Cardiac Paraganglioma – A Rare Case Of A Rare Tumor

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Introduction
Cardiac paraganglioma is a rare entity of an uncommon neuroendocrine tumor. Clinically, non-secreting tumors are often diagnosed because of their growth effects, secreting tumors present symptoms related to catecholamines. Correct diagnosis of a paraganglioma can be reached by biochemical investigations and imaging. Surgical resection is the treatment of choice and has to be planned carefully and interdisciplinarily.

Methods
We report the case of a 42-year old woman whose unspecific symptoms and initial investigations revealed a paracardiac mass, which upon further imagistic examinations led to the suspicion of a cardiac paraganglioma.

Results
The initial clinical presentation was intermittent fever, attacks of sweating, dyspnea and dull epigastric pain and also relapse of previously experienced back pain. The CT and afterwards PET/CT scans revealed the mediastinal mass (Fig. 1). No further pathologic findings were noted and after a dynamic heart-MRI, which showed no clear adherence of the mass with the myocardium, the resection was recommended.

We performed an open tumor resection via right lateral thoracotomy. After complete preparation of the situs and localization of the mass, which showed an infiltration of the left atrium of the heart, cardiopulmonary bypass (CPB) was required to perform resection. For better access to the tumor, the superior right pulmonary vein was temporarily disconnected. After removal of the tumor with resection of the adherent left atrial wall, which was infiltrated by the tumor over an area of 3 cm in diameter, the atrial wall was reconstructed with a bovine pericardium patch. The pulmonary vein was re-anastomosed.

Histopathologically, the typical specimen was encapsulated, 6.4 cm in diameter, with focal invasion of the left atrium, displaying a nested (“Zellballen”) architecture (Fig. 2a). Immunohistochemically chromogranin A, synaptophysin and staining for S100 protein were positive while tumor cells exhibited intense staining for succinate dehydrogenase subunit B (SDHB) (Fig. 2b).

Conclusion
Paragangliomas are extremely rare chromaffin cell tumors that may be treated curatively by resection. Because of unspecific symptoms and clinical presentation, the diagnosis of this type of tumor is a challenge. Surgical resection is the treatment of choice and operative techniques suggested include simple excision, excision with reconstruction, or even cardiac autotransplantation or orthotopic cardiac transplantation.

Literature:
• Matsumoto J, Tanaka N, Yoshida Y, Yamamoto T. Resection of an intrapericardial paraganglioma under cardiopulmonary bypass. Asian Cardiovascular & Thoracic Annals 21(4) 476–478