 80 kg, exhibiting features suggestive of a marfanoid habitus. Furthermore, the patient reported a family history of aortic dissection affecting his father, mother, and sister; however, the ages of these relatives at the time of dissection were not documented.

Figure 1. Chest radiograph demonstrating enlargement of the aortic arch silhouette (arrow), with no focal pulmonary consolidation.

The initial presentation suggested a respiratory illness, with pneumonia and viral respiratory infection considered in the differential diagnosis. However, chest radiography revealed no focal pulmonary consolidation but demonstrated enlargement of the aortic silhouette (Figure 1). Based on this finding, along with marfanoid features and a significant family history, computed tomography angiography (CTA) of the aorta was performed. CTA revealed a dissection extending from the aortic root through the ascending aorta, aortic arch, descending thoracic aorta, and abdominal aorta to the aortic bifurca-

tion, with further extension into the right iliac artery (Figure 2). These findings were consistent with a Stanford type A, DeBakey type I aortic dissection.

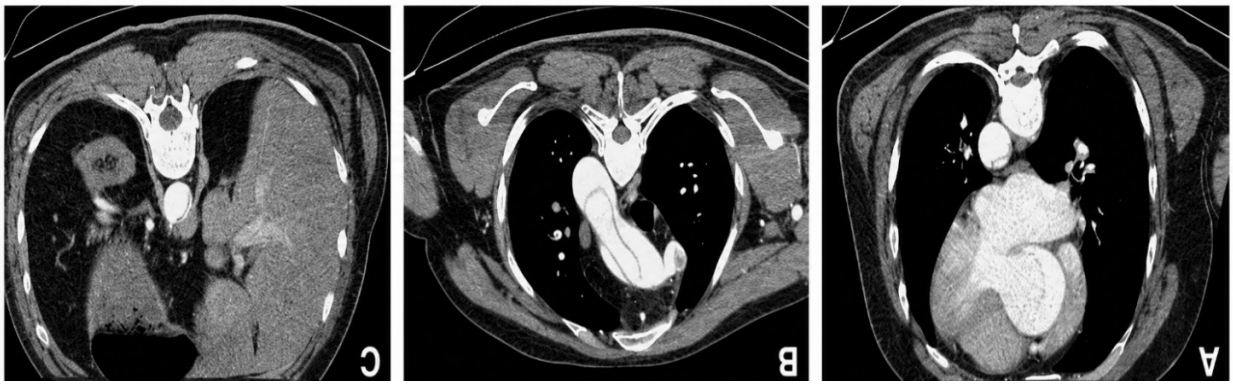

Figure 2. CTA images showing an extensive Stanford type A, DeBakey type I aortic dissection:

(**A**) involvement of the aortic root; (**B**) extension through the ascending aorta and aortic arch;

(**C**) propagation into the abdominal aorta.

Transthoracic echocardiography demonstrated marked dilatation of the proximal aorta and a mobile intimal flap at the level of the right coronary cusp (Figure 3A). The aortic root measured 66 to 68 mm at the sinuses of Valsalva and sinotubular junction, while the ascending aorta measured 66 mm. Severe eccentric aortic regurgitation was present, with a vena contracta width of 0.6 cm (Figure 3B). Despite these findings, left ventricular systolic function was preserved, with an ejection fraction of 62%.

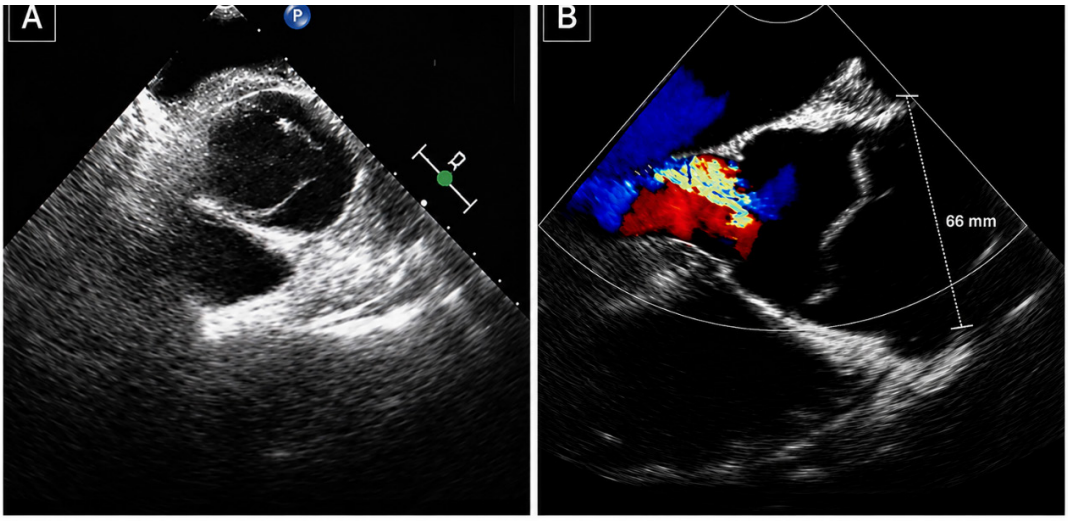

Figure 3. Transthoracic echocardiographic images demonstrating **(A)** an intimal flap at the level of the right coronary cusp and **(B)** marked aortic root dilatation measuring 66 mm, with color Doppler evidence of severe eccentric aortic regurgitation.

The patient subsequently underwent emergency surgical repair. General anesthesia was induced with

etomidate, fentanyl, and rocuronium and maintained with sevoflurane until the initiation of cardiopulmonary bypass, after which anesthesia was maintained with a propofol infusion. Central venous and large-bore venous access were established, alongside invasive femoral arterial pressure monitoring. Cerebral oxygenation was continuously monitored using near-infrared spectroscopy (NIRS), and hemostatic management was guided by rotational thromboelastometry (ROTEM). Cardiopulmonary bypass was established through ascending aortic cannulation and right atrial cannulation with a two-stage venous cannula. A Bentall procedure was performed using a CarboMedics mechanical composite valved conduit comprising a 27-mm mechanical valve and a 30-mm vascular graft (Figure 4). On the first postoperative day, excessive surgical drain output necessitated re-exploration to identify bleeding and surgical hemostasis.

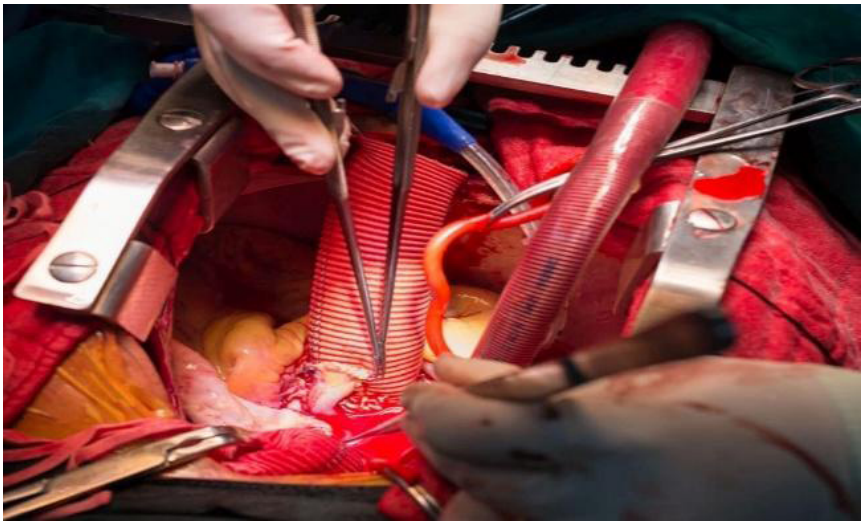

Figure 4. Intraoperative image of aortic root replacement with a mechanical composite valved conduit during the Bentall procedure.

Postoperatively, the patient required temporary mechanical ventilation and vasoactive support but remained in sinus rhythm. He was successfully weaned from mechanical ventilation, and vasoactive therapy was gradually reduced and discontinued. Early mobilization and physical rehabilitation were initiated, and the patient was discharged in stable condition on postoperative day 5 with a prescription for lifelong oral anticoagulation (acenocoumarol) due to the mechanical aortic valve prosthesis.

At follow-up assessments conducted at one week, one month, and six months after surgery, the patient remained clinically stable. Further evaluation was scheduled at one year, followed by annual cardiology assessments and lifelong imaging surveillance of the residual aorta.

The presence of marfanoid features, marked aortic root dilatation, and aortic dissection in three first-degree relatives raised strong suspicion for an underlying heritable thoracic aortic disorder. However, the systemic manifestations required for assessment under the revised Ghent criteria had not been comprehensively documented, so a valid systemic score could not be calculated. Additionally, ophthalmologic evaluation, genetic counseling, and molecular genetic testing had not yet been performed; therefore, Marfan syndrome remained suspected but unconfirmed.

Discussion

Acute type A aortic dissection is a surgical emergency due to the risk of rapid progression to life-threatening complications, including aortic rupture, cardiac tamponade, acute aortic regurgitation, coronary malperfusion, stroke, and organ ischemia (1,2). Current European guidelines recommend immediate surgical intervention upon diagnosis (1), regardless of apparent hemodynamic stability, as significant disease may be present even in stable patients (1,3). In the present case, pronounced proximal aortic dilatation, severe aortic regurgitation, and a dissection extending into the abdominal aorta and right iliac artery signified advanced disease despite initial clinical stability.

Acute aortic dissection typically presents with abrupt, severe chest or back pain (2); however, the absence of typical pain does not exclude the diagnosis. In a population-based study by Smedberg et al., 18% of patients with ATAAD presented without pain, and painless presentation was independently associated with increased 30-day mortality in the overall cohort (4). Data from the International Registry of Acute Aortic Dissection (IRAD) further indicate that painless dissection may instead manifest as syncope, heart failure, or stroke, potentially delaying recognition of the underlying aortic emergency (3). In the present case, the patient experienced only mild chest discomfort and did not report the abrupt, severe pain typically associated with aortic dissection. The predominance of respiratory symptoms and fever initially indicated a possible respiratory or infectious illness. However, the etiology of these symptoms remained unclear, and the possibility of a concurrent respiratory infection could not be excluded. Therefore, these symptoms cannot be definitively attributed to the dissection. Although enlargement of the aortic silhouette on chest radiography was not diagnostic in isolation (1), this finding, together with the patient's marfanoid features and strong family history of aortic dissection, prompted definitive cross-sectional imaging.

CTA is the preferred first-line imaging modality for most hemodynamically stable patients with suspected acute aortic dissection, as it is rapid, widely accessible, and enables comprehensive evaluation of the entire aorta, including the delineation of true and false lumina, identification of the entry tear, assessment of branch-vessel involvement, and detection of major complications (1,2). In the present case, CTA played a pivotal role in establishing the diagnosis, revealing a DeBakey type I dissection extending from the aortic root to the aortic bifurcation and into the right iliac artery. Transthoracic echocardiography provided complementary information by visualizing the proximal intimal flap, quantifying aortic root and ascending aortic dilatation, demonstrating severe eccentric aortic regurgitation, and assessing left ventricular systolic function. The combined use of CTA and echocardiography was therefore essential for detailed anatomical characterization and precise surgical planning.

The patient underwent emergency surgical repair, as the dissection involved the ascending aorta (1,2). The presence of a markedly aneurysmal aortic root measuring 66–68 mm, together with severe aortic regurgitation, provided a clear anatomical rationale for composite aortic root replacement (1). In this case, a Bentall procedure was performed, addressing both the diseased aortic root and the aortic valve in a single operation. Although valve-sparing root replacement may be considered in selected patients, particularly when the valve cusps are suitable for repair and the procedure is performed by an experienced aortic surgical team (1), the substantial root dilatation and severity of aortic regurgitation in this emergency context favored composite root replacement. The implantation of a mechanical valved conduit consequently necessitated lifelong anticoagulation with acenocoumarol (Sintrom).

The patient's relatively young age, marfanoid features, marked aortic root dilatation, and a family

history of aortic dissection in three first-degree relatives strongly suggested an underlying heritable thoracic aortic disorder (1,5). However, neither the presence of marfanoid habitus nor a family history of aortic dissection is, on its own, sufficient to diagnose Marfan syndrome (5,6). According to the revised Ghent criteria, a diagnosis of Marfan syndrome can be established based on specific combinations of aortic root dilatation or dissection, and additional features such as ectopia lentis, a pathogenic FBN1 mutation, a systemic score of ≥7, or a family history of confirmed Marfan syndrome (5,6). In this patient, the lack of comprehensive documentation of systemic features and the absence of both ophthalmologic evaluation and molecular genetic testing precluded a definitive diagnosis, leaving Marfan syndrome suspected but unconfirmed. Accordingly, comprehensive phenotypic assessment, ophthalmological examination, genetic counseling, and molecular testing are recommended to clarify the diagnosis and guide family risk assessment. First-degree relatives should undergo screening aortic imaging, with cascade genetic testing if a pathogenic or likely pathogenic variant is identified (1,5).

The dissection extended beyond the surgically replaced segment, leaving the residual aorta at ongoing risk of progressive dilatation, aneurysmal degeneration, and potentially requiring future reintervention. Therefore, strict blood pressure control and lifelong cross-sectional imaging surveillance of the entire aorta are essential components of long-term management, regardless of apparent clinical stability (1).

Conclusion

This case illustrates that acute type A aortic dissection may present atypically in relatively young patients with features suggesting heritable aortic disease, even in the absence of characteristic severe pain. Early identification of subtle radiographic findings, together with consideration of phenotypic and family history, is critical for timely diagnosis and intervention. Clinicians should maintain a high level of suspicion in at-risk individuals and ensure ongoing assessment of both patients and their first-degree relatives to improve clinical outcomes.

Declarations

Ethics approval and consent to participate: A formal approval of the Ethics Committee was not required for this study.

Informed consent: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of interest: The authors declare no conflict of interest.

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Author contribution statement: All authors contributed to the conception of the case report, data collection, manuscript preparation, and approved the final version of the manuscript.

Artificial Intelligence (AI) statement: AI tools were used for language editing. The authors take full responsibility for the content.

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