

## > THE PATIENT

20-year-old man

## > SIGNS & SYMPTOMS

- Sudden-onset chest pain
- Worsening pain with cough and exertion
- No improvement with naproxen

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## > THE CASE

A 20-year-old man presented to our clinic with a 3-day history of nonradiating chest pain located at the center of his chest. Past medical history included idiopathic neonatal giant-cell hepatitis and subsequent liver transplant at 1 month of age; he had been followed by the transplant team without rejection or infection and was in otherwise good health prior to the chest pain.

On the day of symptom onset, he was walking inside his house and fell to his knees with a chest pain described as “a punch” to the center of the chest that lasted for a few seconds. He was able to continue his daily activities without limitation despite a constant, squeezing, centrally located chest pain. The pain worsened with cough and exertion.

A few hours later, he went to an urgent care center for evaluation. There, he reported, his chest radiograph and electrocardiogram (EKG) results were normal and he was given a diagnosis of musculoskeletal chest pain. Over the next 3 days, his chest pain persisted but did not worsen. He was taking 500 mg of naproxen every 8 hours with no improvement. No other acute or chronic medications were being taken. He had no significant family history. A review of systems was otherwise negative.

On physical exam, his vital statistics included a height of 6'4"; weight, 261 lb; body mass index, 31.8; temperature, 98.7 °F; blood pressure, 134/77 mm Hg; heart rate, 92 beats/min; respiratory rate, 18 breaths/min; and oxygen saturation, 96%. Throughout the exam, he demonstrated no acute distress, appeared well, and was talkative; however, he reported having a “constant, squeezing” chest pain that did not worsen with palpation of the chest. The rest of his physical exam was unremarkable.

Although he reported that his EKG and chest radiograph were normal 3 days prior, repeat chest radiograph and EKG were ordered due to his unexplained, active chest pain and the lack of immediate access to the prior results.

## THE DIAGNOSIS

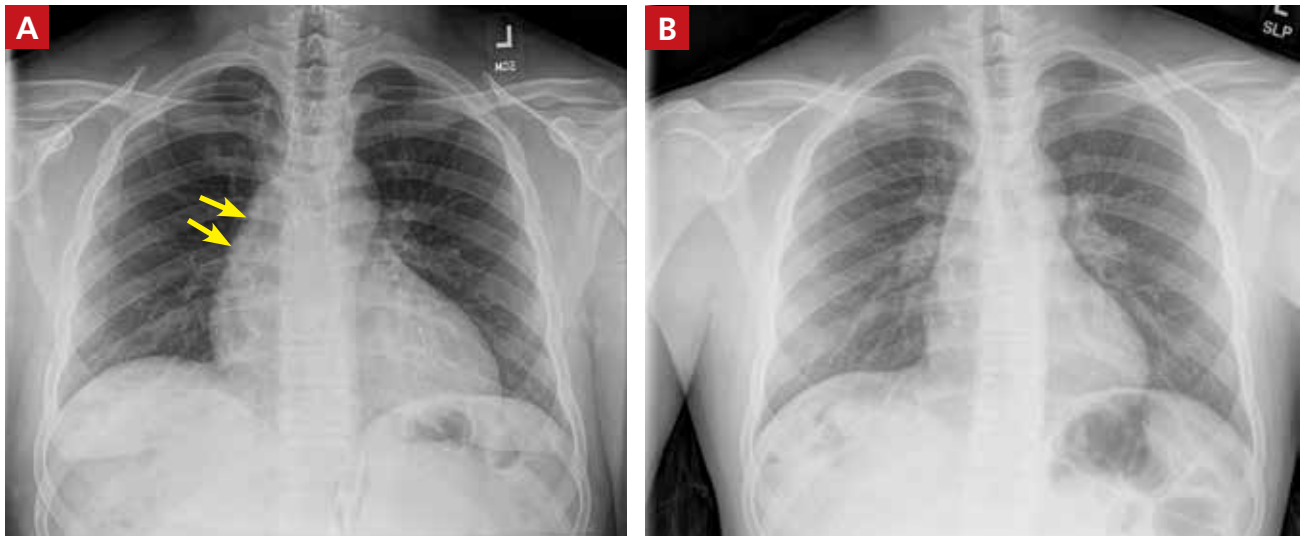
The chest radiograph (FIGURE 1A) showed a “mildly ectatic ascending thoracic aorta” that had increased since a chest radiograph from 6 years prior (FIGURE 1B) and “was concerning for an aneurysm.” Computed tomography (CT) angiography (FIGURE 2) then confirmed a 7-cm aneurysm of the ascending aorta, with findings suggestive of a retrograde ascending aortic dissection.

## DISCUSSION

The average age of a patient with acute aortic dissection (AAD) is 63 years; only 7% occur in people younger than 40.<sup>1</sup> AAD is often accompanied by a predisposing risk factor such

FIGURE 1

## Imaging was ‘concerning for aneurysm’



A chest radiograph showed increasing ectasia of the ascending thoracic aorta (A), which had not been present on a radiograph taken 6 years prior and used for comparison (B).

as a connective tissue disease, bicuspid aortic valve, longstanding hypertension, trauma, or larger aortic dimensions.<sup>2,3</sup> Younger patients are more likely to have predisposing risk factors of Marfan syndrome, prior aortic surgery, or a bicuspid aortic valve.<sup>3</sup>

A literature review did not reveal any known correlation between the patient's history of giant-cell hepatitis or antirejection therapy with thoracic aortic dissection. Furthermore, liver transplant is not known to be a specific risk factor for AAD in pediatric patients or outside the immediate postoperative period. Therefore, there were no known predisposing risk factors for AAD in our patient.

■ **The most common clinical feature of AAD** is chest pain, which occurs in 75% of patients.<sup>1</sup> Other clinical symptoms include hypertension and diaphoresis.<sup>2,4</sup> However, classic clinical findings are not always displayed, making the diagnosis difficult.<sup>2,4</sup> The classical description of “tearing pain” is seen in only 51% of patients, and 5% to 15% of patients present without any pain.<sup>1</sup>

■ **Commonly missed or misdiagnosed.** The diagnosis of AAD has been missed during the initial exam in 38% of patients.<sup>4</sup> As seen in our case, symptoms may be initially diag-

nosed as musculoskeletal chest pain. Based on symptoms, AAD can be incorrectly diagnosed as an acute myocardial infarction or vascular embolization.<sup>2,4</sup>

Every hour after symptom onset, the mortality rate of untreated AAD increases 1% to 2%, with no difference based on age.<sup>3,4</sup> Different reports have shown mortality rates between 7% and 30%.<sup>4</sup>

■ **Effective imaging is crucial** to the diagnosis and treatment of AAD, given the occurrence of atypical presentation, missed diagnosis, and high mortality rate.<sup>4</sup> A chest radiograph will show a widened mediastinum, but the preferred diagnostic tests are a CT or transthoracic echocardiogram.<sup>2,4</sup> Once the diagnosis of AAD is confirmed, an aortic angiogram is the preferred test to determine the extent of the dissection prior to surgical treatment.<sup>2</sup>

■ **Classification dictates treatment.** AAD is classified based on where the dissection of the aorta occurs. If the dissection involves the ascending aorta, it is classified as a type A AAD and should immediately be treated with emergent surgery in order to prevent complications including myocardial infarction, cardiac tamponade, and aortic rupture.<sup>2,4,5</sup> If the

FIGURE 2

## CT angiography confirms the Dx



Computed tomography angiography revealed a 7-cm aneurysm of the ascending aorta.

dissection is limited to the descending aorta, it is classified as a type B AAD and can be medically managed by controlling pain and lowering blood pressure; if symptoms persist, surgical management may be required.<sup>2</sup> After hospital discharge, AAD patients are followed closely with medical therapy, serial imaging, and reoperation if necessary.<sup>4</sup>

■ **Our patient** underwent emergent surgery for aortic root/ascending aortic replacement with a mechanical valve. He tolerated

the procedure well. Surgical tissue pathology of the aortic segment showed a wall of elastic vessel with medial degeneration and dissection, and the tissue pathology of the aorta leaflets showed valvular tissue with myxoid degeneration.

### THE TAKEAWAY

It is critical to keep AAD in the differential diagnosis of a patient presenting with acute onset of chest pain, as AAD often has an atypical presentation and can easily be misdiagnosed. Effective imaging is crucial to diagnosis, and immediate treatment is essential to patient survival. **JFP**

#### CORRESPONDENCE

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