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CASE REPORT

Composite pheochromocytoma with a malignant peripheral nerve sheath tumor: Case report and review of the literature

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Summary Adrenal tumors with more than one cellular component are uncommon. Furthermore, an adrenal tumor composed of a pheochromocytoma and a malignant peripheral nerve sheath tumor is extremely rare. A composite pheochromocytoma with malignant peripheral nerve sheath tumor in a 42-year-old man is reported here. After adequate preoperative control, left adrenalectomy was performed simultaneously with resection of the ipsilateral kidney for spontaneous rupture of the left adrenal tumor. Pathological findings demonstrated pheochromocytoma and malignant peripheral nerve sheath tumor in a ruptured adrenal tumor. To date, there have been only four reported cases of composite pheochromocytoma with malignant peripheral nerve sheath tumor, so the present case is only the fifth case in the world. Despite the very poor prognosis of patients with pheochromocytoma and malignant peripheral nerve sheath tumors reported in the literature, the patient remains well without evidence of recurrence or new metastatic lesions at 36 months postoperatively.

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1. Introduction

The rare coexistence of a pheochromocytoma with neuroblastoma, ganglioneuroblastoma, ganglioneuroma, or malignant peripheral nerve sheath tumor (MPNST) is termed a composite pheochromocytoma.[1] Bolande[2] designated this pathologic mechanism as neurocristopathy, based on the concept that various kinds of tumors originate from developmental arrest and maldevelopment of totipotential neural crest cells. An adrenal tumor composed of pheochromocytoma and MPNST is extremely rare.[1,3–5] To date, only four cases of composite pheochromocytoma with MPNST have been reported.[1,3–5] An unusual case of composite pheochromocytoma with MPNST in a 42-year-old man who was treated surgically is reported here.

2. Case report

In August 2009, a 42-year-old man presented with acute left low back pain. Abdominal and pelvic computed tomography (CT) scan revealed an elliptic, 112 mm $\times$ 88 mm^2, adrenal tumor with massive retroperitoneal hemorrhage (Fig. 1A and B). Magnetic resonance imaging suggested an adrenal lesion with T1 hypointensity and T2 hyperintensity. Preoperative serum and urinary concentrations of catecholamines and catecholamine metabolites were as follows: serum epinephrine 2883 pg/mL [normal range (NR) $\leq$100 pg/mL], serum norepinephrine 36,829 pg/mL (NR 100–450 pg/mL), serum dopamine 761 pg/mL (NR $\leq$20 pg/mL), serum vanillylmandelic acid 43.4 ng/mL (NR 4–5 ng/mL), urinary epinephrine 536.4 μg/d (NR 3.4–26.9 μg/d), urinary norepinephrine 9234.0 μg/d (NR 48.6–168.4 μg/d), urinary dopamine 289.6 μg/d (NR 365.0–961.5 μg/d), and urinary vanillylmandelic acid 27.4 mg/d (NR 1.5–4.3 mg/d). Other laboratory data demonstrated that this adrenal tumor was not hormonally active except for catecholamines. On physical examination, no evidence of neurofibromatosis type 1, such as neurofibromas over his entire skin and café au lait spots, was found.

Finally, a definite diagnosis of a pheochromocytoma was made based on the abovementioned results. After adequate preoperative control, in September 2009, open left adrenalectomy with simultaneous ipsilateral nephrectomy was performed because of posthemorrhagic adhesions.

Macroscopically, the adrenal tumor appeared as a yellow–brown myxoid tumor with hemorrhage and necrosis on cross-section, measuring 90 $\times$ 70 mm^2. Histologically, this tumor consisted of two different components. The main tumor cells forming predominantly zellballen-type nests were typical of a pheochromocytoma (Fig. 2A). These tumor cells were immunoreactive for chromogranin, synaptophysin, CD56, CD57, and, focally, S100 protein. Furthermore, separated from the domain of pheochromocytoma, oval to spindle cells arranged in a streamy fasciculated pattern (Fig. 2B). These spindle cells were positively stained for S100 protein and were positive for Ki-67 within the range of 10–40%. These histological findings were consistent with the features of MPNST. Thus, we concluded a composite pheochromocytoma with MPNST in a ruptured adrenal tumor. Microscopic positive margins were absent.

This patient has been followed regularly and has not undergone any adjuvant therapy. There is no evidence of recurrence or new metastatic lesions on CT and bone scan at 36 months postoperatively.

3. Discussion

During embryogenesis, certain cells in the neural crest migrate ventrally and give rise to the essential cellular components of sympathetic ganglia and adrenal medulla.[3] Migration disturbance and developmental arrest or maldevelopment of neural crest-derived cells may result in various types of neoplasms.[5] Bolande[2] first described the concept of neurocristopathies. Tumors composed of various combinations of varying stages of differentiation have been reported as part of neurocristopathies.[3–5] Furthermore, in a pheochromocytoma and MPNST tumor, sustentacular cells in the stroma of the pheochromocytoma have been suggested as the origin of MPNST recently.[1,4,5] Ch'ng et al[1] suggested that the morphological transition from the pheochromocytoma component to

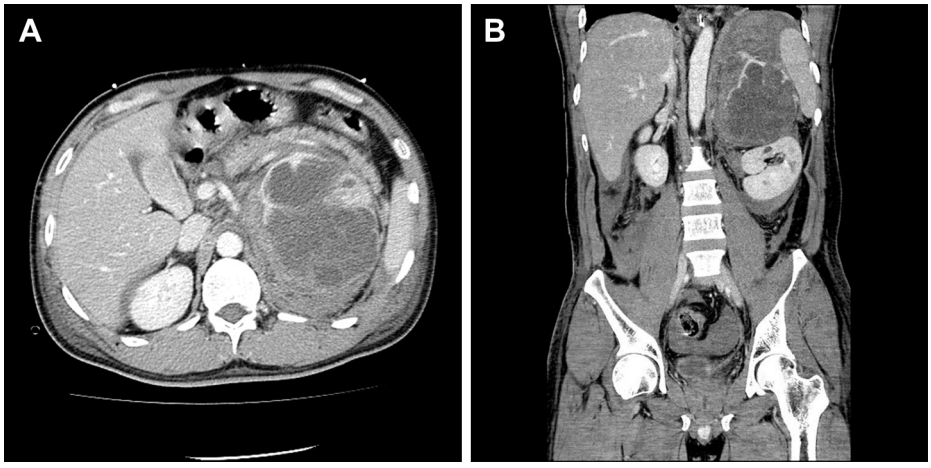

Figure 1 Preoperative computed tomography findings showed the left adrenal lesion with enhancement and large hemorrhage: (A) transverse plane and (B) coronal plane.