
Recurred Malignant Paraganglioma with Liver and Lung Metastasis 17 Years after Resection of Primary Retroperitoneal Paraganglioma

고려대학교 의과대학 외과학교실

최새별, 한형준, 김정윤, 이진석, 김동식, 김완배, 송태진, 서성옥, 김영철, 최상룡

Introduction: Paraganglioma is a neoplasm derived from chromaffin-affinity tissue or parasympathetic ganglion cells that was known as extra-adrenal pheochromocytoma. Most paraganglioma are generally reported to be benign, with approximately 10% of them malignant. It is difficult to distinguish benign tumors from malignant tumors histologically. The only definite criteria for malignancy is the presence of metastasis, however, tumor necrosis, microscopic vascular invasion, and extensive capsular invasion are more common in malignant tumor. There were a few reports about malignant paraganglioma with hepatic metastasis after surgery of primary adrenal lesion. We present herein an interesting case in which hepatic and lung metastasis of malignant paraganglioma was diagnosed 17 years after surgical resection of a primary lesion of the retroperitoneum.

Case Report: The 43-year-old man was referred to our hospital because of huge retroperitoneal mass, multiple liver masses and solid pulmonary nodule on right upper lung zone found 1 month ago during evaluation of diffuse abdominal pain that was recently developed. He underwent retroperitoneal tumor excision 17 years ago due to right-sided retroperitoneal paraganglioma. Review of system and physical examination were normal. Laboratory data and all tumor markers were within normal limits. Urine VMA was 18.61 mg/day (normal: <8 mg/day) and metanephrine was 9.415 mg/day (normal: 0~1.3 mg/day). Chest x-ray revealed 3.9cm-sized nodule in right upper lung. Abdominal computed tomography revealed Infiltrative bulky retroperitoneal tumor is noted in the left with invasion to the left psoas muscle and left diaphragmatic crus and other mass lesion is also noted in the right retroperitoneal area with invasion to the right diaphragmatic crus. Chest computed tomography showed 4.7 cm-sized mass in right upper lung and several small nodules in right lower lung and both lower lung zone. Laparotomy demonstrated that huge retroperitoneal tumor that was infiltrating surrounding structures. Left kidney and adrenal gland were adhered to the mass and right adrenal gland also firmly adhered to the mass. En-bloc resection of retroperitoneal tumor with surrounding structures was performed. Final pathologic reports revealed recurrent paraganglioma in perirenal soft tissue and retroperitoneum. Postoperative course was uneventful. Postoperative adjuvant low-dose ¹³¹I-metaiodobenzylguanidine (MIBG) was performed.