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Case report

Pituitary stalk hemangioblastoma: The fourth case report and review of the literature

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Abstract

Supratentorial hemangioblastomas (HBL) have been rarely described in the literature. Herein we report the fourth case of pituitary stalk HBL diagnosed concurrently with cerebellar HBLs in a 51-year-old woman with von Hippel-Lindau disease. Complete resection of the lesion was achieved using left frontopterional craniotomy and no recurrence was observed after 8 years of follow-up. The clinical presentation, radiological features, pathological findings, and the management of this particular case are discussed in the light of the available literature. To our knowledge, this case is the only pituitary stalk HBL with total resection, and a long follow-up fully documented.

Keywords: Pituitary stalk tumor; Sellar and suprasellar neoplasm; Hemangioblastoma; von Hippel-Lindau disease; Tumor; Radiosurgery

1. Introduction

Hemangioblastomas (HBLs) represent about 1.5–2.5% of all primary intracranial tumors and 7–12% of posterior fossa tumors [1,2]. The most common site of occurrence is the cerebellum; however, they may also occur in the medulla oblongata and spinal cord [3,4]. In 20% of cases, they are associated with von Hippel-Lindau (VHL) disease which is an autosomal dominant syndrome resulting from a germline mutation of the VHL tumor suppressor gene on the short arm of chromosome 3. Patients affected can develop various benign or malignant tumors of the central nervous system, kidneys, adrenal glands, pancreas, and reproductive organs. In a series of 231 cases of VHL disease, Wanebo found 4% of supratentorial HBLs [5]. Pituitary stalk HBLs have been reported exceptionally; to date, only three cases have been described [2,6,7]. Herein we report the fourth case of pituitary stalk HBL with a literature review focusing upon sellar and suprasellar HBLs. In addition, the radiosurgical management of these tumors is discussed.

2. Case report

A 51-year-old woman was admitted to our hospital in 1998 with a 4-month history of progressive impairment of the vision in her right eye. Her medical history revealed that she had already lost her left eye's vision at the age of 18 (1965), following cobalt therapy for a retinal HBL. Five years later, she developed a progressive paraparesis accompanied by back pain and impaired sensation in both lower limbs. An intramedullar hypervascular mass was demonstrated at the T_4 level and was removed completely. Histological findings were consistent with features of HBL. She completely recovered postoperatively and returned to her job. At that time, based on her clinical syndrome, she was diagnosed with VHL disease.

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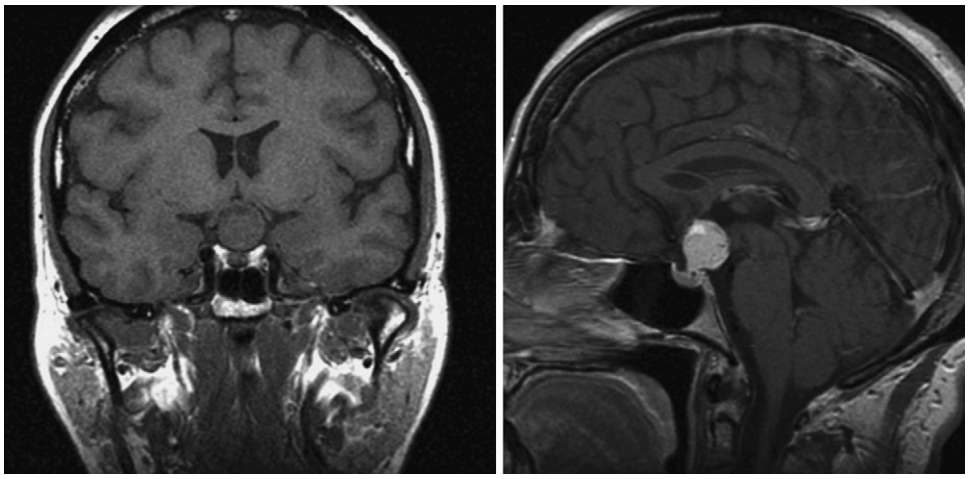

Fig. 1. Preoperative T_1-weighted MR images, coronal without contrast (left) and sagittal with contrast (right), demonstrating an isointense homogenous suprasellar mass which displaced the optic chiasm upwards. The mass strongly enhanced after gadolinium administration and appeared to involve the pituitary stalk.

The opthalmological examination using Goldmann and Humphrey perimetry of her right eye revealed a right inferior temporal quadranopsia. Ophthalmoscopy revealed mild optic disc pallor and excluded a retinal capillary angioma. T_1-weighted magnetic resonance imaging (MRI) revealed an isointense round suprasellar homogenous mass, 20 mm in largest diameter, which strongly enhanced with gadolinium. On T_2-weighted MRI, this mass was hyperintense appearing to arise from the pituitary stalk (Fig. 1) and displacing the optic chiasm superiorly and laterally. Four other infracentimetric lesions were observed in the cerebellum. The preoperative endocrinological profile was normal except a moderate increase of prolactine (80.7 ng/ml). The patient's medical history together with MRI findings suggested hemangioblastoma.

Using a left frontopterional craniotomy, the atrophic nonfunctional left optic nerve was severed to facilitate the access to the tumor. Macroscopically, the tumor appeared hyperemic and reddish without capsule or cyst. It arose from an inflated pituitary stalk. No adhesion to the dura mater was observed.

Histological findings revealed numerous endothelial forming vascular channels which were lined by foamy cells, consistent with the diagnosis of HBL (Fig. 2). A polymerase chain reaction (PCR) test was performed to amplify the entire open reading frame of the VHL gene, followed by sequencing (21M13 dye primer sequencing) for the exons 1–3. The test showed mutation on the exon 2 of the VHL gene (change of cysteine to thymine on the nucleotide 556 responsible for substitution of histidine to thyroxine (H115Y)). This confirmed a diagnosis of VHL.

Postoperatively, the vision of the right eye recovered completely after a few weeks. The resection of the pituitary stalk resulted in panhypopituitarism which was successfully managed with substitutive therapy. Clinically, she was very tired immediately after surgery, and her body weight increased from 56 to 62 kg but normalized within few months. She could return to her job 8 months later. There was no recurrence during a follow-up period of 8 years (Fig. 3). During the follow-up period, one more lesion was observed in the cerebellum making a total of five cerebellar lesions, two at the right side and three at the left side. Two of the cerebellar lesions grew to almost 20 mm in size within 4 years (Fig. 4A), and at that moment the patient complained of some episodes of headache and dizziness. Therefore, a stereotactic radiosurgery (SRS) was applied using Novalis system (BrainLab, Germany) for all the cerebellar HBLs. Six isocenters were used to cover the tumors because the radiosurgical team noticed one additional millimetric lesion. However, 18 months later, she showed symptoms of intracranial hypertension. The radiological work-up revealed the development of severe cerebellar edema (Fig. 4B) and hydrocephalus. Moreover, no decrease in lesions' sizes was noticed. She improved temporarily for a few weeks after endoscopic third

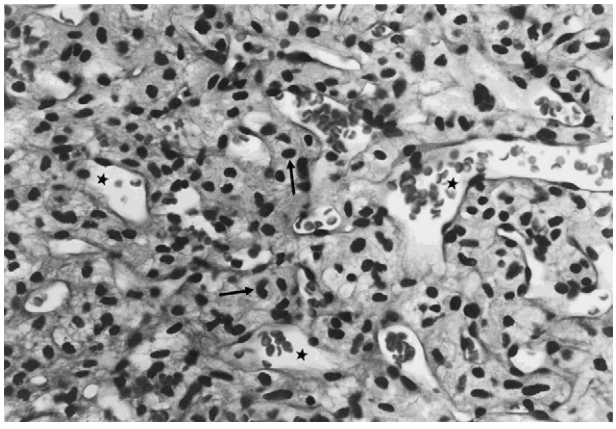

Fig. 2. Microscopic view of the lesion showing typical histological features of HBL: a rich vascular network (stars) surrounded by vacuolated stromal cells (arrows).