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CASE REPORT

Unicuspid aortic valve with false aortic dissection appearance: A case report

Uniküspit aort kapak ve yalancı aort diseksiyonu görünümü: Vaka bildirimi

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Summary– Unicuspid aortic valve (UAV) is a rare congenital anomaly that usually presents with aortic stenosis or mixed stenosis and regurgitation early in life. Ascending aortic aneurysm and aortic dissection are important complications of UAVs. A 27-year-old man presented to the emergency department with a complaint of acute chest pain. Bedside transthoracic echocardiography (TTE) showed dilatation of ascending aorta (47 mm) and mild aortic regurgitation; computed tomography (CT) angiography revealed a suspicious dissection flap within ascending aorta. A cardiovascular surgeon, a radiologist, and a cardiologist were immediately consulted. TTE performed by the cardiologist revealed a unicuspid unicommissural aortic valve and dilated ascending aorta with no signs of dissection. Aortic dissection image on CT angiogram was interpreted by an experienced radiologist and the cardiovascular surgeon as superior pericardial recess and considered as a false-positive dissection image. Given the patient was pain-free, the CT image was considered false positive and as TTE clearly visualized the ascending aorta, the heart team decided that no further imaging is required. After excluding acute aortic syndrome, acute coronary syndrome, and other causes of acute chest pain, the patient was discharged with close follow-up. Diagnosis of aortic dissection is based on noninvasive imaging modalities, and CT is the first-line imaging choice in most emergency departments. Depending on a single imaging modality may cause false interpretations and lead to unnecessary surgical explorations.

Özet– Uniküspit aort kapak, genellikle erken yaşlarda aort darlığı veya kombine aort darlığı ve yetersizliği şeklinde prezente olan nadir bir konjenital anomalidir. Asendan aort anevrizması ve aort diseksiyonu uniküspit aort kapağın önemli komplikasyonlarındandır. 27 yaşında erkek hasta acil servise yeni başlayan göğüs ağrısıyla başvurdu. Yatakbaşı transtorasik ekokardiyografi (TTE) ile asendan aort genişlemesi (47mm) ve hafif aort yetersizliği görüldü, ardından yapılan bilgisayarlı tomografik (BT) anjiyografi ile asendan aortada şüpheli diseksiyon flepi izlendi. Kalp damar cerrahı, radyolog ve kardiyolog acil olarak konsülte edildi. Kardiyolog tarafından yapılan TTE sonucu uniküspit unikomissural aort kapak görüldü, asendan aorta dilate izlendi fakat diseksiyon bulgusu saptanmadı. BT görüntüsündeki şüpheli diseksiyon hattı deneyimli radyolog ve kalp damar cerrahı tarafından perikart katlantısı olarak yorumlandı ve yalancı diseksiyon görüntüsü olarak değerlendirildi. Göğüs ağrısı geçen, BT görüntüsü yalancı pozitif olarak yorumlanan ve TTE'de asendan aort net olarak değerlendirilen hastada kalp ekibi tarafından ek görüntülemeye ihtiyaç duyulmadı. Akut aortik sendrom, akut koroner sendrom ve akut başlangıçlı göğüs ağrısının diğer sebepleri dışlanan hasta yakın takip önerilerek taburcu edildi. Aort diseksiyonu tanısı non-invaziv görüntüleme yöntemleri ile konur ve BT çoğu acil serviste ilk tercih edilen görüntüleme yöntemidir. Tek bir görüntüleme yöntemine bağlı kalmak yanlış yorumlara ve dolayısıyla gereksiz cerrahi müdahalelere yol açabilmektedir.

Unicuspid aortic valve (UAV) is a rare congenital malformation. UAVs usually present with aortic stenosis or mixed stenosis and regurgitation.[1] Valvular dysfunction, dystrophic calcification, aortic dilatation, and aortic dissection are common features of both UAV and bicuspid aortic valve (BAV).[1] As with aortic valve abnormalities, diagnosis of aortic dissection is based on noninvasive imaging modalities: transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), and computed tomography (CT). Relying solely on 1 imaging modality may cause false interpretations and lead to unnecessary surgical explorations, resulting in increased morbidity and mortality.

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CASE REPORT

A 27-year-old male patient presented to the emergency department with sudden onset, stabbing, sharp, and midsternal chest and back pain. He had no prior medical history but had a family history of sudden cardiac death. He was an active smoker for 5 years. His physical examination was positive for a systolic ejection murmur 3/6 heard at right sternal border. His electrocardiography (ECG) was normal, and chest X-ray showed dilatation of aorta. There was no significant finding in routine laboratory workup; hs-Troponin and d-Dimer were also negative.

Bedside TTE (Toshiba Artida Medical System) performed in the emergency department showed normal left ventricular function, moderate aortic stenosis, and dilatation of ascending aorta with a diameter of 48 mm. No intimal flap or dissection image was observed. A thoracoabdominal CT angiogram was performed because of chest pain, back pain, and aortic dilatation. CT angiogram revealed suspicious aortic dissection within ascending aorta (Fig. 1). Because of the discrepancy between 2 imaging modalities, a radiologist, a cardiovascular surgeon, and a senior cardiologist were immediately consulted before any surgical decision was made. TTE performed using an advanced echocardiography device (Philips EPIQ 7C; Philips Medical Systems, Andover, MA, USA) revealed a unicuspid and unicommissural aortic valve. Ascending aorta was dilated, and no signs of dissection were observed (Fig. 2A). Parasternal-short-axis images showed a single commissural zone of attachment and eccentric valvular orifice during systole (Fig. 2B). Echocardiography confirmed moderate aortic stenosis with a peak/mean gradient of 45/28 mm Hg across the aortic valve and mild aortic regurgitation. Patient's left ventricular function was preserved; left ventricular systolic and diastolic diameters were in normal range. Three-dimensional (3D) TTE confirmed unicuspid unicommissural aortic valve (Video 1*). Presence of the optimal images obtained by advanced TTE with both 2D and 3D views provided us certainty of the absence of aortic dissection.

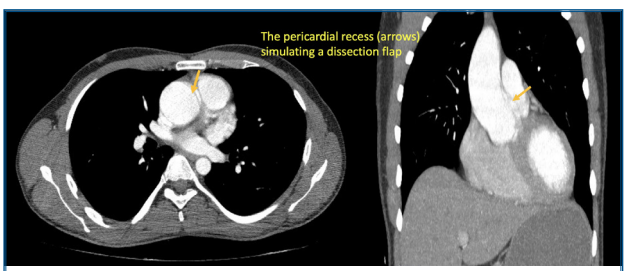

Figure 1. Computed tomography angiography showing the pericardial recess simulating a dissection flap.

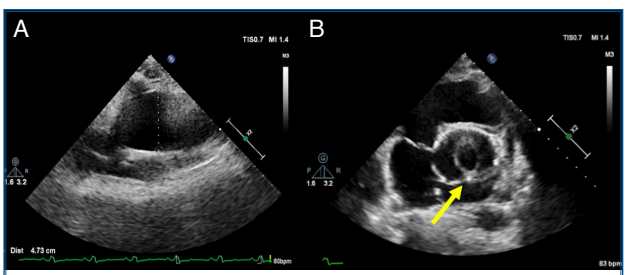

Figure 2. (A) Transthoracic echocardiography showing dilated ascending aorta; no signs of dissection were observed. **(B)** Transthoracic echocardiography showing single commissural zone of attachment and eccentric valvular orifice during systole.

Abbreviations:

3D	*Three-dimensional*
BAV	*Bicuspid aortic valve*
CRP	*C-reactive protein*
CT	*Computed tomography*
ECG	*Electrocardiography*
TEE	*Transesophageal echocardiography*
TTE	*Transthoracic echocardiography*
UAV	*Unicuspid aortic valve*

Aortic dissection image on CT angiogram was interpreted by an experienced radiologist and a cardiovascular surgeon, as superior pericardial recess and considered as a false-positive dissection image.

The patient was followed in the emergency department, and further diagnostic work such as troponin, d-Dimer, repeated ECG, control chest X-ray, and C-reactive protein (CRP) level were obtained. Control chest X-ray, CT scan, and CRP level were negative for the diagnosis of pulmonary embolism, pneumonia, and pneumothorax. Acute coronary syndrome was also excluded after cardiac troponin and ECG follow-up. After excluding possible cardiovascular and pulmonary etiologies, we had a clear diagnosis of noncardiac chest pain. Patient was discharged with close follow-up and medical advice.

DISCUSSION

UAV is a rare congenital malformation with an estimated incidence of approximately 0.02% in patients referred for echocardiography.[2] Two subtypes of UAV have been described: pinhole-shaped acommissural and slit-shaped unicommissural. Unlike acommissural valves, unicommissural valves are usually asymptomatic early in life.

Diagnosis of UAV and differentiating it from BAV can sometimes be challenging. Thus, modalities such as 3D TTE and TEE and, for associated complications, cardiac CT and magnetic resonance imaging might be needed. Several echocardiographic criteria have been defined for differentiating UAV from BAV.[3] Those are single commissural zone of attachment, rounded leaflet-free edge on opposite side of the commissural attachment zone, and eccentric valvular orifice during systole. As in our case, TTE imaging was capable to diagnose UAV, and no further imaging required.

As with BAV, ascending aortic aneurysm is an important associated complication of the UAV.[1] Younger patients tend to present with more aggressive forms of UAV with aortic dilatation.[4] Risk of aortic dissection in UAV is 5- to 9-fold higher than in normal population.[5]

CT angiography is the first-line imaging modality for aortic dissection in most emergency departments, with sensitivity and specificity of 95% and 87%-100%, respectively.[6] Although CT scan is a low-cost and readily available imaging modality in emergency departments, it also has several pitfalls in the diagnosis of acute aortic syndrome.[7] These are the left innominate vein, the left pulmonary vein, streak artifacts simulating an intimal flap, the superior pericardial recess, and motion artifacts. The superior pericardial recess, which is the extension of pericardium over ascending aorta, created a diagnostic confusion in our patient with suspected aortic dissection. To overcome those diagnostic difficulties, ECG-gated CT is the preferred modality.[8] In such cases, false interpretations of anatomic structures and artifacts may lead to unnecessary sternotomy. Thus, it is crucial to be aware of potential radiologic pitfalls to avoid increased morbidity and mortality in these patients.

In conclusion, CT angiography is the primary imaging modality in the diagnosis of aortic dissection. Recognizing imaging artifacts and adjacent anatomic structures is vital. It is pivotal to notice these potential pitfalls and use supportive imaging modalities to avoid false-positive results.

*Supplementary video files associated with this article can be found in the online version of the journal.

Informed Consent: Informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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Anahtar Kelimeler: Aort diseksiyonu, aort kapak, aort anevrizması, ekokardiyografi