A rare localization and Presentation of cardiac Paraganglioma

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Abstract

Paragangliomas (PGLs) are rare neuroendocrine tumors that arise from the autonomic ganglia. A 34-year-old woman was admitted to our department with palpitation, fatigue and chest pain under stress. Past medical history included a para-carotid PGL surgically removed 5 years earlier. Her family history was notable for familiar PGL with SDH mutation. At stress-echocardiography, was recorded a marked ST-elevation on precordial-leads, concomitant to new onset anterior septum hypo-kinesis, and therefore the patient was proposed for coronary angiography. Although angiographic evidence of atherosclerosis were not recorded, was found that Left Anterior Descending Coronary Artery gave off some branches towards a vascular structure (3×2 cm) in contiguity with the left coronary sinus of the aortic root. Subsequent Compute-Tomography and Magnetic-Resonance clarified the anatomical relationships of the mass with adjacent cardiac structures, in particular the mass was located behind the pulmonary artery trunk and lateral to aortic root, lying above the first portion of the anterior descending coronary artery. Subsequently a careful discussion about the risks and benefits of all therapeutic opportunity, the open surgical resection was deemed the best option for the patient despite the complex clinical anatomy. After standard cannulation for CPB and cardioplegic arrest, transection of the pulmonary artery and the aorta was performed granting adequate exposure of the mass and identification of afferent and efferent vascularization for complete resection though a cleavage plane. To ensure adequate hemostasis on a such critical area, the continuity of the pulmonary trunk was ensured only after removing aortic cross-clamp. The post-operative course was uneventful

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Running Head: Imaging of Cardiac Paraganglioma

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Text

Paragangliomas (PGLs) are rare neuroendocrine tumors that arise from the autonomic ganglia. The familial PGLs have been associated most commonly with succinate-dehydrogenase (SDH) complex mutation of the electron transport chain.(1)

A 34-year-old woman was admitted to our department with palpitation, fatigue and chest pain under stress. Past medical history included a para-carotid PGL surgically removed 5 years earlier. Her family history was notable for familiar PGL with SDH mutation.

At admission was asymptomatic at rest without history of smoking, hypertension, diabetes, coronary artery disease, or peripheral atherosclerosis.

At stress-echocardiography, was recorded a marked ST-elevation on precordial-leads, concomitant to new onset anterior septum hypo-kinesis, and therefore the patient was proposed for coronary angiography. Although angiographic evidence of atherosclerosis were not recorded, was found that Left Anterior Descending Coronary Artery gave off some branches towards a vascular structure (3×2 cm) in contiguity with the left coronary sinus of the aortic root (Fg2 + Vid). Subsequent Compute- Tomography and Magnetic-Resonance clarified the anatomical relationships of the mass with adjacent cardiac structures (Fig 2 – 4), in particular the mass was located behind the pulmonary artery trunk and lateral to aortic root, lying above the first portion of the anterior descending coronary artery.

Subsequently a careful discussion about the risks and benefits of all therapeutic opportunity (2), the open surgical resection was deemed the best option for the patient despite the complex clinical anatomy(3).

After standard cannulation for CPB and cardioplegic arrest, a transection of the pulmonary artery and the aorta was performed granting adequate exposure of the mass and identification of afferent and efferent vascularization for complete resection though a cleavage plane (Fig 1). To ensure adequate hemostasis on a such critical area, the continuity of the pulmonary trunk was ensured only after removing aortic cross-clamp.

The post-operative course was uneventful.

References

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Figure Legend:

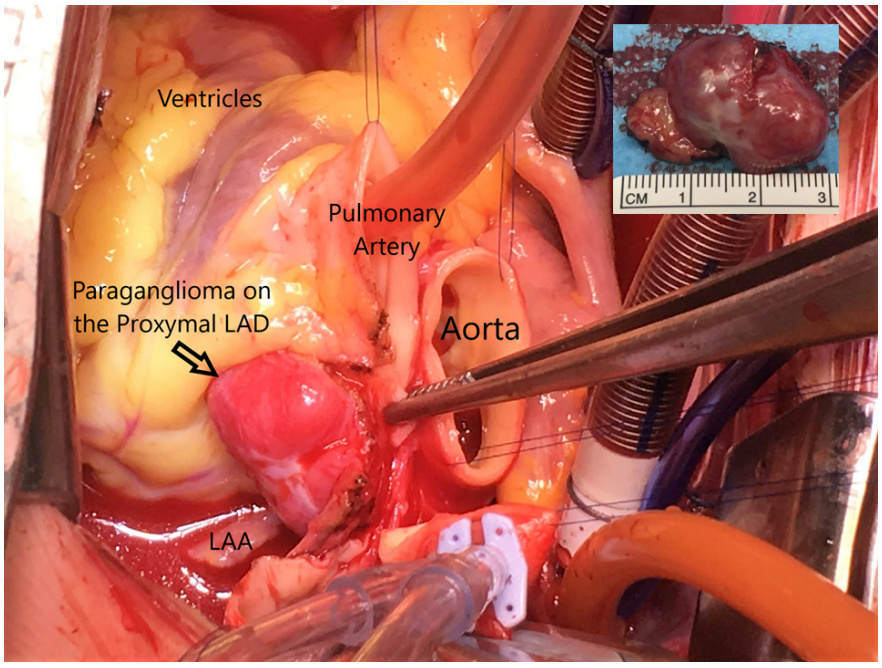
Figure 1 (Central Picture): Surgical View of PGL: to be noted the transected Aorta and Pulmonary Artery. LAA: Left Atrial Appendage; in the right upper corner: surgical specimen after removal

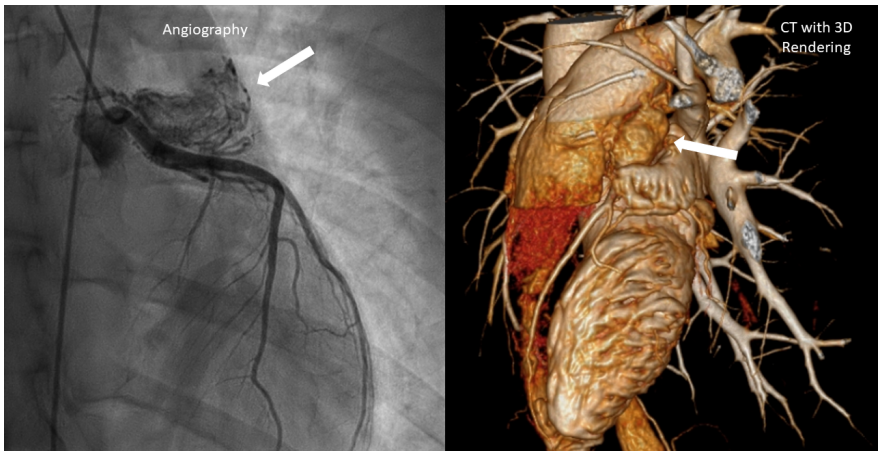
Fig 2: Coronary Angiography and Computerized Tomography with 3D Rendering; arrows are pointing at the PGL

Fig 3: Echocardiographic (Trans-esophageal) (TEE) and Magnetic Resonance View (MR); arrows pointing at the PGL.

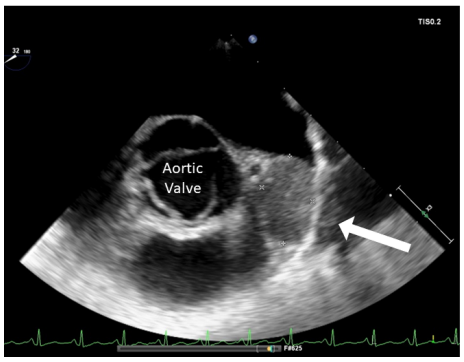
Video Legend:

Coronary angiography





TEE



MR