Images in Cardiovascular Medicine

Cardiac Fibroma Presenting With Left Bundle Branch Block in an Adult With Gorlin Syndrome

Kumar Jatti, MD, MRCP¹; Ramya Dhandapani, FRCR²; Vishal Sharma, MD, MRCP¹; Balazs Ruzsics, MD, PhD¹

¹Department of Cardiology, Royal Liverpool University Hospitals NHS Foundation Trust, Royal Liverpool University Hospital, Liverpool, United Kingdom

²Department of Radiology, Royal Liverpool University Hospitals NHS Foundation Trust, Royal Liverpool University Hospital, Liverpool, United Kingdom.

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Case Description

A 60-year-old man presented with chest pain and palpitations. His troponin level was normal, and electrocardiography showed left bundle branch block (LBBB) (Fig. 1). Echocardiogram demonstrated moderate left ventricular systolic dysfunction, and no mass was clearly reported (Fig. 2). His invasive coronary angiogram was normal. Cardiac magnetic resonance imaging showed a solitary, ovoid (5×2.7 cm), well-circumscribed intramural mass in the mid- to apical lateral segments. The lesion showed low intensity relative to the myo-



Fig. 1 Image of the patient's electrocardiogram with Gorlin syndrome shows left bundle branch block.



Fig. 2 Echocardiogram showing A) 2-chamber and B) 4-chamber images. Echo windows were poor with suspected rib shadow over the mass (which, in retrospect, was suspected to be attached to lateral segments but with no clear visual conclusion).

Supplemental motion image is available for Figures 2A and 2B.

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Corresponding author: Balazs Ruzsics, MD, PhD, Department of Cardiology, Link 7Z, Royal Liverpool University Hospitals NHS Foundation Trust, Royal Liverpool University Hospital, Prescot Street, Liverpool, United Kingdom, L7 8XP (Balazs.Ruzsics@liverpoolft.nhs.uk) © 2023 by The Texas Heart® Institute, Houston



Fig. 3 Cardiac magnetic resonance imaging shows the lesion with predominantly low intensity relative to the myocardium on **A**) T1w images and **B**) T2w images. On late gadolinium contrast–enhanced images **C**), the mass demonstrated intense homogenous delayed hyperenhancement. Arrow shows cardiac fibroma location.

Abbreviations and Acronyms

LBBB left bundle branch block

cardium on T1w images (Fig. 3A) and T2w images (Fig. 3B). On late gadolinium-enhanced images, the mass demonstrated intense homogenous enhancement (Fig. 3C). Imaging characteristics of the mass were in keeping with cardiac fibroma. In retrospect, an unclear, round mass was suspected in some of the echocardiogram images; this is a rare manifestation that can be missed even by an experienced operator.

On review of the patients' previous imaging records, x-ray and computed tomography of the mandible demonstrated multiple lytic lesions consistent with keratocystic odontogenic tumors (Fig. 4). Biopsy results of multiple skin lesions confirmed basal cell nevi. A conclusive diagnosis of cardiac fibroma associated with Gorlin syndrome was established and supported by further genetic testing.

Comment

Gorlin syndrome is a rare autosomal dominant disorder with complete penetrance and variable expressivity,¹ with estimated prevalence of 1 in 19,000.² The



Fig. 4 X-ray of mandible **A**) and corresponding computed tomography of mandible **B**) show multiple lytic lesions (arrows) in association with Gorlin syndrome.

syndrome is associated with multiple basal cell carcinoma, and additional features may include craniofacial, central nervous system, musculoskeletal, and genitourinary anomalies. Approximately 3% to 5% of cases are associated with cardiac fibromas. Cardiac fibroma is a common pediatric cardiac tumor with only approximately 15% of cases occurring in adults.¹ Arrhythmia is a common presentation, but in the index case, conduction abnormality (LBBB) was noted.

Magnetic resonance imaging shows unique T1 and T2 characteristics, including intense homogenous late gadolinium enhancement.³ Left bundle branch block usually is a sign of underlying heart disease. This case is cardiac fibroma presenting with LBBB. Cardiac magnetic resonance imaging can be an important diagnostic step in patients with newly found LBBB.

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