

Nonfunctioning Huge Malignant Pheochromocytoma with Liver Metastasis

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A 27-year-old woman with a 4-month history of right upper quadrant dull pain and normal blood pressure was admitted in May 2001. Abdominal ultrasonography and abdominal computed tomography revealed a huge retroperitoneal mass with hepatic nodules. Histologic diagnosis was paraganglioma based on gun biopsy. Hormonal study for pheochromocytoma was negative. I-metaiodobenzylguanidine (MIBG) scintigraphy and angiography were performed for operation. The tumor was removed en bloc with part of the pancreas, spleen and hepatic mass. Pathologic examination with immunohistochemical staining revealed a malignant pheochromocytoma growing exophytically from the left adrenal and metastasizing to the liver. The postoperative course has been uneventful and no recurrence has been noted over a 2-months follow-up period. (*J Korean Surg Soc* 2002;63:345-349)

Key Words: Malignant pheochromocytoma, Nonfunctioning pheochromocytoma, Asymptomatic pheochromocytoma

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0.1% 가 가
 (1)
 10%
 가
 가 (2)
 , I-me-
 taiodobenzylguanidine (MIBG) scintigraphy
 (3,4)

 27 4
 가
 가 6 , 7 , 8
 가
 가
 B , C 24
 (Table 1).
 CA 129 97.5 U/ml, AFP 3.6 ng/ml, CEA
 0.9 U/ml, CA 19-9 4.9 CA 129 가
 가

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(Fig. 1). 8

가

chromogranin , S-100
sustentacular 가

MIBG scintigraphy cold area가 MIBG
tumor uptake neuroendocrine

MIBG 가

(Fig. 2).

Table 1. The patient's endocrinologic laboratory value

	Hormone	Level	Normal
Serum	VMA* (ng/ml)	6.1	3 9
	epinephrin (pg/ml)	23.00	0 120
	norepinephrin (pg/ml)	116.7	100 410
	dopamine (pg/ml)	47.1	0 30
24 hr. urine	epinephrine (μg/day)	4.6	0 40
	norepinephrine (μg/day)	17.8	0 80
	metanephrine (mg/day)	0.27	52 341
	VMA (mg/day)	1.84	2 10

*VMA = vanillyl mandelic acid.



Fig. 1. Enhanced abdominal CT scan shows retroperitoneal huge mass and lymphatic invasion with liver metastasis.

6 , 7 , 8

(Fig. 3A, B).

fibrous capsule
15 × 16 × 11 cm, 1,600 g

가

chromogranin synaptophysin . S-100
sustentacular cell zellballen

(Fig. 4).

mitochondria (), lipid vacuole ()
가

adrenalin mitochondria () 가
(Fig. 5).

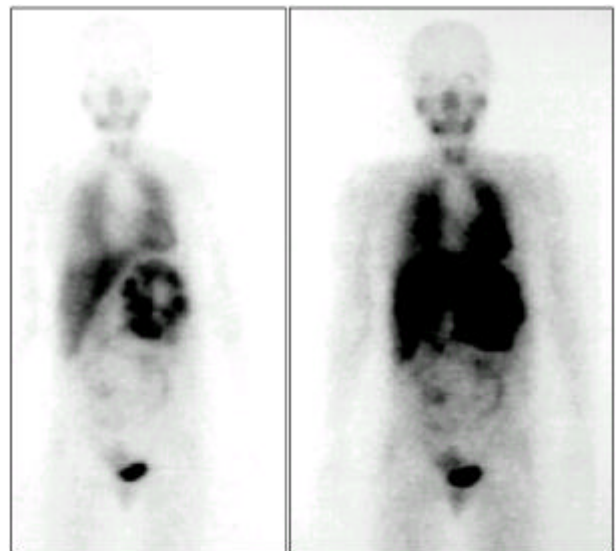


Fig. 2. MIBG scintigraphy shows MIBG uptake of large mass on left upper quadrant with central cold area and hepatic nodule.

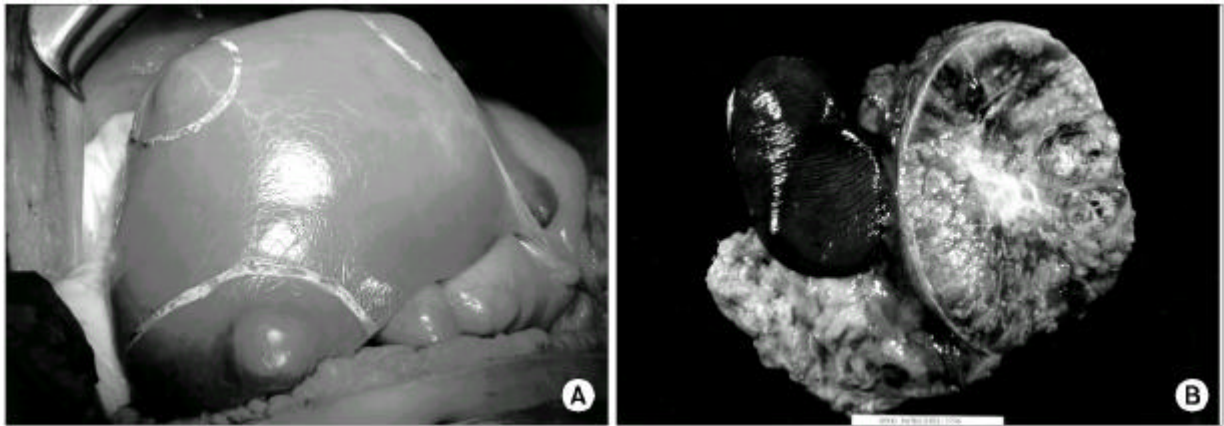


Fig. 3. In operative field, multiple liver metastatic nodules are noted (A), a huge left adrenal mass is adhered to spleen and pancreatic tail (B).

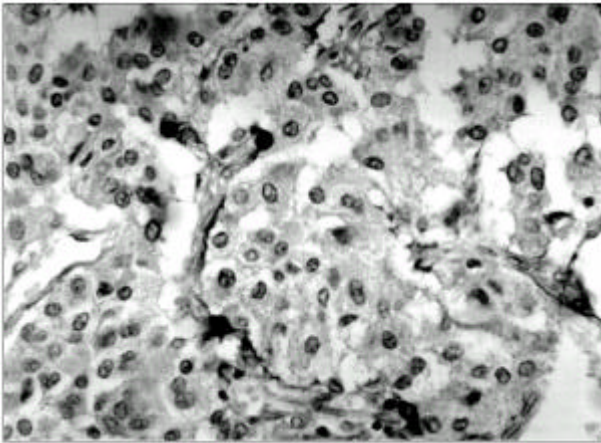


Fig. 4. The tumor nests are supported by thin fibrovascular stroma with S-100 positive sustentacular cells (S-100 × 400).

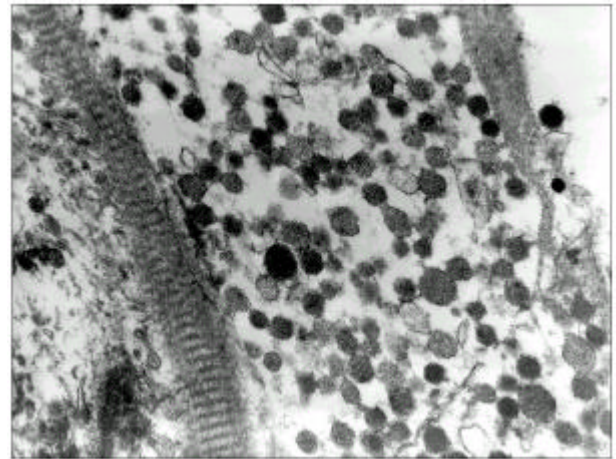


Fig. 5. Electron micrograph of liver mass shows numerous membrane bound dense core epinephrine type granules, ranging from 128nm to 276.8nm in diameter, Luse body is noted at the periphery of nests (EM original magnification × 17,000).

MIBG
 가
 symphoadrenal system
 dichromate 'dusky'
 가
 paraganglioma
 (, organ of Zuckerkandl)

가 가 .(5)
 1,000 1
 . Mayo Clinic
 13.1% 72.3%가
 8 .(6) 가
 PASS (Pheochromocytoma of the adrenal gland scaled score)가 .(7)
 가
 (1), (1),
 (2), large nests or diffuse growth (2), focal of confluent necrosis (2), high cellularity (2), tumor cell spindling (2),

cellular monotomy (2), increased mitotic figures (2), atypical mitotic figures (2), profound nuclear pleomorphism (1), hyperchromasia (1) 가 , 4 가

2 14% .(8,9) Mornex (10) 34 14 case 가 가 Crout Sjoerdsma(11)

50 g , 50 g 가 metaiodobenzylguanidine scintigraphy (MIBG) scintigraphy 가 .(12,13) , Maurea (14) 가 MIBG uptake

가 가 MIBG scintigraphy, 가 가 ¹³¹I-metaiodobenzylguanidine scintigraphy 가 6-[18F]fluorodopamine positron emission tomography (PET) scanning .(15)

MIBG 가 (3.7) (9.0) .(17)

가 50% .(18) 가 가

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