Pituitary Infundibulum Hemangioblastoma Detected by Dynamic Enhancement MRI

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Hemangioblastomas in the pituitary stalk are extremely rare; only 13 cases have been described in the past.1-6 Detected by a dynamic contrast-enhanced magnetic resonance imaging (MRI), a case of pituitary infundibulum hemangioblastomas (HBL) was reported by us with tumor volume only 0.016 cm³.

CASE REPORT

A 28-year-old woman was admitted to our hospital with a 27-month history of galactorrhea and alopecia. No obvious tumor on the pituitary infundibulum was found by a cranial MRI (Figure 1), however, it indicated multiple masses in the vermis cerebellum, medulla oblongata, and dorsal portion of thoracic spinal cord 1-2 and syringomyelia from cervical to thoracic spinal cord. Previous medical history showed she was healthy and she was on no medications. Her menstruation was regular without a history of oligomenorrhea and no diabetes insipidus was found. On admission, the neurological examination revealed her slight dissociated sensory loss and slight weakness on the left hand. Her left extremities’ strength was V- The ophthalmological examination was normal and abdominal computer tomography scans indicated a pancreatic cyst. Von Hippel-Lindau (VHL) disease was diagnosed based on the clinical criteria. Serum hormone levels including total thyroxine 3, total thyroxine 4, thyroid stimulating hormone, free thyroxine 3, free thyroxine 4, estradiol, progesterone, human growth hormone, and testosterone were in the normal ranges. The concentration of prolactin in the serum was slightly elevated (28.35ng/ml, normal range 1.39-24.20ng/ml), luteinizing hormone (0.38mIU/ml, normal range 0.40-20.00IU/ml), follicle-stimulating hormone (1.43mIU/ml, normal range 2.00-13.00IU/ml), cortisol (23.66ng/ml, normal range 50.00-250.00ng/ml) were slightly decreased.

A suboccipital craniotomy and a C1, T1-T3 laminectomy were performed. Macroscopically, lesions in the vermis cerebellum, medulla oblongata and thoracic spinal cord 1-2 were explored and removed completely. Hemangioblastomas were confirmed through pathologic study.

Symptoms of galactorrhea and alopecia of the patient still existed postoperatively. Magnetic resonance imaging (including dynamic contrast enhancement MRI with thin coronal scans through the sella) was performed at one week and three months after the surgery. On the second dynamic contrast-enhanced MRI, (Figure 2 a-d) a tiny enhanced mass with volume 4x2x2 mm³ was detected on the pituitary infundibulum. Based on the clinical data and criteria,6 it was diagnosed as pituitary infundibulum hemangioblastoma. Three months after the surgery serum hormone levels including total thyroxine 3, total thyroxine 4, thyroid stimulating hormone, free thyroxine 3, free thyroxine 4, luteinizing hormone, follicle-stimulating hormone, estradiol, progesterone, human growth hormone, cortisol, and testosterone were within normal ranges; the concentration of prolactin in the serum (29.44ng/ml, normal range 1.39-24.20ng/ml) was still slightly elevated. She was prescribed 2.5 mg/day of bromocriptine. Two weeks later the symptom of galactorrhea was relieved to some degree, and the concentration of prolactin in the serum dropped to 19.23ng/ml (normal range 1.39-24.20ng/ml).

DISCUSSION

Compared with pituitary stalk HBLs reported so far1-6 the tumor volume in our case was the smallest. On MRI the tumor...
could be found just to the left of the pituitary infundibulum. We could easily clarify its origin, where the tumor arose from in the pituitary stalk, because of its tiny volume.

In the Lonser’s series⁶ these tumor volume ranged from 0.08 to 2.8 cm³, however, no patients presented with or developed signs or symptoms attributable to a pituitary stalk tumor and laboratory endocrine profiles of all patients were normal. In our case HBL volume was only 0.016 mm³ and the patient complained of the galactorrhea and alopecia that was thought as a presentation of hyperprolactinemia and hypopituitarism. Some women have excess prolactin secretion but with normal pituitary MRI.⁷ Our patient was in a different situation: her symptoms of galactorrhea, increase of prolactin secretion, and pituitary stalk lesion shown by the pituitary MRI corroborated each other. Therefore we speculated that the small tumor might be the etiology of the hyperprolactinemia. From a functional point of view, the tumor, even when it was very small, had influenced stalk input of hypothalamic dopamine.

In our case the tumor was too small to catch by a conventional enhancement MRI. It was the dynamic contrast enhancement MRI which helped capture the image of the tumor. Dynamic contrast enhancement MRI is performed after the administration of intravenous contrast medium to noninvasively access tumor vascular characteristics.⁸ It can provide useful hemodynamic information about intra-axial brain tumors that is not provided by standard MRI and therefore contributes to the differential diagnosis.⁹ The normal pituitary stalk lacks a blood-brain barrier, and it enhances intensely with administration of gadolinium.¹⁰ However, being a vascular tumor, hemangioblastoma has an immediately and more quickly increasing enhancement after injection of gadolinium; the hemangioblastoma’s enhancement happens earlier than the pituitary stalk’s when using dynamic
contrast MRI. Therefore, dynamic contrast enhancement MRI has the capability to distinguish a tiny hemangioblastoma from the pituitary stalk.

Excluding the case presented in Neumann’s report\(^3\) in which details were not described, previous cases of pituitary stalk hemangioblastomas took surgical resection or observation without surgery.\(^1\)\(^6\) In our case we chose medical treatment. Oral bromocriptine brought relief of symptoms to the patient.

Pituitary infundibulum hemangioblastoma can affect the function of the pituitary gland, even when tumor volume is very small. For hemangioblastomas, particularly in VHL disease, dynamic contrast enhancement MRI with thin coronal scans through the sella is necessary when patients present pituitary dysfunction symptoms. Oral bromocriptine can control the symptoms of hyperprolactinemia caused by pituitary infundibulum hemangioblastoma.

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REFERENCES