A Rare Case of Acute Coronary Syndrome in a Patient With Turner Syndrome

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Received 2015 August 25; Revised 2015 September 16; Accepted 2015 October 30.

Abstract

Introduction: In Turner syndrome, cardiovascular complications are the most important causes of early mortality. Congenital cardiovascular abnormalities are found in approximately one third of Turner syndrome patients. Developments in diagnosis and treatment have decreased the rate of mortality related to these abnormalities. In recent years, many papers have mentioned that coronary artery disease developing at early ages in patients with Turner syndrome causes sudden deaths.

Case Presentation: The patient, a 27-year-old female was admitted to the emergency room with chest pain at rest. She was diagnosed with Turner Syndrome in her teenage years due to amenorrhea. Patients with ECG changes and cardiac enzyme elevations were treated with acute coronary syndrome.

Conclusions: The young woman with Turner Syndrome have several risk factors for early Coronary Artery Disease development. In such cases, dramatic results like sudden death or heart attack at an early age may occur in cases of insufficient follow-up and treatment.

Keywords: Turner Syndrome, Acute Coronary Syndrome, Karyotype

1. Introduction

Turner syndrome (TS) is seen in female infants at a rate of 1 in 2,500, and it occurs as a result of partial or complete absence of an X chromosome (1). It is detected in about 30% of all congenital cardiac abnormalities (2). Although many cardiac abnormalities have been identified, the most common are obstructive abnormalities in the left side of the heart, especially in the bicuspid aortic valve (16%), and aortic coarctation (11%) (3). These patients carry an obvious risk for coronary artery disease (CAD), which is still an important cause of mortality and morbidity worldwide. Many major risk factors (such as hypertension, hyperlipidemia, and diabetes) that contribute to the development of CAD appear at an early age in patients with Turner syndrome.

2. Case Presentation

The patient, a 27-year-old female, was diagnosed with TS in her teenage years due to amenorrhea and short stature. Cytogenetically, the patient is mosaic 45XO/46XX. The patient was taking metaprolol (50 mg daily) for hypertension. Her family history was negative for CAD, and she was not a smoker. The patient was admitted to the emergency room with chest pain at rest, her blood pressure was 168/85 mmHg, and her pulse rate was 110 beats/minute. She was 139 cm tall, weighed 47 kilograms, and had a body mass index of 24.3 kg/m². Electrocardiography showed sinus tachycardia and 2 mm ST depression in leads D1, aVL, and V3 – V6. The admission troponin value was 3.25 ng/ml. Biochemical analysis showed a fasting glucose level of 128 mg/dl, total cholesterol of 265 mg/dl, high-density lipoprotein (HDL) cholesterol of 48 mg/dl, triglycerides of 238 mg/dl, and low-density lipoprotein (LDL) cholesterol of 169 mg/dl. Two-dimensional echocardiography showed a mildly reduced ejection fraction (40%) and wall motion abnormalities in the anterior, anterior septum, and the anterolateral wall in the parasternal short axis image and in the anterior septum and apex in the apical 2 - 4 chamber image. The aortic valve was tricuspid; it showed moderate insufficiency, and aortic coarctation was observed in the suprasternal window. The patient underwent cardiac catheterization. The left main coronary artery (LMCA) was absent. The left anterior descending artery (LAD) and the left circumflex artery (CX) originated from different orifices. The LAD was a small vessel and had a 99% stenosis before the first diagonal branch (Figure 1). The CX and the right coronary artery (RCA) were irregular. The aortic coarctation was detected via aortography (Figure 2). Consequently, this TS patient was diagnosed with severe LAD stenosis and aortic coarctation, which were treated with stent placement.
3. Discussion

CAD is still the most important cause of death in the general population. Female patients with TS have increased risk for not only congenital heart disease but also CAD (4). TS patients have many major risk factors for CAD. Hypertension affects approximately 50% of TS patients. Altered autonomic innervation of the heart and increased heart rate are also commonly seen in patients with TS (5). Type 2 diabetes is also common. In addition to defective insulin secretion, more than half of TS patients have impaired glucose tolerance or diabetes. Although insulin sensitivity is present in many patients, a strong familial history of type 2 diabetes and obesity decrease this sensitivity (6). In female TS patients, LDL and triglycerides are generally higher, and lipid particle size is smaller than in the general female population (7). Moreover, estrogen deficiency is commonly seen in these patients, which causes intimal thickening and changes in artery wall dynamics (8). All of these conditions facilitate the development of coronary artery disease and may cause undesired results in cases of insufficient risk monitoring and treatment.

3.1. Conclusion

The young woman with TS discussed above seems to have several risk factors for early CAD development. In such cases, dramatic results like sudden death or heart attack at an early age may occur in cases of insufficient follow-up and treatment. On the basis of these considerations, every TS patient should be evaluated for coronary artery disease, regardless of whether they have cardiac symptoms.

Footnote

Authors’ Contribution: All author have contributed to the report, diagnosis and treatment of the case.

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