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/CASE REPORT

Extrinsic compression of an anomalous left main coronary artery in a patient with pulmonary arterial hypertension presenting with myocardial injury

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Extrinsic compression of the left main coronary artery (LMCA) caused by severe pulmonary arterial dilatation in the setting of pulmonary arterial hypertension (PAH) is a recognised entity.[1] This can present with angina, cardiogenic shock, malignant arrhythmias or sudden cardiac death.[2] We report the case of a 17-year-old female with a history of primary PAH who presented with acute chest discomfort, elevated biomarkers and ECG changes. Invasive and non-invasive imaging confirmed her diagnosis and identified an anomalous origin of the left main coronary artery and significant LMCA extrinsic compression by an enlarged main pulmonary artery (MPA). The abnormal anatomical location of the LMCA resulting from its anomalous origin could have further contributed to the risk of compression.[3]

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Introduction

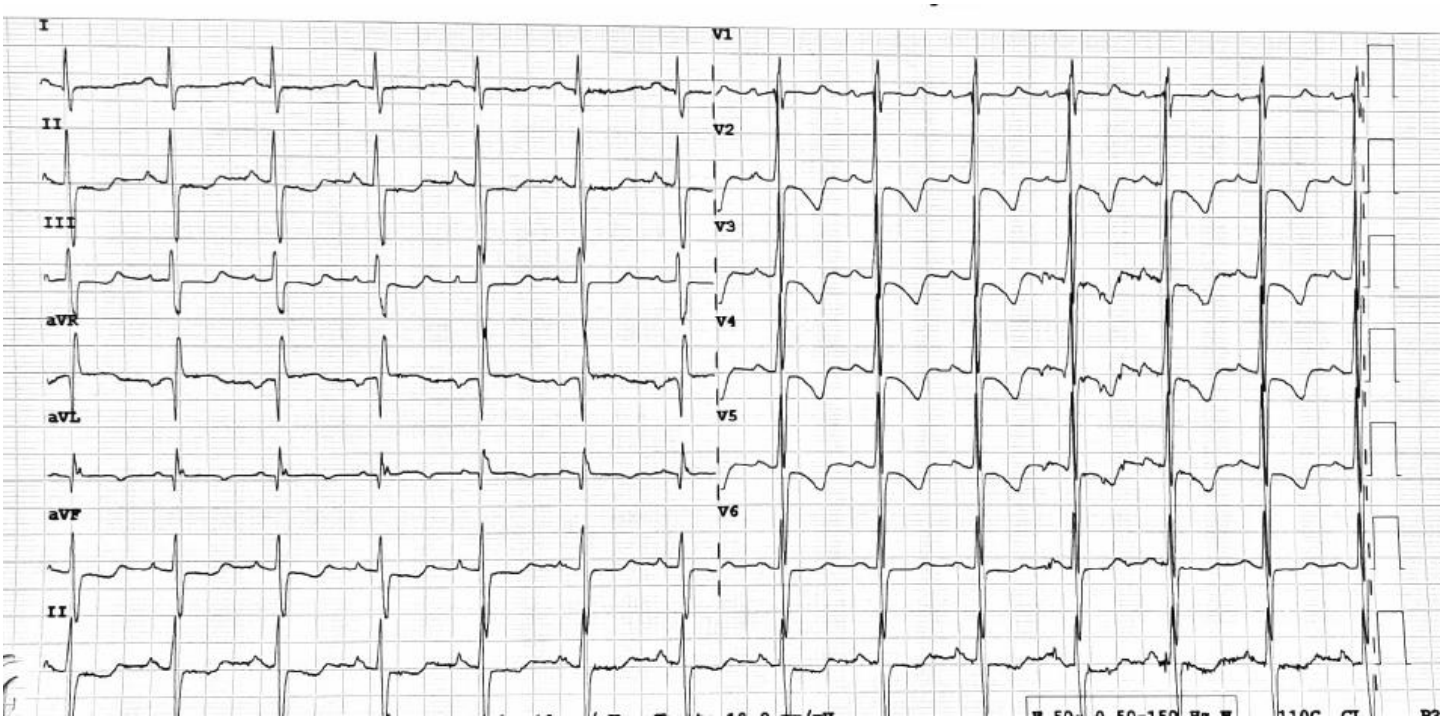
Extrinsic compression of coronary arteries by severely dilated pulmonary arteries in severe PAH is a recognised phenomenon.[1,3] This can lead to angina with/without myocardial injury, cardiogenic shock, arrhythmias and even sudden cardiac death.[2]

Case Report

A 17-year-old female with severe primary PAH diagnosed at the age of 13 years and established on sildenafil since, presented to the Accident and Emergency Department of our institution with compressive chest pain and presyncope. She was also being followed up by an inherited cardiomyopathy specialist in view of suspected biventricular non-compaction cardiomyopathy. She had an implantable loop recorder (ILR) *in situ* for several months in view of a history of presyncope and syncope.

On clinical examination, she had a split second heart sound with a loud pulmonary component and an end diastolic murmur at the left upper sternal border all in keeping with her known PAH. A high sensitivity troponin T of 1087ng/L (UL 14ng/L) was recorded upon admission which peaked at 1446ng/L within a few hours. A 12-leadECG upon admission showed wide spread downsloping ST-segment depressions and deep T wave inversions as well as subtle ST segment elevations in lead aVR (Figure 1). No arrhythmias were detected on ILR check.

Figure 1 Twelve-lead electrocardiogram (ECG) at time of admission showing widespread downsloping ST segment depressions and deep T wave inversions as well as subtle ST segment elevations in lead aVR.



An invasive right and left heart catheter study confirmed severe PAH with near-systemic pulmonary artery pressures (PAP) (systolic PAP = 75mmHg vs. systolic aortic pressure = 80mmHg) and a mean PAP of 52mmHg under general anaesthesia. Aortography revealed the LMCA to be originated anomalously from the aorta at the level of the sinotubular junction. A tight long stenosis of its proximal segment was also clearly evident (Figure 2) which is the likely cause of the documented myocardial injury. Retrospective review of a recent cardiac computed tomography (CT) scan confirmed the anomalous coronary origin with extrinsic compression by an adjacent severely dilated MPA with the latter measuring 45 x 48mm (Figure 3).

Figure 2 Aortographic image of the anomalous origin of the left main coronary artery and a tight stenosis on a long proximal segment.

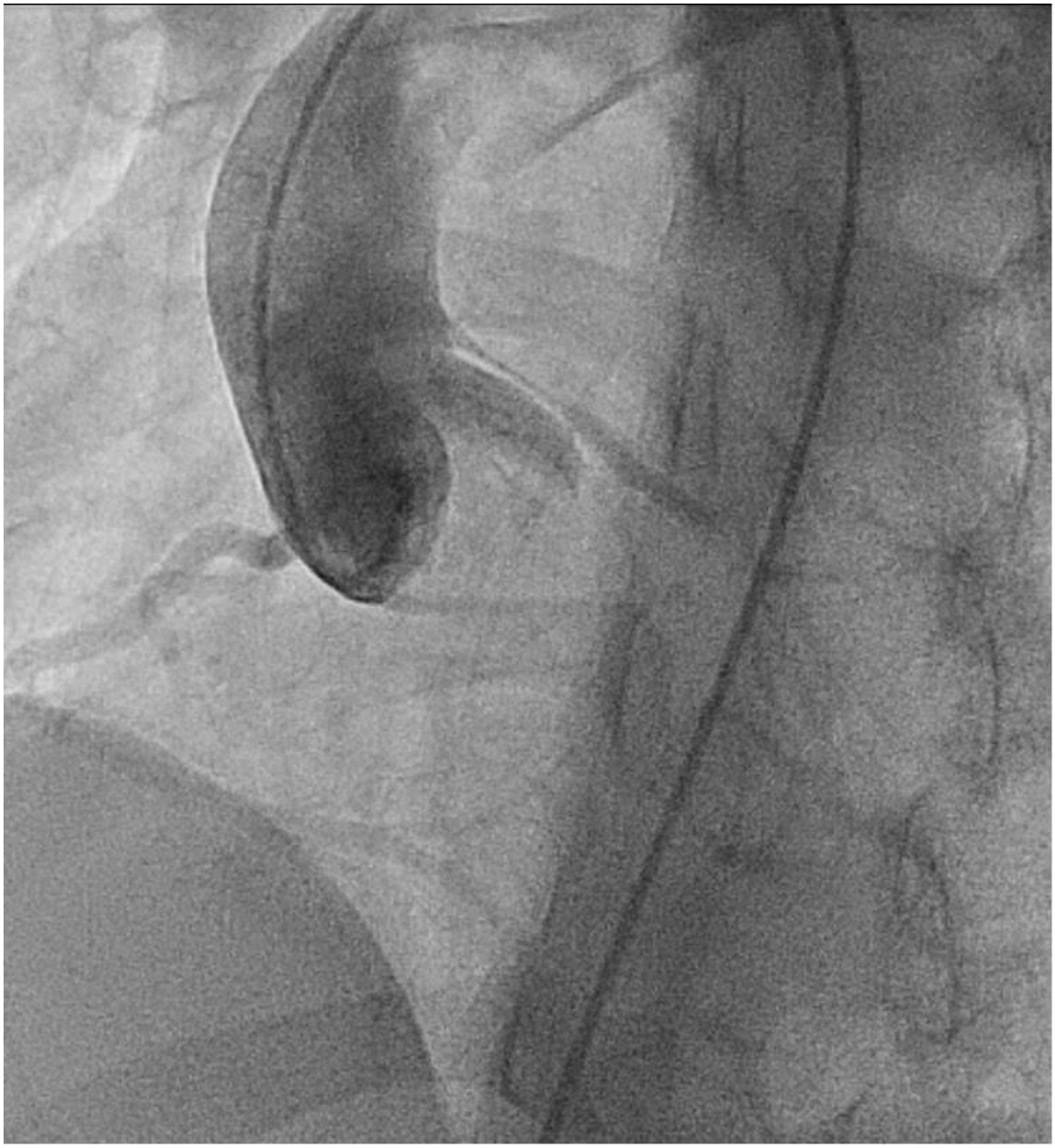
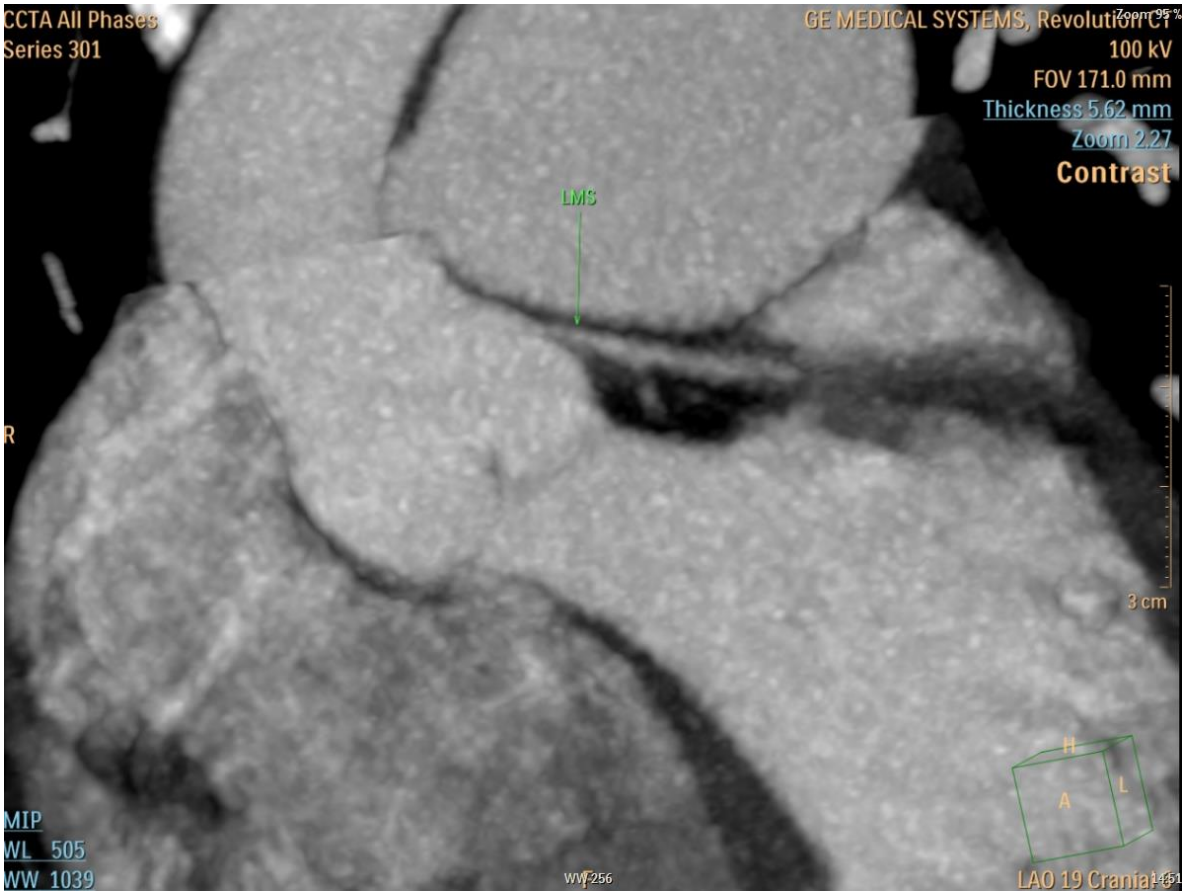


Figure 3 Extrinsic left main coronary artery compression by the enlarged pulmonary arteries. Cardiac CT also shows the anomalous origin of the left main artery above the sinus of valsalva.



Following extensive discussion about a high-risk LMCA stenting, it was decided to uptitrate advance pulmonary vasodilator treatment in the first instance. She responded well to the addition of macitentan and has reported improvement in her breathlessness and chest discomfort and no further syncopal episodes.

Discussion

We report the case of a patient that sustained myocardial injury as a result of severe extrinsic compression of the LMCA by a severely dilated main PA in the setting of primary PAH. Myocardial injury identified by elevated cardiac troponin markers has been documented in patients with PAH and is associated with an adverse prognosis. LMCA compression in the setting of PAH is probably an underestimated cause of anginal chest pain.[5] Nonetheless, the prevalence of LMCA compression in patients with pulmonary hypertension and angina is high and various imaging modalities including CTCA have indentified significant LMCA compression in patients with chest pain.[1] A number of risk factors have been associated with LMCA compression in raised pulmonary pressures. These include young age, pulmonary trunk dilation of more than 40mm and a PA trunk / aorta ratio of >1.2.[6] In keeping with our case, Albadri et al highlighted the high anomalous of origin the LMCA is possibly another risk factor for LMCA compression.[3] Percutaneous coronary intervention of the compressed

coronary artery segment in addition to pulmonary hypertension treatment appears to offer an effective and favourable outcome.[1,3,5]

Our case is rather unique in that the compressed coronary artery had an anomalous origin. To the best of our knowledge, there has been only one similar case of an anomalous LMCA extrinsic compression reported in the literature. [4]

In conclusion, chest pains in patients with severe PAH should always alert clinicians to the possibility of coronary compression, especially if accompanied by ischaemic ECG changes and/or cardiac biomarker elevation.

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