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An exceptionally rare cause of myocardial ischemia: a case report

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Introduction: Paragangliomas are rare, slow-growing tumors derived from neural crest cells. Paragangliomas may exhibit locally invasive behavior or metastatic disease. Few cases of paragangliomas located in the pericardium are reported. We describe a case of pericardial paraganglioma followed-up with cardiac MR for 7 years, until it caused coronary artery compression and myocardial ischemia.

Case report: A 35–year-old woman with a history of partial resection of latero-cervical paraganglioma (positive for SDHB mutation-c.744C > G) treated with radiometabolic radiotherapy, was referred to our Hospital for characterization of an increased Gallium signal on PET-TC at the level of left atrial appendage.

The patient underwent Cardiac Magnetic Resonance (CMR) (panel A), which documented a well-defined, highly vascular, paraaortic, intra-pericardial mass between the aortic bulb and the pulmonary artery. MR signal was hyperintense on T2 STIR, hypointense on T1 and with scarce, inhomogeneous LGE with a fibrotic labrum suggesting a possible myocardial relapse of paraganglioma.

CMR 5 and 7 years follow-up (panel B-C) showed an increase in mass diameter with initial compression of the left Valsalva sinus, superiorly to the left main/ LAD artery and inferiorly to the proximal portion of the common trunk of the pulmonary artery.

Stress-EKG was positive for ST elevation in aVR with diffuse ST depression (II, III, aVF, V4-5-6) suggesting inducible ischemia due to mass compression . Coronary angiography showed normal coronary arteries.

The patient underwent successful resection of the mass (Panel D). Histology confirmed the diagnosis of extra-suprarenal paraganglioma, with no vascular invasion: hematoxylin and eosin staining showed a well-defined nest of cuboidal cells; immunohistochemistry was strongly positive for chromogranin A and synaptophysin and negative for cytokeratins, excluding a carcinoid tumor.

Conclusion: Intrapericardial paragangliomas are slowly growing masses; they have the capability to infiltrate or compress the coronary arteries: in this case a paraganglioma caused inducible myocardial ischemia.

CMR is ideal for depicting the site, signal and dimension of these rare intrapericardial tumors. In case of masses adjacent to the aortic root, a stress test can help the clinical management.

Abstract P402 Figure.