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Pituicytoma, About Two Cases and Review of The Literature

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Abstract: *Pituicytoma previously known as Infundibuloma or posterior pituitary astrocytoma, is a rare tumor of the sellar and suprasellar region. This tumor is considered as a benign lesion without any malignant transformation reported to date. Given its rarity, this lesion must be recognized and distinguished from the other most common suprasellar tumors. We report two cases of pituicytomas, clinically imitating a macroadenoma, with a brief review of the literature.*
Keywords: *pituicytoma, sellar region, endoscopic approach*

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I. Introduction:

Pituicytoma is an extremely rare low-grade glioma of the sellaire region. Pituicytes are specialized modified glial cells located in the neurohypophysis. These tumors can be easily confused with pituitary adenomas, craniopharyngiomas or meningiomas due to their location in the sellar and suprasellar region. In 2007, the pituicytoma was named a distinct entity according to WHO classification.

II. Material And Methods:

Case 1: A 45-year-old male, presented a decline in bilateral visual acuity for a year associated with endocrine disorders. The brain scan showed a hyperdense suprasellar mass with intense enhancement after contrast product injection. Diagnosis of meningioma a suprasellar was raised. An endoscopic biopsy was performed in this patient.

The pituitary gland appeared normal and was split to reveal a highly vascularized tumor, extending superiorly above the diaphragm sellae. A transcranial approach has been performed by the frontopterional approach. The microscopic study of the resected tissue revealed a well- circumscribed tumor, adherent to the pars tuberalis of the adenohypophysis. The post-operative consequences were straightforward. The histological study came back in favor of a pituicytoma.

Case 2: A 53-year-old male, who presented three years previously with endocrine disorders; impotence and decreased libido, with the onset two years later of a gradual decrease in visual acuity. The cerebral MRI had objectified an expansive intra and suprasellar process having been considered like a macroadenoma. The patient was first operated on by endonasal endoscopy, but given the very hemorrhagic nature of the lesion, the diagnosis of macroadenoma was ruled out, and the histologic study had come back in favor of a pituicytoma. The patient was operated on by the same approach with a total excision of the lesion. The postoperative effects were marked by a transient unilateral impairment of the III rd nerve seen in tumor extension to the cavernous sinus.

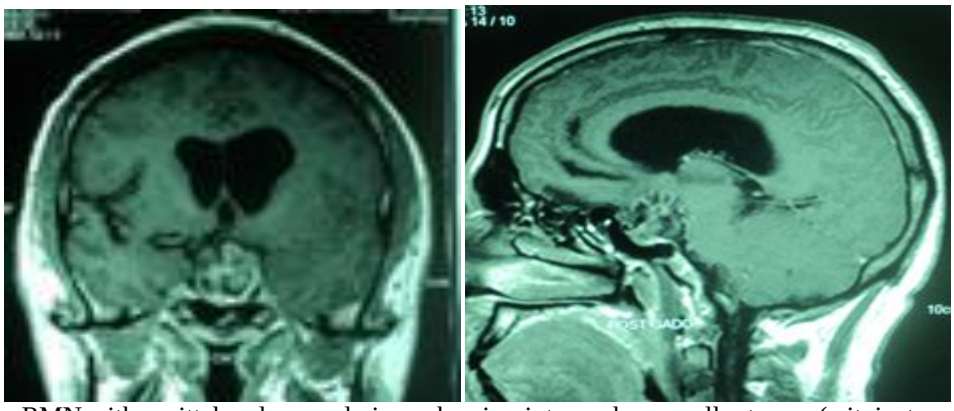

Fig:RMN with sagittal and coronal views showing intra and suprasellar tumor (pituicytoma).

III. Results:

The two patients were operated on, and benefited from partial resection for the first patient and subtotal resection for the second patient.

We observed an IIIrd nerve damage in the immediate postoperative period given the extension of the tumor towards the cavernous sinus; Improved visual function for both patients.

Both patients were followed for more than 06 years. Only one patient was referred for radiotherapy, currently the remaining tumor is stable.

IV. Discussion:

Pituicytomas are rare primary brain tumors originating from pituicytes in the neurohypophysis and the pituitary gland (1). The Pituicytes were brought to light by PC Bucy in 1932. These cells are responsible for the secretion of oxytocin and vasopressin.

Y. Takai classified the pituicytes in 4 types: (i) primary cells, (ii) dark cells, (iii) cancer cells, (iv) ependymal cells (2). Most pituicytomas are thought to originate from major cells and dark cells. The first case of pituicytoma was described by Scothorne in 1955. The term pituicytoma was proposed in 2000 by Brat et al (1). However, the tumor has been designated as a separate entity in the 2007 WHO classification (3, 4). A total of 78 cases of pituicytomas have been published in the literature until today (5). Those tumors occur mainly in adults, with an average age of 46.9 years old at diagnosis and a slight male predominance (1.7: 1).

The most common symptoms of pituicytomas are visual impairment (56.4%), headache (43.6%), hypopituitarism (21.8%) and sexual dysfunction (20.5%).

The diagnostic delay varies from a few months to several years (6). Acute presentation secondary to Intratumoral hemorrhage has been described. The preoperative diagnosis of pituicytomas is often difficult due to the lack of distinct radiological features differentiating them from other pituitary tumors.

Brain MRI is the imagery of choice for the diagnosis. Pituicytomas usually appear as solid, round, or well circumscribed oval masses in the sellar region, with or without suprasellar extension. They are generally hypo to isointense in T1 sequences, moderately hyperintense in T2 sequences, with homogeneous or heterogeneous contrast enhancement. The heterogeneous appearance, cystic or calcifications were rarely observed (7, 8).

Differential diagnoses include pituitary adenoma, meningioma, craniopharyngioma, hemangiopericytoma, and metastatic tumors.

Surgical resection is the treatment of choice for pituicytoma, with a rate of extremely low recurrences after complete resection (4.3%) (9). Intraoperatively, the tumors appear pink and solid, well defined and benign in appearance. Hypervascularization is a common characteristic to pituicytomas, which could hinder the performance of total tumor resection. Current surgical approaches include the transcranial and endoscopic endonasal approach (10). Feng et al reported a complete resection via an extended endonasal endoscopic approach (11). Recurrence after subtotal resection is frequent according to the literature. The interval to recurrence after subtotal tumor resection is usually long and no case of malignant transformation or cerebrospinal spread has been reported. Radiotherapy has been recommended after subtotal resection, but there is little evidence to suggest its benefit. The most common postoperative complications in this pathology are diabetes insipidus, hypopituitarism and worsening of visual function.

The definitive diagnosis of pituicytoma always depends on anatomopathological examination (12). Under the microscope, pituicytomas are composed of round spindle-shaped cells with a pattern of fascicular or storiform growth (13).

V. CONCLUSION

Pituicytomas are extremely rare, low-grade, slow-growing glial tumors. Transcranial resection or endoscopic endonasal surgery should be carried out based on the information provided by the brain MRI which must be read carefully. In addition, the good knowledge of the surrounding anatomical structures is crucial in order to facilitate a surgical resection as complete as possible of the tumor with the preservation of neurological functions. The role and effectiveness of radiotherapy and chemotherapy are not yet clear and require further study.

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