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Diffuse Large B-Cell Lymphoma Mimicking Intramural Hematoma of the Thoracic Aorta

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ABSTRACT

Diffuse large B-cell lymphoma (DLBCL) is an aggressive lymphoma that is fatal if left untreated. Few cases have been reported of involvement of the aorta. Here we present a case of DLBCL that was diagnosed by periaortic computed tomography–guided biopsy. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2023;15:101858) © 2023 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 73-year-old man with a complex past medical history notable for recent thoracic endovascular aortic repair (TEVAR) for presumed intramural hematoma (IMH) in the descending thoracic aorta presented to our facility (University Hospitals Cleveland Medical Center, Cleveland, Ohio, USA) as a new patient with recurrent chest pain and bilateral flank pain. Chest symptoms started suddenly while he was working in his yard, and soon afterward he developed flank pain. When conservative management, including rest and over-the-counter pain medications, failed to bring relief, the patient presented to the nearest emergency department for formal evaluation. Chest pain was reported as mild, intermittent, right sided, and nonpleuritic. Review of systems was significant for a 9-lb weight loss over the last 2 months. On presentation, his vital signs were normal (blood pressure, 115/65 mm Hg; pulse, 89 beats/min), and oxygen saturation was normal on room air. Physical examination demonstrated equal radial, dorsalis pedis, and posterior tibial pulses bilaterally.

LEARNING OBJECTIVES

- To be able to formulate a differential diagnosis of periaortic fluid collections with CT.
- To understand the management of DLBCL and the fatality of the disease when untreated.

PAST MEDICAL HISTORY

The patient had a past medical history of hypothyroidism (thyroid stimulating hormone, 2.48 mIU/L) treated with levothyroxine, and hypertension treated with lisinopril. Three months earlier, the patient presented to a different hospital with sharp, sudden onset, severe chest pain, at which time he was ultimately given a diagnosis of aortic IMH. He subsequently underwent TEVAR with placement of a Bolton Relay thoracic stent. The procedure was uncomplicated, and the patient was discharged safely home without issue.

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ABBREVIATIONS AND ACRONYMS

CT = computed tomography

DLBCL = diffuse large B-cell lymphoma

IMH = intramural hematoma

PET = positron emission tomography

TEVAR = thoracic endovascular aortic repair

DIFFERENTIAL DIAGNOSIS

The patient's presentation with chest pain and flank pain in the setting of recent TEVAR placement raised concern for stent graft infection, type 1 endoleak, extension of known aortic disease, acute coronary syndrome, and noncardiac chest pain.[1-4]

INVESTIGATIONS

Given his recent TEVAR 3 months earlier, a computed tomography (CT) angiogram of the chest, abdomen, and pelvis was done. The scan revealed a periaortic fluid collection extending from level of the T8 to T11 vertebral bodies, with no active extravasation of contrast material (**Figure 1**). Cardiac surgery admitted the patient to the hospital for management, and an extensive infectious work-up was completed and found to be unrevealing, with negative blood culture results and an absence of leukocytosis or other inflammatory markers. A positron emission tomography (PET) scan revealed a hypermetabolic periaortic bulky mass surrounding the majority of the adjacent aorta, engulfing the adjacent spine at the T8 through T11 vertebral bodies and extending into the spinal canal (**Figures 2 and 3**). The patient underwent CT-guided biopsy of the periaortic soft tissue, and findings were consistent with diffuse large B-cell lymphoma (DLBCL). Pathologic examination further characterized his malignant disease as DLBCL, non-germinal center subtype (CD10[-], BCL-6[-], MUM1[+]).

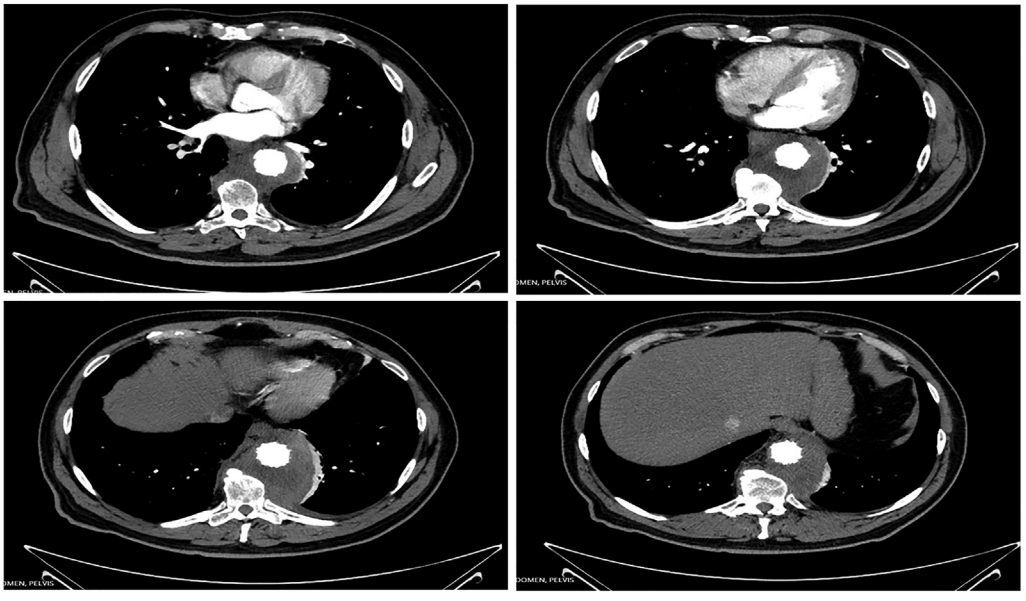
MANAGEMENT

The patient was referred to oncology, with expedited induction of chemotherapy (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisone). He tolerated 6 cycles of treatment without any major complications. A follow-up PET scan 6 months later revealed metabolic resolution of the previously paraspinal mass and periaortic lymphadenopathy, with no evidence of hypermetabolic extranodal disease or distal lymphomatous involvement (**Figures 4 and 5**).

DISCUSSION

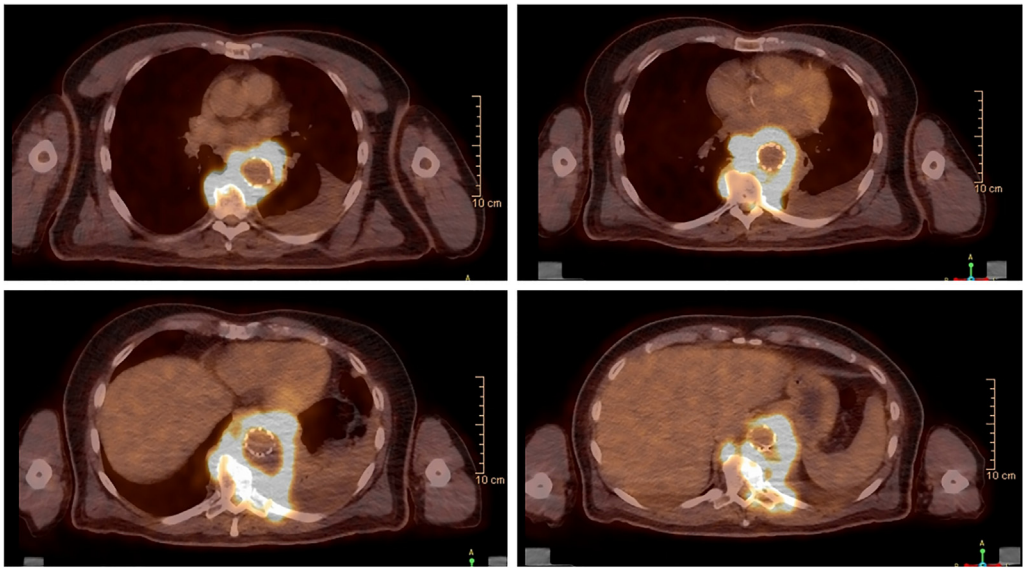
Acute aortic syndromes (acute aortic dissection, IMH, and penetrating aortic ulcer) are life-threatening conditions that require prompt, accurate diagnosis. Unless aortic rupture has occurred, patients are typically hypertensive and tachycardic, and they report sudden onset of severe pain involving the chest, back, abdomen, and/or lower extremities, depending on the location and extent of the aortic

FIGURE 1 Computed Tomography Angiogram at Initial Presentation



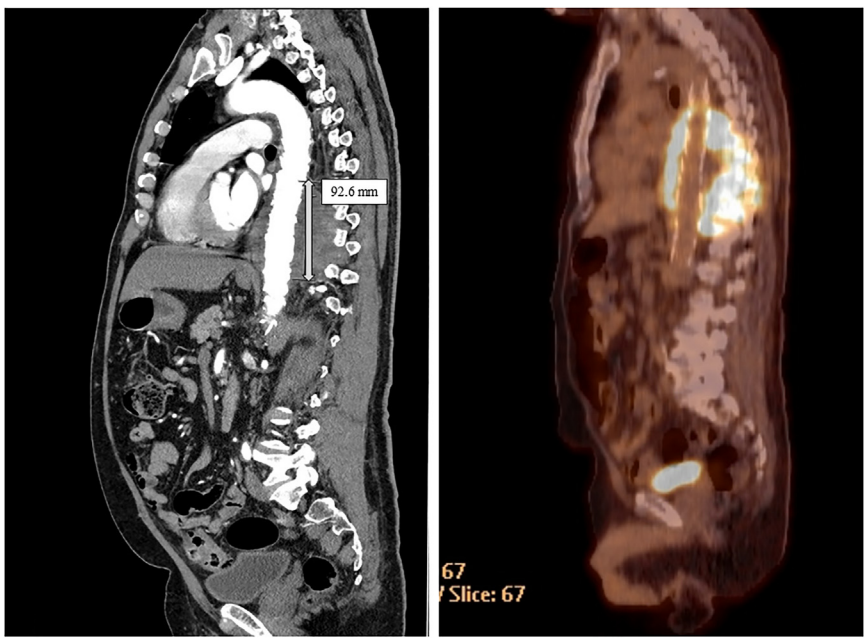
Axial views of periaortic lymphoma at levels T8 to T11, with an appearance "mimicking" that of a contained aortic rupture or hematoma.

FIGURE 2 **Initial Positron Emission Tomography Staging Scan**



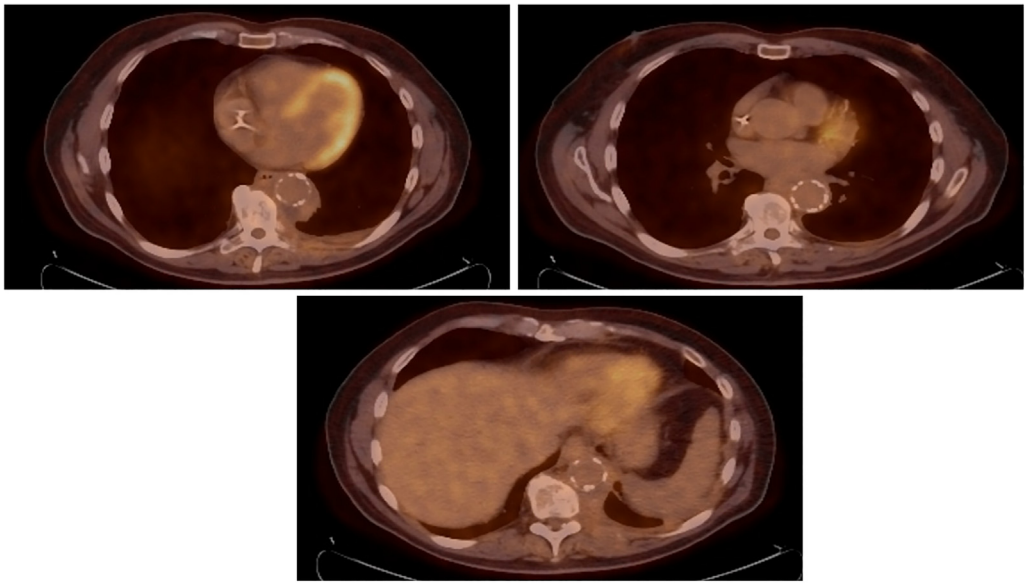
Pretreatment scan, axial views from T8 to T11.The mass is seen surrounding the aorta and with extension to the spine noted.

FIGURE 3 **Initial Positron Emission Tomography Staging Scan With Comparison With Computed Tomography Angiography Scan**



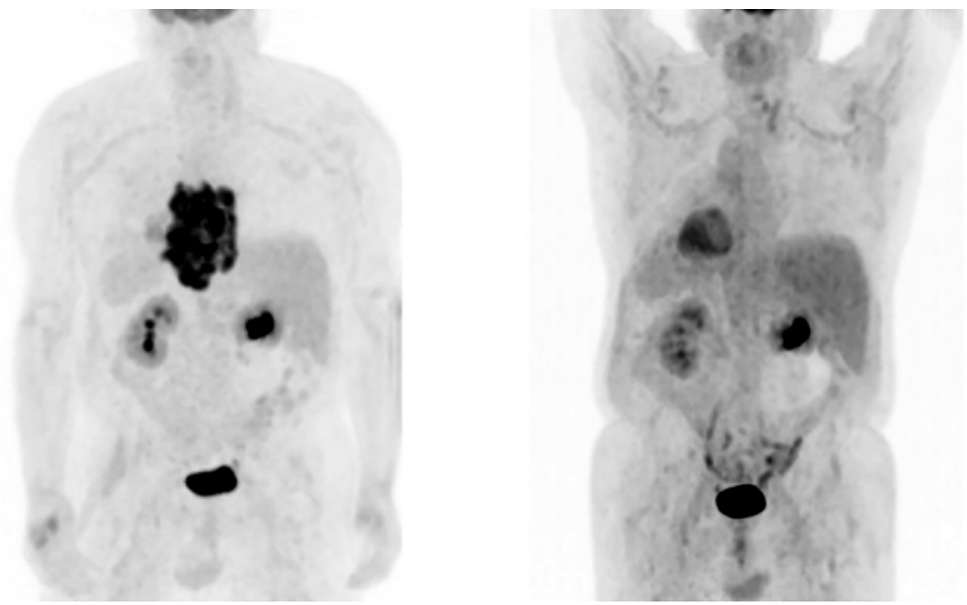
Pretreatment scans, sagittal view. The tumor measures approximately 92.6 mm in length. **(Left)** Computed tomography angiography. **(Right)** Positron emission tomography.

FIGURE 4 Follow-Up Positron Emission Tomography Scan



Axial views 6 months post-treatment with resolution of active disease surrounding the aorta. Noted hypermetabolic activity is now absent.

FIGURE 5 Comparison Positron Emission Tomography Scans Pre- and Post-Treatment



(Left) Pretreatment scan. **(Right)** Post-treatment scan.

disease. In the correct clinical context, health care providers should maintain a high index of suspicion for acute aortic syndromes and initiate the appropriate work-up, which usually includes electrocardiogram-gated high-resolution CT or magnetic resonance angiography.[1] Once the diagnosis is confirmed, the standard of care includes aggressive management to achieve strict impulse control, often followed by open or endovascular repair.[2-5]

We present a unique case of DLBCL that masqueraded as IMH. Three months earlier, the patient presented to an outside hospital in the usual fashion for acute aortic syndrome, at which time he was found to have radiographic findings suggestive of IMH. The patient underwent successful TEVAR and was discharged home. Over the subsequent months, the patient experienced unintentional weight loss, and he slowly developed chest symptoms and flank pain that led him to present to the hospital again. When advanced imaging of the chest failed to show evidence of an endoleak, and the result of an extensive infectious disease work-up was negative, the cause of the periaortic fluid collection came into question. On careful review of his historical records, it was noted that the fluid collection had grown considerably over the intervening months, and it demonstrated evidence of direct extension into surrounding tissue as it grew posteriorly toward the spine. This change guided the decision to pursue PET imaging and then biopsy, which confirmed the diagnosis of DLBCL.

DLBCL comprises approximately 30% of non-Hodgkin's lymphoma cases in the United States, and it typically arises from either nodal or extranodal lymphatic tissue.[6] Because of the aggressive nature of DLBCL, prompt diagnosis is important for initiation of lifesaving treatment. The patient's history of unintentional weight loss was an additional clue to malignancy in this case. We treated the patient with anthracycline-based therapy, consisting of cyclophosphamide, doxorubicin, vincristine, and prednisone in addition to rituximab[6,7]; resolution of disease occurred after 6 cycles of treatment. The finding of this report demonstrates the importance of considering DLBCL as a diagnosis in patients with large, persistent periaortic collections that resemble hematoma. Although there have been a few cases of DLBCL diagnosed incidentally from aortic tissue sampled during surgery, this is the first case to our knowledge of periaortic DLBCL reported with imaging findings similar to those of IMH.[8-10]

FOLLOW-UP

At his 10-month follow-up, the patient had CT imaging with no evidence of recurrent disease in the chest, abdomen, or pelvis. The patient denied any chest pain or back pain at this follow-up visit. The patient will undergo continued surveillance imaging with oncology to monitor for disease recurrence.

CONCLUSIONS

We present a novel case of DLBCL with an appearance of IMH of the thoracic aorta. DLBCL is an important diagnosis to consider in patients with persistent periaortic collections. Once DLBCL is identified, it is vital to have prompt initiation of chemotherapy of this clinically aggressive malignant disease.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS aortic syndromes, diffuse large B-cell lymphoma