Anticoagulation Management in Loeys-Dietz Syndrome with Atrial Fibrillation

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Introduction

- Loeys-Dietz syndrome (LDS): Rare autosomal dominant connective tissue disorder
- Heterozygous mutations along the transforming growth factor-beta (TGFβ) signaling pathway
- Associated with multiple vasculopathies, skeletal abnormalities, and other systemic manifestations
- Increased risk for thromboembolic events in connective tissue disorders without evidence-based guidelines for anticoagulation

Clinical Presentation

66-year-old male with a PMHx significant for LDS, Thoracic Aortic Aneurysm repair, Aortic Valve repair, HTN, BPH, and Atrial Fibrillation s/P PM presented with epigastric pain, nausea, vomiting, and bloody diarrhea

Infographics

<table>
<thead>
<tr>
<th>Letter</th>
<th>Risk Factor</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>C</td>
<td>Congestive Heart Failure/LV dysfunction</td>
<td>1</td>
</tr>
<tr>
<td>H</td>
<td>Hypertension</td>
<td>1</td>
</tr>
<tr>
<td>A_2</td>
<td>Age ≥ 75</td>
<td>2</td>
</tr>
<tr>
<td>D</td>
<td>Diabetes mellitus</td>
<td>1</td>
</tr>
<tr>
<td>S_2</td>
<td>Stroke/TIA/thromboembolism</td>
<td>2</td>
</tr>
<tr>
<td>V</td>
<td>Vascular disease*</td>
<td>1</td>
</tr>
<tr>
<td>A</td>
<td>Age 65-74</td>
<td>1</td>
</tr>
<tr>
<td>Sc</td>
<td>Sex category (i.e., female sex)</td>
<td>1</td>
</tr>
</tbody>
</table>

CHA₂DS₂-VASc Score: Score; LV dysfunction means LV EF ≤ 40%. Hypertension includes the patient's current antihypertensive medication. *Prior myocardial infarction, peripheral vascular disease, chronic kidney disease, atrial fibrillation

Discussion

- Anticoagulation was held in the setting of a suspected GI bleed
- After a GI bleed was ruled out and the patient was subsequently treated, a clinical conundrum was present at the time of discharge
- While our patient’s chronic atrial fibrillation is managed with oral anticoagulation, dosing and regimen seem to be unaffected by the underlying rare connective tissue disorder of LDS
- This syndrome often presents with damaged blood vessels, vascular fragility, altered blood flow, arterial aneurysms and dissections, and inflammation which - in totality - activates the clotting cascade, increasing the risk of thrombosis
- Review of literature reveals that this is not limited to the likes of LDS but also habitually characterizes other connective tissue disorders
- As such, this clinical picture raises concerns regarding anticoagulation management of connective tissue disorders, especially in the setting of a thoroughly-studied chronic condition, like that of atrial fibrillation
- Currently, there is a lack of consensus on the use of anticoagulation for prophylaxis of thromboembolic events in connective tissue disorders, let alone those with concomitant atrial fibrillation
- Thus, management guidelines should be developed in those with varying forms of CTD, especially in the accomplishment of cardiovascular risk factors, provided that the potential benefits outweigh bleeding risks

Clinical Pearls

- LDS, among other connective tissue disorders, increases the risk for thromboembolic events
- The CHA₂DS₂-VASc Score identifies a need for anticoagulation but does not qualify connective tissue disorders as a risk factor
- The lack of guidelines for connective tissue disorders, let alone those with concomitant atrial fibrillation, leaves uncertainty about optimal anticoagulation management

References


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