Sellar Hemangioblastoma Mimicking a Macroadenoma

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Abstract-

Purpose: Hemangioblastomas (HGB) are slow growing benign vascular tumors that arise almost always from the cerebellum and the spinal cord. Supratentorial location is extremely rare with approximately 130 cases published to date. We present a case of a sellar hemangioblastoma.

Case Report: An eleven year-old girl presented with a sellar and suprasellar tumor that seemed to be a macroadenoma. A transsphenoidal approach was attempted but excessive intraoperative bleeding made the resection not feasible. A second transcranial approach was successful in partially removing the lesion and decompressing the optic chiasm and the pituitary stalk. Pathological review revealed a sellar hemangioblastoma. Screening for Von Hippel Lindeau Syndrome was negative.

Conclusion: The preoperative diagnosis of HGB is extremely difficult in this case but would have been essential in order to choose the right surgical approach.

Key Words: Hemangioblastoma, von Hippel Lindeau, Transsphenoidal approach

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INTRODUCTION

Hemangioblastomas (HBL) are benign, slow growing vascular tumors that occur commonly in adults and arise mainly from the Central Nervous System[1,2]. They are often associated with VHL syndrome, a genetic multi-system disorder[3-5]. The most frequent locations of HGB are the cerebellum and spinal cord[6,7]. Supratentorial location is particularly uncommon and almost always associated with VHL. Only very few sellar HGB have been described in the literature to date[8-12].

The risk of intraoperative hemorrhage in these tumors is difficult to predict but if a preoperative diagnosis is available the surgical approach chosen should guarantee a strict control of intraoperative tumor bleeding[13].

We report an infrequent case of a sellar hemangioblastoma which was attempt to be resected by a transsphenoidal approach because of lacking an accurate preoperative diagnosis.

CLINICAL CASE

A 12-year-old female patient presented with headache and visual complains. Visual field test showed bitemporal hemianopsia. A Magnetic Resonance Image

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(MRI) was performed and revealed a sellar expanding lesion with homogeneous contrast enhancement and optic chiasm compression (Fig. 1, 2). Blood hormone analysis showed hypopituitarism. A transnasal surgery was proposed with a preoperative diagnosis of non-functioning pituitary adenoma, despite being extremely rare in pediatric population. A transsphenoidal approach was attempted but excessive intraoperative bleeding made the resection not feasible. A second transcranial approach was successful in partially removing a fragile, bleeding mass and in decompressing the optic chiasm and the pituitary stalk. Pathological review revealed an hemangioblastoma. Because of the frequent association of these lesions with Von Hippel Linedeau Syndrome (VHL) an abdominal MRI scan was performed and it ruled out renal cell carcinoma and pheochromocytoma. A genetic test was also carried out on peripheral blood leukocytes with no proof of VHL mutation. Patient's follow-up revealed marked improvement in her visual field and hormone values 3 months after surgery.

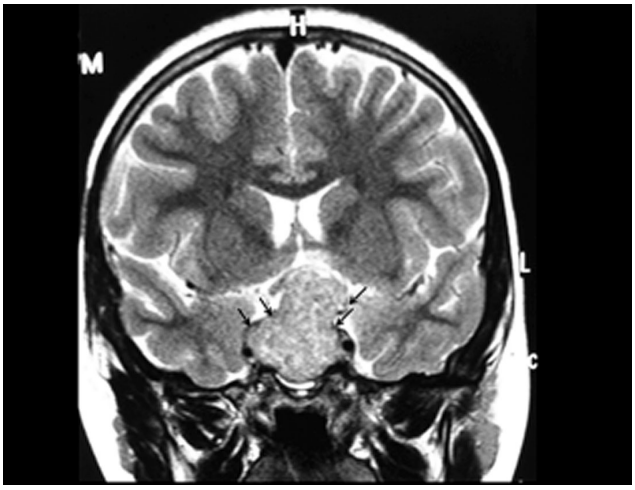

Figure 1. Coronal T2-weighted MRI, multiple arrows pointing a large sellar tumor, engorged vessels adjacent and inside the lesion can be identified

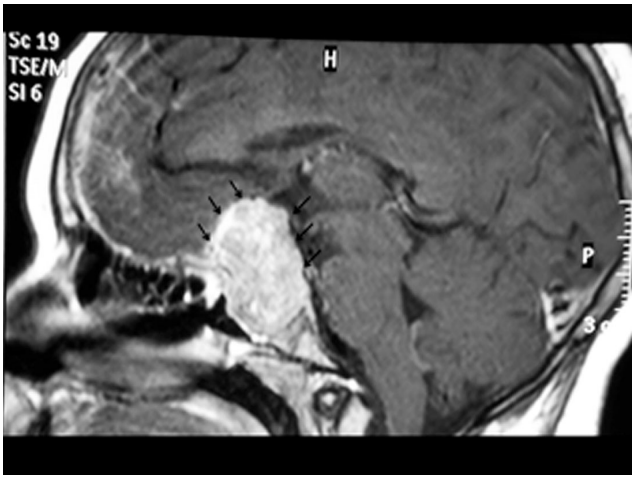

Figure 2. Sagittal contrast-enhanced T1-weighted MRI brain scan, multiple arrows showing a sellar and suprasellar tumor with homogeneous contrast enhancement.

DISCUSSION

Although pituitary adenoma was though as a probable diagnosis, this kind of tumors are extremely infrequent in pediatric population, and in the presence of a sellar mass in these patients, it must be taken in mind that the differential diagnosis should include germ cell tumor, Langherns' cell hisitiocytoma, chordoma and sarcoma. When the lesion arises from the sellar floor or the skull base, as in our case, the patient presents with panhypopituitarism or visual defect and it is more likely to be a chordoma or sarcoma. Furthermore, the enhanced MRI showed an image with dura-tail appearance, which is typical of meningioma. Thus, despite occurring almost always in elder patients, meningioma could be another possible diagnosis of this case.

Hemangioblastomas are benign tumors of uncertain histogenesis[14]. They represent 1 to 2.5% of all brain tumors[15] and are mostly located at the posterior fossa and spinal cord[16]. Approximately 25% of all HGB are associated with VHL disease[17]; being the supratentorial location especially linked with this entity, which makes this case remarkable .

Only very few cases of sellar HGB have been described in the literature to date[8-12]; three of them associated with VHL, a situation that favors the presumptive diagnosis of HBL and thus the choice of the optimal approach. Because of the imaging features of the current case, a sellar adenoma was considered as first diagnosis; therefore, a transsphenoidal approach was carried out. The resection by this route was not possible due to massive bleeding[11] and the procedure had to be ended. A

month later, after total recovery of the patient, a frontopterional approach with a subtotal resection of the suprasellar component and decompression of the optic chiasm was succesfully performed. The patient recovered the visual field and improved her degree of hypopituitarism.

If there are engorged drainage veins and strong enhancement on MRI, HGB may be suspected. These vessels are clearly shown on T2-weighted sequence. An useful option is to indicate an angiography, which can demonstrate the tumor vascularization in a very anatomical way, and to embolize it if it is allowed by the tumor features and its vascular supply. If the last one is not possible, radiosurgery with cyberknife or gammaknife may be an alternative option to treat this entity avoiding a massive intraoperative bleeding. In those patients in whom embolization was feasible, surgical resection may be done in a second step, when the tumor shrinks.

Correct surgical approach is mostly based on the location of the lesion and rarely the histological presumptive diagnosis influences surgical planning. However, HBG are an exception to this rule and minimally invasive procedures or transsphenoidal approaches should not be performed as first treatment options.

In reviewing the images we found small vessels inside the tumor already describe by Rumboldt et al[11], who describe a similar case in which a primary approach was used instead of a transnasal route based on preoperative image findings. A probable HGB diagnosis based on the MRI could have changed the surgical approach in our case.

In this patient, a HGB diagnosis was not suspected because no other lesions were present in the posterior fossa, signs and symptoms matched those of a pituitary adenoma and neither the patient nor her family had a record of VHL Syndrome.

CONCLUSION

The transsphenoidal surgery is a very safe approach for most sellar lesions. However, the surgical control in case of massive bleeding is very limited by this route. Therefore, high vascular sellar lesions must be thoroughly studied before a transsphenoidal approach is attempted in order to rule out a HGB diagnosis. VHL screening and a careful review of the images looking for intratumoral vessels represent highly useful tools. If there was a massive bleeding in the first surgery, as in our case, an angiography must be undertaken previously to a second surgical resection. It may exclude and prevent a vascular lesion, which would be a life-threatening one.

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