

1. Abstract

Pheochromocytoma/ paraganglioma (FEO/PGL) may be developed on the basis of an inherited genetic mutation of different genes. They are associated with a high risk of developing of secondary hypertension, organ damage and metastatic disease that can be fatal. The aim was to focus on the possibility of genetic testing in patients with FEO/PGL, especially in patients with malignant tumors. The issue FEO/PGL, however, concerns not only the examination and assessment of risks arising therefrom, as well as other therapies and monitoring, including appropriate recommendations for clinical practice.

We demonstrated a 20% incidence of cardiovascular (CV) complications before determining the final diagnosis of FEO/PGL, mainly arrhythmic, followed by complications of myocardial ischemia and accentuate atherosclerosis. Elevated levels of vitamin C and decreased levels of malondialdehyde (MDA) following the successful removal of the tumor demonstrated reduction of oxidative stress postoperatively. We found that early postoperative testing of levels of plasma metanephrines to confirm the success of surgical removal of FEO/PGL is already possible, since there was no significant correlation between plasma levels of metanephrines and postoperative examination interval. Distribution of frequency of metastatic tumors with mutations in *SDHB* gene and sporadic tumors (ST) in adult patients is approximately the same, in children we found 5 times more FEO/PGL with mutations in *SDHB* gene, but with longer survival than in adult patients with this mutation. Tumors greater than 4.5 cm were more primary metastatic and noradrenergic PGL. Our data did not show significant differences between overproduction of different types of catecholamines or metanephrines in connection with age. Metastatic tumors are similarly aggressive in both children and adults. Adult patients with ST have better 10-year survival interval. The results of our study support recommendations for long-term follow up of patients, examination of *SDHB* mutations in all tumors diagnosed in childhood, increase the frequency of follow up in elderly and patients with tumors greater than 4.5 cm, whole body imaging in patients with *SDHB* mutations for a high probability of detection of metastatic disease in bone at the time of diagnosis.