Pheochromocytoma Crisis

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摘要：

Pheochromocytomas and paragangliomas (PPGLs) are rare neuroendocrine tumors derived from neural crest-derived cells of the sympathetic and parasympathetic nervous systems. Pheochromocytoma crisis (PCC) was defined as an acute catecholamine-induced hemodynamic instability causing multiple organ dysfunction. Pheochromocytomas are approximately ten times more frequently related to PCC compared with paragangliomas. The heart is particularly susceptible to damage from acute elevated catecholamine concentration, and profound myocardial depression with cardiogenic shock may occur. Clinical spectrum of organ dysfunction in PCC involved cardiac, respiratory, neurological, renal, hepatic, gastrointestinal, metabolic, musculoskeletal, and vascular dysfunction. Precipitants of PCC included insufficient/compromised tumor blood supply, direct physical stimulus to tumor, indirect physical stimulus to tumor, general anaesthesia/ nonadrenal surgery, drugs and pregnancy. Steroid-induced PCC and pheochromocytoma in pregnancy warrants special consideration. Up to 65% of practitioners fail to diagnose pheochromocytoma in their pregnant patients before delivery despite the high maternal and fetal morbidity and mortality rates (40.3% and 56%, respectively). The common diagnoses mimicked by PCC with associated misleading clinical features were acute coronary syndrome/ myocardial infarction, cardiomyopathy andcardiogenic shock, septic shock, pre- eclampsia or amniotic fluid embolism, and so on. The incidence of PCC in pheochromocytoma was around 7-18%. Multidisciplinary interventions, included intra-aortic balloon pump, extracorporeal membrane oxygenation (ECMO), continuous hemodiafiltration, and neuroprotective

therapeutic hypothermia, were needed for medical stablization. Alpha-blockade is the
most widely accepted method of presurgical medical preparation. In almost all cases,
it would only be appropriate to attempt surgery after medical stabilization has been
achieved.