A case of paraganglioma with cyanotic congenital heart disease

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Abstrak

Co-occurrence of cyanotic congenital heart disease (CCHD) and phaeochromocytoma (PCC) and paraganglioma (PGL) are rare, although some cases have been reported. We report a case of left paraganglioma in a 20-year-old lady with an underlying CCHD who underwent palliative Glenn shunt, subsequently developed polycythaemia and cavernous sinus thrombosis presented with palpitation, sweating, headache and hypertension of 3-months duration at the age of 17. The abdominal CT scan revealed an enhancing left paraaortic mass measuring 5.2 cm x 4.4 cm x 3.8 cm. A 24-hour urine catecholamine demonstrated raised noradrenaline level to six times upper limit of normal and hence diagnosis of left sympathetic (sPGL) was made. In view of the delayed diagnosis and significant morbidity associated with her condition, surgical treatment is no longer an option. Therefore, vigilant screening and early treatment of PCC-PGL in patients with CCHD are crucial in order to avoid significant morbidity and ensure a good quality of life.