Interdisciplinary Neurosurgery: Advanced Techniques and Case Management 10 (2017) 130–134

Contents lists available at ScienceDirect

Interdisciplinary Neurosurgery: Advanced Techniques and Case Management

journal homepage: www.inat-journal.com

Cerebral metastasis of malignant pheochromocytoma 28 years after of disease onset☆

Makiko Miyahara, M.D. [a,b,*], Kouichiro Okamoto, M.D. [a,b], Ryuichi Noda, M.D. [a], Masato Inoue, M.D. [a], Akiyo Tanabe, M.D., Ph.D. [c], Tetsuo Hara, M.D., Ph.D. [a,b]

[a] Department of Neurosurgery, Center Hospital of the National Center for Global Health and Medicine, Tokyo, Japan
[b] Course of Advanced and Specialized Medicine, Juntendo University Graduate School of Medicine, Tokyo, Japan
[c] Department of Endocrinology, Metabolism, and Diabetes, Center Hospital of the National Center for Global Health and Medicine, Tokyo, Japan

ARTICLE INFO

Article history:
Received 31 July 2017
Accepted 6 August 2017
Available online xxxx

Keywords:
Brain metastasis
Malignant pheochromocytoma
Treatment

ABSTRACT

Background: Malignant pheochromocytoma is a very rare disease; moreover, brain metastasis is extremely rare, and there have been only a few published reports. Therefore, there are no established methods of treatment as of yet. We describe an extremely rare case of a patient with malignant pheochromocytoma that metastasized to the brain who was treated 28 years after the onset of pheochromocytoma, and review the relevant literature.
Case description: A 58-year-old man who had first been diagnosed with malignant pheochromocytoma 28 years earlier presented with right hemiplegia. We performed initial magnetic resonance imaging that showed a coin-like enhanced region in the left deep parietal lobe. His symptoms were stable after stereotactic radiosurgery was performed; however, the tumor size had not changed. Twelve months later, a new necrotic area, likely a result of irradiation, had grown in the peritumoral region. His symptoms worsened day by day, prompting us to perform craniotomy and total tumor removal. His catecholamine levels remained stable, and there were no perioperative events. His condition rapidly improved and there were few sequelae.
Conclusion: Cerebral metastasis from malignant pheochromocytoma is extremely rare; surgery of patients with low catecholamine levels may be performed safely.

1. Background

Malignant pheochromocytoma (PCC) can metastasize to multiple organs. However, cerebral metastases in malignant PCC are exceptionally rare. Metastases mostly involve the skeletal system, liver, lung, and regional lymph nodes [1–3]. To our knowledge, this is the first case report to provide radiotherapy treatment and operation details for malignant PCC and related brain metastasis. We also summarize previous reports and review the relevant literature.

2. Case report

A 57-year-old-man presented with headache, right hemiparesis, aphasia, and dysarthria. He had been diagnosed as having left adrenal PCC 28 years earlier when he first presented with hyperthermia, and had undergone left adrenalectomy. Twenty-three years earlier, multiple metastases to both lungs had been discovered, which led to a diagnosis of malignant PCC. The initial operation for lung metastasis was discontinued because of blood pressure fluctuation. Subsequently, he had undergone a second (at age 50) and third (at age 51) operation for lung metastasis. Even though a small lesion in the left mediastinum remained, his catecholamine level and blood pressure were well controlled.

In the present examination, we performed an initial brain magnetic resonance imaging (MRI) that revealed an enhancing lesion in the left parietal lobe, which was entirely enhanced on metaiodobenzylguanidine (MIBG) scintigraphy. On fluorodeoxyglucose positron emission tomography (FDG-PET) and methionine PET, there was accumulation at the center of the tumor (Fig. 1). During hospitalization, his symptoms improved. Catecholamine levels remained within normal limits (Table 1). After diagnosis of brain metastasis from these examinations, he chose stereotactic radiosurgery (SRS) because of his reluctance to undergo surgery, fearing the worsening of neurological symptoms. Brain SRS at a dose of 24 Gy in 1 fraction was performed. Subsequently, his symptoms remained unchanged.

Abbreviations: FDG-PET, fluorodeoxyglucose-positron emission tomography; MIBG, metaiodobenzylguanidine; MRI, Magnetic resonance imaging; PCC, pheochromocytoma; SRS, stereotactic radiosurgery.

☆ Conflict of interest: None.Disclosure of funding: None.

* Corresponding author at: Department of Neurosurgery, Center Hospital of the National Center for Global Health and Medicine, Toyama 1-21-1, Shinjuku-ku, Tokyo 162-8655, Japan.
E-mail address: mmiyahara@hosp.ncgm.go.jp (M. Miyahara).

http://dx.doi.org/10.1016/j.inat.2017.08.003

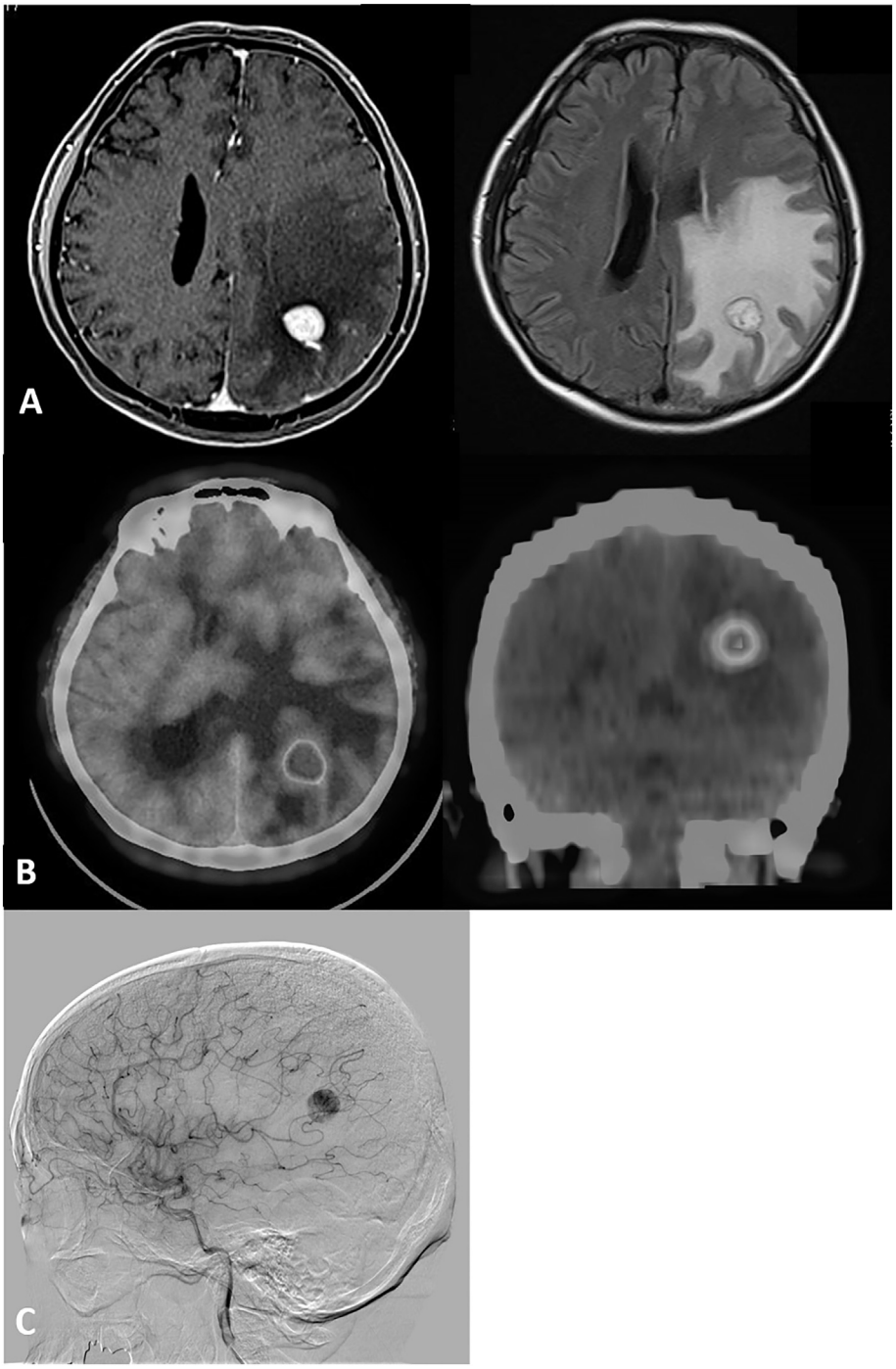

Fig. 1. A: T1-weighted magnetic resonance image enhanced by gadolinium (Gd)-diethylenetriamine penta-acetic acid demonstrating a strongly contrasted coin-like lesion in the left parietal lobe on admission (left); FLAIR MRI also showing severe peritumoral edema (right). B: Methionine positron emission tomography (PET) image (left) and metaiodobenzylguanidine fluorodeoxy scintigraphy (right) demonstrating high accumulation in the tumor. C: Left internal carotid artery angiogram showing marked vascular staining via a branch of the angular artery.

One year later, his aphasia and hemiplegia had rapidly worsened; MRI and FDG-PET showed radiation necrosis in the peritumoral area and marked worsening of peritumoral edema (Fig. 2A). To determine the exact diagnosis and relieve his symptoms, total tumor removal was performed 1 month after his second hospitalization. Presurgical tumor vascular embolization for bleeding control was attempted, because the tumor was receiving a rich blood flow. However, we had to abandon the attempt, because the tumor was fed from the distal angular artery which was difficult to cannulate.

2.1. Operation

Ten days before surgery, a sufficient amount of alpha blocker (doxazosin increased from 2 mg/day to 32 mg/day) was administered. Nifedipine was prepared for hypertension during operation, and a large craniotomy was performed. A high parietal corticotomy was performed, and brain parenchyma lying between the cortex and the surface of the tumor was removed, in order to mitigate the edema that had developed. Thickening and reddening of the draining vein in the posterior