

## CASE REPORT

## Left ventricular non-compaction in a patient with Shone's complex

## Shone anomalisi olan hastada sol ventriküler nonkompaksiyon

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**Summary**– Shone's complex is a rare congenital cardiac malformation characterized by serial obstructive lesions of the left heart at multiple levels. Presently described is an unusual case of an adult male patient who presented with palpitations and worsening dyspnea. An echocardiographic evaluation revealed Shone's complex associated with left ventricular non-compaction cardiomyopathy (NCC). To our knowledge, an association between NCC and Shone's complex has not been previously described.

**Özet**– Shone kompleksi sol kalpte birçok düzeyde bir dizi obstrüktif lezyonla karakterize seyrek görülen bir doğuştan kardiyak malformasyondur. Burada çarpıntı ve artan nefes darlığı olan bir erişkin erkek hastaya ait olağandışı bir olgu sunuyoruz. Ekokardiyografik değerlendirme sol ventriküler nonkompaksiyon kardiyomiyopatiyle ilişkili Shone kompleksi lehineydi. Bildiğimiz kadarıyla bu kompleksle nonkompaksiyon kardiyomiyopatinin ilişkisi daha önce tanımlanmamıştı.

Shone's anomaly/syndrome/complex is a rare congenital cardiac malformation that was first described by John Shone in 1963,<sup>[1]</sup> and is characterized by 4 serial obstructive lesions of the left heart: a defect in the supra-ventricular mitral membrane, parachute mitral valve, muscular or membranous subaortic stenosis, and coarctation of the aorta.<sup>[2]</sup> The full spectrum of serial obstruction completes Shone's complex; however, fewer pathological lesions may be present in incomplete forms.<sup>[1]</sup> This anomaly is most often diagnosed during childhood, but the mortality rate in patients who reach adulthood is low.<sup>[3,4]</sup> In comparison with other congenital heart diseases, the obstructive lesions in cases of Shone's complex deteriorate over time, and the long-term prognosis of the disease significantly depends on the extent of obstruction of the inflow and outflow of the left ventricle (LV).

Non-compaction cardiomyopathy (NCC) is also a rare congenital failure of trabecular compaction during myocardial development. It may involve both ventricles and is considered to be the result of an ar-

rest in normal embryogenesis.

<sup>[5]</sup> Although NCC has frequently been reported in parallel with other cardiac and non-cardiac neuromuscular disorders,<sup>[6]</sup> an association with Shone's complex has yet to be reported. The present report is a description of a case of Shone's complex in which the patient also had a structural morphology of the LV consistent with NCC.

**Abbreviations:**

LV	Left ventricle
NCC	Non-compaction cardiomyopathy

**CASE REPORT**

A 20-year-old man with no history of medical or surgical illnesses was referred to the outpatient cardiology clinic for evaluation of complaints of palpitations and progressive dyspnea. There was no family history of congenital heart disease or sudden cardiac death. Upon presentation, the patient's pulse rate was 95 bpm with a blood pressure of 145/80 mm Hg measured in the right upper arm. There was no sign of jugular vein distension, cyanosis, or clubbing of the fingers. The apex beat was localized to the sixth intercostal space

Received: February 18, 2018 Accepted: May 08, 2018

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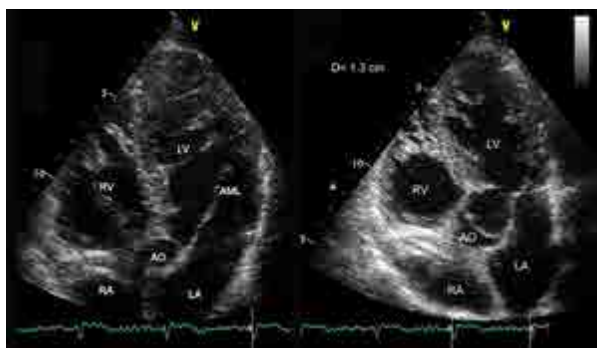
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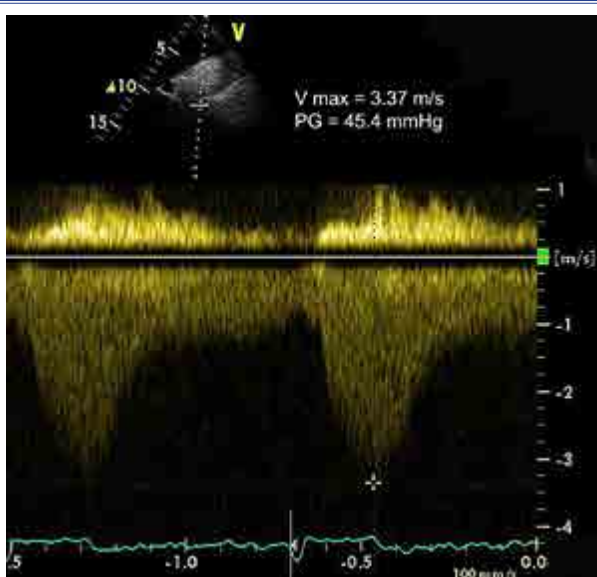


mid-clavicular line with no evidence of parasternal heave. On cardiac auscultation, a grade 2/6 systolic murmur was detected over the right parasternal border.

An electrocardiogram revealed a sinus rhythm with non-specific ST-T changes. A transthoracic echocardiogram was performed and indicated a normal ejection fraction with evidence of mitral valve malformation compatible with a parachute-type morphology and the presence of a single papillary muscle, along with non-significant obstruction to mitral valve inflow and only mild regurgitation (Fig. 1, Video 1 and 2\*). Bicuspid morphology was detected upon evaluation of



**Figure 1.** Apical 5-chamber echocardiographic view showing the parachute mitral valve morphology and multiple false tendons in the left ventricle. The distance between the subaortic web and the right coronary cusp is 13 mm. AML: Anterior mitral valve leaflet; Ao: Aorta, LA: Left atrium; LV: Left ventricle; RA: Right atrium; RV: Right ventricle.



**Figure 2.** Doppler recording from the suprasternal view illustrating significant coarctation of the aorta with a peak gradient of 45 mm Hg.

the aortic valve, along with a subvalvular web, leading to mild obstruction with a peak gradient of 32 mm Hg. There were multiple false tendons in the LV with hypertrabeculation involving the apical and lateral segments of the LV (Video 3\*). The Stollberger criteria for the diagnosis of NCC were fulfilled: a 2-layered myocardial structure with a thicker, non-compacted layer; >3 prominent trabeculations apical to the papillary muscles; and perfused inter-trabecular spaces. Additionally, there was significant coarctation of the aorta with a peak gradient of 45 mm Hg (Fig. 2). Mild tricuspid regurgitation with a peak gradient of 25 mm Hg was also present. According to the normal size and respiratory collapse of the inferior vena cava, systolic pulmonary pressure was estimated at 30 mm Hg.

## DISCUSSION

The current report is a description of multilevel congenital anomalies of the systemic circulation that met all of the criteria of Shone's complex in addition to non-compaction morphology of the LV. In addition to the NCC component, the remaining developmental cardiac anomalies in this case completed the spectrum of findings originally described by Shone et al. in 1963. The parachute deformity of the mitral valve represents a congenital form of mitral stenosis, which is characterized by the unifocal attachment of chordae to a single or fused papillary muscle. The close adjacency of the valvular leaflets secondary to their attachment to a single papillary muscle generally results in the development of restriction in mitral valve inflow. Isolated occurrence of this malformation is rare, as it is almost always seen in combination with other cardiovascular anomalies.<sup>[7,8]</sup> Although it was not the case here, the supra-valvular mitral membrane is frequently characterized by an abnormal circumferential ridge of connective tissue on the atrial side of the mitral valve that disturbs the opening of the mitral valve.

Obstruction of LV outflow may occur at multiple levels in Shone complex. The first level of restriction of LV outflow results from the presence of subaortic webs, which is generally followed by more restrictive flow pattern further away through a coarctation of the aorta at the level of ligamentum arteriosus. The presence of aortic coarctation should raise a suspicion that there may be other associated cardiac anomalies, since it frequently occurs in conjunction with valvular anomalies.

Shone's complex has been reported to coincide with other developmental cardiac disorders, such as dextrocardia and situs inversus totalis,<sup>[9]</sup> a myxomatous Eustachian valve,<sup>[10]</sup> bicuspid aortic stenosis, and hypoplasia of the aortic arch and left superior vena cava.<sup>[3]</sup> Ours is believed to be the first report of co-occurring NCC and Shone's complex. Like the anomalies in Shone's complex, NCC is a developmental disorder of the LV that results from formation of multiple deep trabeculae within the ventricular wall.<sup>[11]</sup> The mechanism of the development of NCC in Shone's complex is unclear; however, an arrest of normal compaction of the loose interwoven mesh of myocardial fibers in the embryo is the most probable cause for this association.<sup>[12]</sup> As with other developmental disorders, NCC has been associated with several congenital cardiac anomalies, including Ebstein's anomaly, congenitally corrected transposition, a bicuspid aortic valve, and ventricular septal defects.<sup>[13]</sup>

The overall prognosis of Shone's complex depends on the severity of the associated pathologies and the progressive nature of the restrictive flow pattern. It has been reported that the natural course of undiagnosed and untreated patients is the development of heart failure and pulmonary hypertension.<sup>[2]</sup> The presence of NCC will further degrade the prognosis of the whole complex since the patient will be prone to ventricular arrhythmia, thromboembolic events, and progressive heart failure.<sup>[13,14]</sup> The long-term prognosis can be improved in cases of Shone's complex with early corrective surgical interventions (with priority given to correcting the aortic coarctation) before the onset of pulmonary hypertension.<sup>[3,15]</sup> We highlight the importance of coexisting NCC in a patient with Shone's complex. It warrants a particularly careful medical treatment strategy, and early surgical intervention may be appropriate in order to preserve cardiac function before irreversible remodeling and changes in the left heart occur.

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

**Peer-review:** Externally peer-reviewed.

**Conflict-of-interest:** None.

**Funding resources:** None

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

**Authorship contributions:** Concept: M.T., L.P., A.B.; Design: A.B., N.N.; Supervision: M.T., N.N.; Material: M.T., L.P.; Data collection: M.T., N.N., L.P., A.B.; Literature search: A.B., L.P.; Writing: A.B., N.N., L.P., M.T.

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**Keywords:** Echocardiography; non-compaction cardiomyopathy; Shone's complex.

**Anahtar sözcükler:** Ekokardiyografi; nonkompaksiyon kardiyomiyo-  
pati; Shone kompleksi.