Exploring Complexities in Differential Diagnosis: Torn Chord Tendinea in the Aortic Valve

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Introduction

The prevalence of anomalous chordae in the aortic valve is rare in clinical practice.[1] These anomalies, although uncommon, can have significant clinical implications, including valve insufficiency, arrhythmias and/or embolic risk. Accurate diagnosis and understanding of its pathophysiology are essential for adequate patient management.

The uncommon nature of anomalous chordae in the aortic valve means that their presence can often go unnoticed or misdiagnosed. Medical literature presents few reports on these structures of probable embryonic origin, leaving gaps in clinical knowledge.[2] Furthermore, due to its ability to influence cardiac hemodynamics and potential to complicate clinical conditions, it is essential to expand the understanding of its incidence, clinical manifestations, and best management practices.

Case report

A 59-year-old female patient with a history of gastroesophageal reflux disease (GERD) attended an outpatient consultation with a cardiologist reporting dyspnea on exertion and unintentional weight loss (around 10 kilos) in 6 months. She denied other cardiovascular symptoms and fever, without infectious stigmata. Physical examination was unremarkable; however, during dental evaluation, impaired oral hygiene and the presence of signs of infection with abscess were noted in a dental unit. During the initial investigation, the transthoracic echocardiogram (TTE) revealed cardiac cavities with normal dimensions (left atrium volume: 25mL/m^2, diastolic diameter 48mm, systolic diameter 30mm, end-diastolic volume 107mL), mild mitral and tricuspid valve reflux, normal diastolic function (E/e': 06), myocardial contractility and preserved biventricular systolic function (LVEF: 67%, estimated using the Simpson method). Furthermore, a filamentary lesion was identified in the aortic valve on its ventricular surface. This lesion, characteristically mobile, had dimensions of approximately 11 mm. Given the clinical relevance of this observation and the potential implication of endocarditis or other cardiac pathologies, blood cultures were collected to identify possible infectious agents.

Transesophageal echocardiography (TEE) assessed the aortic valve lesion with greater precision, identifying incipient signs of degeneration with minimal reflux. Additionally, an image with a serpiginous appearance was detected, with a maximum diameter of up to 21 mm (Figures 1 and 2), adhered to the ventricular surface of the non-coronary valve, suggestive of vegetation.

In addition to evaluating inflammatory markers and searching for infectious foci, additional screening was conducted to investigate autoimmune diseases. The tests performed, which included lupus anticoagulant, rheumatoid factor, anti-Beta-2 glycoprotein antibodies, antinuclear antibodies (ANA), anti-Sm, anti-RNA, and anti-DNA, all returned within normal parameters, not indicating autoimmune disease activity. White blood cell count showed 5.82 thousand/mm^3, C-Reactive Protein (CRP) (5.2mg/L; RV: < 10mg/dL) and procalcitonin remained at normal levels (0.05ng/mL; RV : < 0 .5ng/mL). NT-proBNP was at a level that rules out acute heart failure – 136pg/mL; reference value (RV): < 450pg/mL. Hemoglobin and hematocrit were slightly reduced (12.0g/dL and 36.4%, respectively), indicating mild anemia, but renal function was confirmed as normal (Urea 30mg/dL, Creatinine 0.92mg/dL). In parallel to the infectious and immunological investigation, a complementary evaluation was carried out with a tomography of the chest, sinuses and abdomen, without significant changes, as well as an annual screening mammogram, carried out prior to hospitalization, without relevant findings.

The cardiovascular surgery team was consulted, and the therapeutic approach was discussed in the Heart Team: clinical *versus* surgical treatment. The patient remained afebrile and stable during the hospitalization period, with negative blood cultures. Empirical antibiotic therapy was started (1st set: ceftriaxone associated with oxacillin and gentamicin, later replaced by the 2nd set, with daptomycin associated with ceftriaxone).

After 6 weeks of clinical treatment, during which there was no reduction of the lesion or symptomatic improvement, and considering the significant extension of the lesion with a large mobile component in the left ventricular outflow tract, the medical team decided, together with the patient, to proceed with surgical intervention. This decision was based on the potential risk of embolization, given the mobility and location of the lesion, which substantially increased the possibility of serious embolic events. Prolonged clinical management,

Keywords

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without evidence of regression of the injury, reinforced the need for a more definitive resolution to mitigate the associated risks and promote a more effective recovery.

Intraoperatively, a 5-cm lesion suggestive of chorda tendineae was identified and removed (Figure 1). Anatomopathological evaluation confirmed valve tissue, and culture was negative. Seven days after surgery, the patient was discharged from the hospital, showing subsequent improvement in symptoms and, upon reassessment with TTE, maintained biventricular systolic function and minimal aortic insufficiency, without residual lesions.

Discussion

The chordae tendineae of the aortic valve, also called fibrous "strands", formed between the aortic valve and other cardiac structures, are embryonic remnants of the valve formation process.[3,4] In terms of diagnosis, there are descriptions of its association with embolic events or endocarditis, although its clinical relevance is still subject to debate. Therefore, careful evaluation and close clinical follow-up are recommended when such structures are identified.[5,6]

Although this congenital alteration is a rare observation on imaging exams, its identification is of fundamental importance, as, in some cases, it may be associated with complications.[3,4] Among these, we may consider the possibility of embolic events and progressive aortic regurgitation, including signs of ectasia of the proximal aortic segments and/or consequent eccentric left ventricular hypertrophy due to volumetric overload.

Anomalous aortic chordae tendineae can be associated with valve lesions and manifest itself both in chronic conditions, with an intact cord, and in acute cases of aortic insufficiency due to its rupture. This rupture, whether spontaneous or resulting from trauma, ischemia or senile degeneration, can give rise to a mobile filamentary image on exams, complicating the differential diagnosis with thrombi or vegetation, as demonstrated in the clinical case at hand.[5]

The change in flow generated by the presence of chorda tendineae in an anomalous position can alter the phenotypic expression of endocardial cells in the left ventricular outflow tract, resulting in fibroproliferative lesions that can increase the incidence of cardiac complications.[1] Furthermore, this tendon anomaly can also lead to the formation of a gradient in the left ventricular outflow tract, even resulting in subvalvular aortic stenosis.[4]

During the intraoperative surgical approach, it was possible to identify in detail the specific sites of insertion of the anomalous chorda tendineae (Figure 2), located in the interventricular septum, in the region of the mitral aortic window and, additionally, in the Arantius node belonging to the right coronary valve. Notably, the cord associated with this last site was broken, which determined the finding correlated with the TTE (Figures 3 and 4).

The detection of an anomalous chordae tendineae in the aortic valve, especially when considering the patient in question, highlights the complexity and diagnostic challenges faced in clinical practice. In the present case, the extremely mobile lesion observed in the left ventricular outflow tract introduced an ambiguous diagnostic scenario, where the possibility of endocarditis emerged as a differential diagnosis. This case highlights the importance of an in-depth understanding and a meticulous approach in evaluating such anomalies, thus ensuring diagnostic accuracy and appropriate clinical management.

Conclusion

The presence of chordae tendineae in the aortic valve, although often underdiagnosed, can have significant implications for cardiac hemodynamics. When this tendinous chord is ruptured with a large component of mobility, it can be confused with vegetation, commonly associated with endocarditis, introducing complexity to the differential diagnosis.

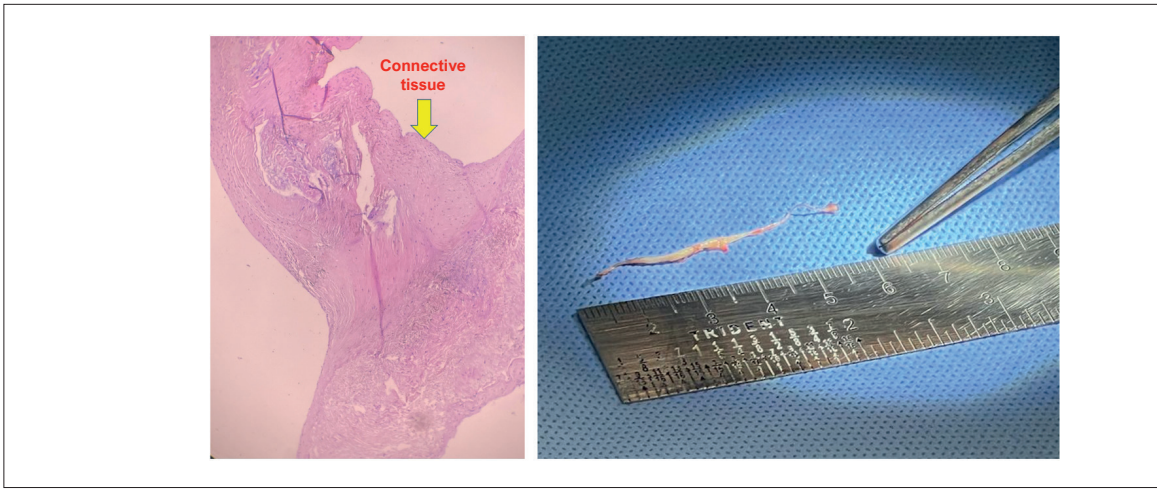

Figure 1 – *On the left, histological section with connective tissue characteristic of valve tissue with the presence of myxomatous areas in between. On the right, anatomical image of the specimen after excision of the lesion.*

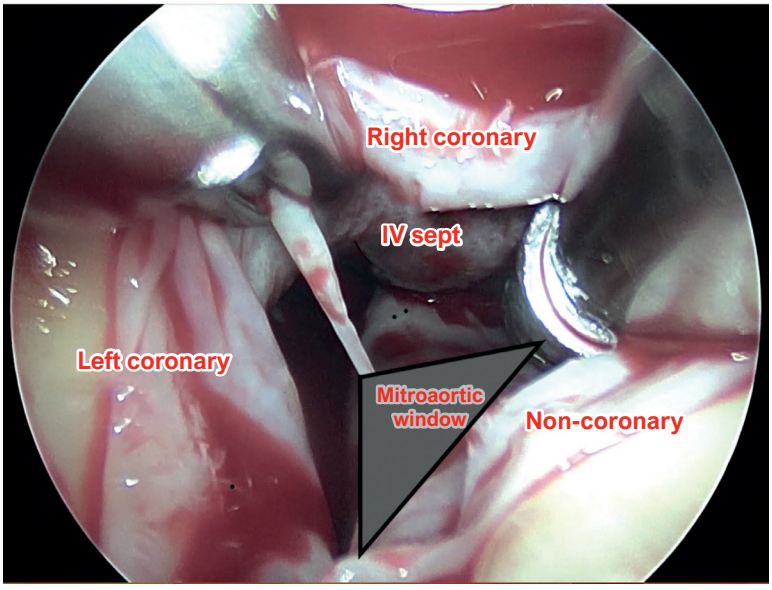

Figure 2 – *Surgeon's view through videothoracoscopy in minimally invasive surgery: after transverse aortotomy and suspension of the aortic valve leaflets. An anomalous cord is identified in the left ventricular outflow tract with fixed ends in the interventricular septum and Mitroaortic Fibrosa (FIMA).*

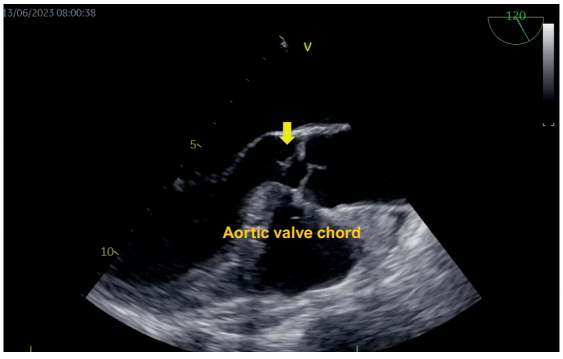

Figure 3 – *Transesophageal echocardiogram at 120°, showing a filamentary lesion related to the ventricular surface of the aortic valve, markedly mobile and measuring up to 21mm.*

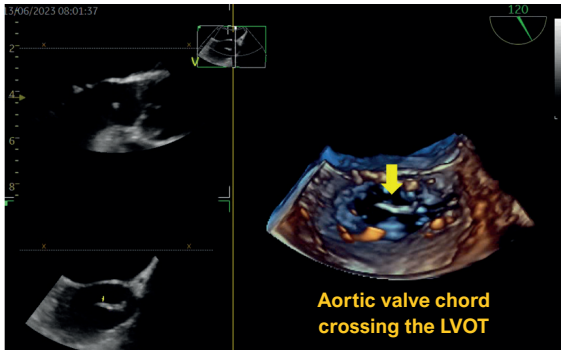

Figure 4 – *3D Transesophageal Echocardiogram, in 4D Zoom mode, with visualization from the ventricular view, showing a filamentary lesion crossing the left ventricular outflow tract.*

Author Contributions

Conception and design of the research: Costa A; acquisition of data: Costa A, Alves ES, Pinheiro P, Flausino L; analysis and interpretation of the data: Freire MV; writing of the manuscript: Costa A, Freire MV, Pinheiro P; critical revision of the manuscript for intellectual content: Morel RV.

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Study Association

This study is not associated with any thesis or dissertation work.

Ethics Approval and Consent to Participate

This article does not contain any studies with human participants or animals performed by any of the authors.

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