عنوان مقاله:

Cardiac Paraganglioma, Recent Advances in Clinicopathologic Features

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خلاصه مقاله:

Cardiac paragangliomas (PGs) are rare neuroendocrine tumors comprising less than ۱% of cardiac tumors. Few cardiac PG case reports and series are described in the literature. Cardiac PGs may be sporadic or arise from a syndromic association. Clinical presentations vary depending on the biochemical activity and location of the tumor. The left atrium, the right atrium, AP window, left ventricle, and atrioventricular groove are the most common sites for cardiac PGs, respectively. Many cardiac PGs are associated with succinate dehydrogenase (SDH) gene mutations. SDH-mutated PGs have aggressive histologic and clinical behavior. Therefore, PG patients should be screened for SDH mutations and provided with appropriate genetic counseling. SDH immunostaining can be used as a substitute diagnostic modality for SDH gene mutation and negative staining is associated with SDH mutation. Biochemical analysis, anatomical imaging, and functional imaging are also used for diagnostic workup of the tumor. Surgery is the only curative treatment for this tumor. Adrenoceptor blockers should be administered in functional PGs. PGs are highly vascular and frequently situated close to vital vessels. Accordingly, surgical complications such as bleeding are a leading cause of mortality in PGs. Metastatic PGs are only seen in a small subset of patients and are associated with poor clinical outcomes. Herein, we summarize clinical and pathological advances in cardiac PGs.

کلمات کلیدی:

Heart, Paraganglioma, Neuroendocrine Tumors, Succinate Dehydrogenase, Heart Atria