Case Report
Pituicytoma

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INTRODUCTION
Pituicytomas are very rare primary tumors of the neurohypophysis and can affect both the sellar and suprasellar regions. Few cases have been described in the literature[13]. Until now there are 31 cases reported, all of them are described in Table 1, which contribute to the poor characterization of the tumor and consequent diagnostic difficulties.

Pituicytomas originate from pituicytes, modified glial cells derived from ependymal lineage which are found in the stalk and posterior lobe of pituitary gland.[13,15]

On neuroimaging, pituicytomas are solid, uniformly contrast-enhancing masses. They are histologically low grade, featuring only mild nuclear atypia and no mitotic activity.[2] We describe here a case of pituicytoma diagnosed at our service and discuss it in relation to the data available in the literature.

CASE REPORT

Description
Clinical presentation: A 17-year-old boy was reported with a history of persistent headache and recent onset of visual disturbances. Examination revealed bitemporal...
Table 1: Summary of the reported 31 cases of pituicytoma

| Patient no. | Series (ref. no.) | Age (year)/sex | Presentation | Imaging | Resection | Follow up | Recurrence/complications | Radiation therapy |
|-------------|------------------|----------------|--------------|---------|-----------|-----------|--------------------------|------------------|
| 1           | Hurley et al., 1994 (7) | 26/F | Decreased visual acuity and hemianopsia | 2-cm enhancing sellar mass, T1-isointense, T2-hyperintense | TSP/STR | 3 year | None/transient Deficit | 5040 cGy |
| 2           | Brat et al., 2000 (2) | 55/F | Visual deficit | Suprasellar, enhancing | GTR | 1 year | None | None |
| 3           | Brat et al., 2000 (2) | 30/M | Headache | Suprasellar, enhancing | GTR | 1 year | None | None |
| 4           | Brat et al., 2000 (2) | 39/M | Headache | Solid and cystic, intrasellar, enhancing | TSP/GTR | 2 year | None | None |
| 5           | Brat et al., 2000 (2) | 42/M | Hypopituitarism/hemianopsia | Intrasellar, enlarged over 2 years of observation | TSP/STR | 2.5 year | 2 year progression with resection | None |
| 6           | Brat et al., 2000 (2) | 42/M | Visual deficit/decreased libido | Solid, suprasellar enhancing | STR | 1 year | Re-resection for recurrence x 2 at 5 and 15 months | None |
| 7           | Brat et al., 2000 (2) | 46/M | Hypopituitarism | Solid, suprasellar enhancing | GTR | 8 year | None | None |
| 8           | Brat et al., 2000 (2) | 83/F | Visual deficit | Suprasellar | GTR | 2 year | None | None |
| 9           | Brat et al., 2000 (2) | 48/M | Hypogonadism | 2-cm solid, suprasellar mass encasing vessels | GTR | 8 year | Recurrence at 5 months with subtotal re-resection | None |
| 10          | Brat et al., 2000 (2) | 51/F | Visual deficit | Solid, enhancing sellar mass consistent with adenoma | GTR | 4 year | None/transient hemiparesis | None |
| 11          | Schultz et al., 2001 (16) | 66/M | Decreased visual acuity, visual field deficit | 2-cm enhancing, T1-isointense, T2-hyperintense | TSP/GTR | 2 year | None | None |
| 12          | Cenacchi et al., 2001 (3) | 79/F | Hypopituitarism/visual disturbances | Unknown | TSP/GTR | 6 months | None | None |
| 13          | Figarella-Branger et al., 2002 (5) | 59/M | Hypopituitarism | Solid, enhancing | TSP/STR | 11 year | None | None |
| 14          | Figarella-Branger et al., 2002 (5) | 46/M | Decreased libido/hypogonadism | Solid, enhancing, suprasellar | GTR | 4 year | None/transient hemiparesis | None |
| 15          | Figarella-Branger et al., 2002 (5) | 58/M | Hypopituitarism/memory deficits | Solid, enhancing, mimicking posterior clinoid meningioma | GTR | 2 year | None/DI | None |
| 16          | Uesaka et al., 2002 (20) | 34/M | Decreased visual acuity | Solid, enhancing, T1-isointense, T2-hyperintense | TSP/STR | 3 months | None | None |
| 17          | Katsuta et al., 2003 (8) | 32/F | Amenorrhea/visual field defect | Intrasellar, isointense T1 and T2, enhancing | TSP/GTR | 2 year | None/DI | None |
| 18          | Ulm et al., 2004 (21) | 45/M | Decreased libido/low testosterone | 2-cm solid, enhancing, suprasellar | Craniotomy/STR | Unknown | None | Stereotactic radiation |
| 19          | Kowalski et al., 2004 (9) | 52/M | Panhypopituitarism | Solid, heterogeneously enhancing sellar/suprasellar mass | TSP/STR | 11 months | Recurrence | Fractionated radiation after recurrence | None |
| 20          | Shah et al., 2005 (17) | 32/F | Amenorrhea/headache | Heterogeneously enhancing posterior pituitary mass, T1-isointense, T2-hyperintense | TSP/STR | 5 year | Recurrence with re-resection TSP | None |
| Patient no. | Series (ref. no.) | Age (year)/sex | Presentation | Imaging | Resection | Follow up | Recurrence/ complications | Radiation therapy |
|------------|------------------|----------------|--------------|---------|-----------|-----------|----------------------------|------------------|
| 21         | Shah et al., 2005 (17) | 45/F          | Headache     | Enhancing sellar/suprasellar mass, T1-isointense, T2-hypointense | TSP       | Unknown   | Unknown                   | Unknown           |
| 22         | Chen, 2005 (4)    | 54/M          | Headache     | Enhancing sellar/suprasellar mass | TSP/STR   | 16 months | None                       | None              |
| 23         | Takei et al., 2005 (18) | 54/F          | Incidental at autopsy | None                     | None       | None       | None                       | None              |
| 24         | Nakasu et al., 2006 (12) | 42/F          | Amenorrhea   | Homogeneously enhancing sellar/suprasellar mass | Craniotomy/ STR | 5 year    | None                       | None              |
| 25         | Nakasu et al., 2006 (12) | 62/F          | Headache/fatigue | Homogeneously enhancing sellar/suprasellar mass | Craniotomy/ STR | 1.5 year | None/transient DI/hypopituitarism | None              |
| 26         | Benveniste et al., 2006 (1) | 47/M          | Hemorrhage/low LH/FSH | Hemorrhagic suprasellar mass with IVH | Craniotomy/ STR | None     | Unknown                   | None              |
| 27         | Gibbs et al., 2006 (6) | 64/M          | Bitemporal hemianopsia | Homogeneously enhancing 3-cm suprasellar mass, T1-isointense, T2-hyperintense, angiogram hypervascular capillary blush from ICA only | Cranio-orbitalzygomatic craniotomy/ GTR, very vascular | Unknown | Unknown                   | Unknown           |
| 28         | Thiryayi et al., 2007 (19) | 77/M          | Hypogonadism Quadrantanopia bitemporal inferior Suprasellar level | TSP/ STR | TSP/ STR | None       | None                       | None              |
| 29         | Wolfe et al. 2008 (22) | 71/F          | Decreased visual acuity and visual field defects | Solid, enhancing mass | TSP/STR | 1.5 year | None                       | None              |
| 30         | Orrego J. T. 2009(14) | 55/M          | Decreased libido Dysfunction erectile ginecomastia | Suprasella mass isointense on T1 | TSP/STR | None     | None                       | None              |
| 31         | Brandao and Braga et al. 2010 | 17/M          | Headache visual Disturbance | Contrast-enhance solid sellar | TSP/STR | 24 months | None                       | None              |

TSP: Transphenoidal Approach, STR: subtotal resection, GTR: gross-total resection

heteronymous hemianopsia and swelling of the left optic papilla. The remaining neurological exam was normal. Magnetic resonance imaging (MRI) of the brain revealed a contrast-enhancing, expansive solid sellar and suprasellar mass with an intermediate signal intensity on T1- and T2-weighted images, measuring about 2.6 cm × 1.6 cm × 1.5 cm [Figure 1]. The mass occupied the sella turcica, extending from the suprasellar cisterna and compressing optic chiasm and chiasmatic recess of the third ventricle. Endocrinological analysis demonstrated mild hyperprolactinemia (31.75 ng/ml, reference: 2.1−17.7 ng/ml), with the other pituitary hormones being normal.

**Surgery:** The patient was submitted to partial...
transphenoidal resection of the tumor. The surgical aspect did not differ from that found in a pituitary adenoma, both in consistency, color, as well as in bleeding. The cavity was filled with autologous fat.

Postoperative period: The patient presented no major complications, except for diabetes insipidus detected during the early postoperative period which was completely controlled. Residual tumor was identified, but no tumor recurrence was observed after a follow-up period of 24 months.

Pathological anatomy: Microscopy showed a predominantly fusocellular tumor consisting of pleomorphic cells with a fascicular growth pattern. The cells were characterized by abundant eosinophilic cytoplasm, vesiculous nuclei with mild atipia and clearly visible nucleoli. Mitotic figures were occasionally observed [Figure 2].

The material was analyzed by immunohistochemical study. Antibodies against several antigens were tested and are shown in Table 2. Most tumor cells were reactive to the cell proliferation antigen Ki-67 and to protein S-100. There was no reaction to the other antigens tested, including glial fibrillary acidic protein (GFAP). According to the criteria proposed by Brat et al.,[4] the diagnosis of pituicytoma was thus confirmed.

DISCUSSION

The neurohypophysis comprises the posterior portion of the pituitary, infundibulum, and tuber cinerium.[7,17] The cellular elements that form the posterior part of the pituitary include microglia, pituicytes, and terminal axons of secretory neurons of the hypothalamus. Pituicytes are modified microglial cells that occupy perivascular areas of the neurohypophysis and participate in the regulation of the release of hypothalamic hormones. The cells are spindle shaped and normally react to GFAP.[16] Five types of pituicytes have been described based on the histopathological criteria of Takei et al.,[7,10,18] (1) major pituicytes, the most common type characterized by an oval or irregular nucleus, distinct nucleoli, and cytoplasm containing various organelles; (2) dark pituicytes which present the same structure as major pituicytes, but have an electron-dense cytoplasm; (3) oncocytic pituicytes characterized by a large number of mitochondria; (4) ependymal pituicytes which are rudimentary ependymal cells; and (5) granular pituicytes which contain numerous electron-dense granules and give origin to granular cell tumors or choristomas.

The most common pituitary tumors are adenomas originating from the adenohypophysis. Although rare, posterior pituitary tumors include hamartomas, craniopharyngiomas, germinomas, granular cell tumors, meningiomas, pituicytomas, and pilocytic astrocytomas.[2,7]

Few cases of primary tumors of the neurohypophysis have been described, a fact impairing the classification of these tumors. So far, 30 cases of pituicytomas have been published [Table 1].[1,3,5,9,11,12,14,16,19-22] In 2000, Brat et al.,[2] described nine cases of pituicytomas and provided a more precise and detailed characterization of the clinical and pathological findings related to this tumor. In addition, the authors proposed a clearer and more objective definition of the tumor.[6,8]

Table 2: Antibodies tested in immunohistochemical study

| Antibodies                               | Clone    | Results |
|------------------------------------------|----------|---------|
| Proliferation antigen Ki-67              | MIB-1    | Positive|
| Epithelial membrane antigen — EMA        | E29      | Negative|
| Cytokeratins of 40, 48, 50, and 50.6 kDa | AE1/AE3  | Negative|
| Protein S-100                            | Policlonal| Positive|
| Glial fibrillary acidic protein — GFAP   | Policlonal| Negative|
| Synaptophysin                            | Sy38 AFP | Negative|
| CD68 – lysosomal protein                 | KP1      | Negative|
| Nerve growth factor receptor             | NGFR5    | Negative|
| Melanoma                                 | PNL2     | Negative|
| Melanoma-associated gp100 antigen        | HMB-45   | Negative|
| Melanoma antigen recognized by T cells – | A103     | Negative|
| Melan A/MART-1                           |          |         |

Figure 2: (A) Microscopic view showing a fusocellular tumor consisting of pleomorphic cells with a fascicular growth pattern. (B) Cells with eosinophilic cytoplasm and clearly visible nucleoli. (C) Immunohistochemical reaction with S-100 protein.
Pituicytomas are rare, noninfiltrative tumors of glial origin, which arise in the neurohypophysis that comprises the posterior region of the pituitary and the pituitary stalk.[2] In the past, the term pituicytoma frequently included granular cell tumors or choristomas and pilocytic astrocytomas.[6] Today, this term is reserved for low-grade gliomas classified as grade I by the World Health Organization and differing from astrocytomas.[11] Thus, as in the present case, pituicytomas are solid well-defined tumors, which appear isointense on T1-weighted MRI and are characterized by marked vascular proliferation and intense contrast enhancement. These findings are nonspecific and confirmation by anatomopathological analysis is necessary.[6,11]

Histologically, pituicytomas are characterized by spindle-shaped cells arranged in interlacing fascicles. Nuclei is oval to elongated with a mild irregularity. The citoplasm is eosinophilic and homogeneous which presents little or no granulation or vacuolization and necrosis is absent.[5] Pituicytomas are histologically classified as low-grade gliomas which present little nuclear atypia and rare mitotic activity. Most tumor cells are reactive to the cell proliferation antigen Ki-67 and to protein S-100. The cells normally react to GFAP and present little or no cytoplasmic reactivity to epithelial membrane antigen (EMA). In the series described by Brat et al.,[2] one of the nine cases showed no reactivity to GFAP and wide variation in the intensity of the reaction was observed in the remaining eight positive cases. In the present patient, the tumor consisted of spindle-shaped cells with a fascicular organization and abundant granular cytoplasm and showed intense positivity to the Ki-67 and protein S-100 antigens, findings compatible with pituicytoma, and no reactivity to EMA or GFAP.

The most common clinical presentation was headache and bitemporal hemianopia due to compression of the optic chiasm.[11] Other signs frequently observed are panhypopituitarism and mild hyperprolactinemia.[2] The patient may even be completely asymptomatic, and the tumor is detected incidentally. The present patient had mild hyperprolactinemia, with the other pituitary hormones being normal, associated with persistent headache and bitemporal hemianopia. These findings, together with the imaging detection of a solid and well-delimited mass in the sellar region, may lead to the clinical diagnosis of pituitary adenoma. The indicated treatment is surgery which can be performed by the transsphenoidal approach. The present patient underwent transsphenoidal resection of the tumor. Tumor consistency and increased bleeding observed during surgery are essential data that help the pathologist with the diagnosis of the tumor. Some authors have reported a greater bleeding tendency of these tumors during surgery.[7] In the present case, no perioperative alterations that would distinguish the tumor from a macroadenoma were observed and the diagnosis was exclusively made by histological analysis.

No tumor recurrence was observed in the related case after a follow-up period of 24 months. Although few studies are available in the literature, data suggest a high rate of tumor recurrence after partial resection and a good prognosis, with little or no recurrence, after total resection.[4]

**CONCLUSION**

Pituicytomas are rare tumors of the neurohypophysis derived from pituicytes. Their clinical presentation resembles that of non-functional pituitary adenomas, but these two types of tumors are histologically well distinct. Surgery is the indicated treatment with a good prognosis if the tumor is completely resected.

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