Multiple endocrine neoplasia type 2 with malignant pheochromocytoma
—Long term follow-up of a case by $^{131}$I-meta-iodobenzylguanidine 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 $^{131}$I-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 $^{131}$I-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 exacer-
bation of the medullary thyroid carcinoma. Compared with any other roentgenological
device and hormonal data, $^{131}$I-MIBG scintigraphy was seen to be a good tool for evaluating
the localization and the progression of tumors. $^{131}$I-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, $^{131}$I-MIBG, MEN type IIa, long term survival