Multiple endocrine neoplasia type 2 with malignant pheochromocytoma —Long term follow-up of a case by ¹³¹I-metaiodobenzylguanidine scintigraphy—

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The case of a 33-year-old Japanese man, who has Multiple Endocrine Neoplasia Type 2 (MEN IIa) (Sipple's syndrome) with malignant pheochromocytoma, is reported. He had survived for twelve years since the initial diagnosis of malignant pheochromocytoma. Within this period, he had undergone ¹³¹I-meta-iodobenzylguanidine scintigraphy twice, in 1983 and 1990. This is the first case in Japan of a longterm surviving patient with malignant pheochromocytoma followed up by ¹³¹I-MIBG scintigraphy. Although he had no exacerbation of clinical symptoms or urinary catecholamine levels, second scintigraphy clearly showed an increase in the tumor size, new metastasis of the malignant pheochromocytoma and exacerbation of the medullary thyroid carcinoma. Compared with any other roentgenological device and hormonal data, ¹³¹I-MIBG scintigraphy was seen to be a good tool for evaluating the localization and the progression of tumors. ¹³¹I-MIBG scintigraphy is a useful procedure not only for initial diagnosis but also for judging progression in a case of advanced malignant pheochromocytoma.

Key words: malignant pheochromocytoma, ¹³¹I-MIBG, MEN type IIa, long term survival