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Introduction The concept of benign and malignant Pheochromocytoma/Paraganglioma has been replaced in the last WHO classification by the idea that all tumors could have some metastatic potential. The risk of malignancy is reported to be extremely low for patients affected by Von Hippel-Lindau disease. Methods We demonstrate the case of a female patient affected by Von Hippel-Lindau disease, which presented six years after right retroperitoneoscopic adrenalectomy at our institution, with increased blood pressure and typical “attacks” that required further investigation for suspected recurrent Pheochromocytoma. The biochemical testing revealed increased circulating noradrenalin levels and a DOTATOC-PET/CT detected pathological lymphonodular, lung and bone uptake congruent to metastatic disease. Results The paracaval and paraaortal nodes situated at the level of the renal vessels were removed by the retroperitoneoscopic route while the infraduodenal interaortocaval node was resected laparoscopically. The procedure was started by redo right retroperitoneoscopy. The operating time was 105 minutes. There were neither intraoperative nor postoperative complications. The blood loss was <10 ml. Histology confirmed retroperitoneal soft tissue metastasis of Pheochromocytoma. Conclusions Minimally invasive treatment can be offered to patients with metastatic disease. In this particular case combination of posterior and anterior approach was used showing that both routes should be part of the surgical armamentarium of a dedicate surgeon.