Pituitary Carcinoma with Cerebral Spinal Fluid Dissemination Presenting with Spinal Cord Compression


carcinoma de hipófise com disseminação liquórica apresentando-se com compressão medular

João Pedro Einsfeld Britz
Guilherme Reghelin Goulart
Ericson Sfreddo
André Martins de Lima Cecchini
Felipe Martins de Lima Cecchini
Tobias Ludwig do Nascimento
Gerson Evandro Perondi

ABSTRACT
Pituitary carcinoma is a rare tumor of the anterior pituitary, and by definition occurs in the presence of distant metastasis. Treatment involves surgical resection, radiotherapy and adjuvant chemotherapy. The authors report the case of a patient with pituitary carcinoma that evolved with spinal cord compression.

Keywords: Pituitary neoplasms; Pituitary tumors; Pituitary adenoma; Spinal cord compression

RESUMO
O carcinoma de hipófise é um tumor raro da hipófise anterior e, por definição, ocorre na presença de metástase à distância. O tratamento envolve a ressecção cirúrgica, radioterapia e quimioterapia adjuvante. Os autores relatam o caso de um paciente com carcinoma de hipófise que evoluiu com compressão medular.

Palavras-chave: Neoplasias de hipófise; Tumores de hipófise; Adenoma hipofisário; Compressão da medula espinhal
INTRODUCTION

Despite the relatively high prevalence of pituitary tumors, only a minority evolve to carcinoma. Pituitary carcinomas originate from the anterior pituitary, and by definition occur in the presence of metastases, which may be systemic or in the central nervous system. The vast majority are hormone producers, especially ACTH, prolactin and GH\(^1\). The authors report in the present work the case of a patient with pituitary carcinoma that evolved with spinal cord compression after leptomeningeal dissemination of the disease.

CASE PRESENTATION

A 54-year-old female patient, with no known previous pathologies, presented progressive visual deterioration and ptosis of the right eye with 4 months of evolution, which subsequently evolved with low bilateral visual acuity. She investigated the condition with magnetic resonance imaging (MRI), which showed an expansive lesion with an intra and suprasellar component, suggestive of a pituitary macroadenoma (Figure 1). She underwent surgery for resection via the transphenoidal route, with partial removal of the tumor, which was complemented with a transcranial route during the same hospital stay. She presented partial improvement of vision after the procedure.

The anatomopathological and immunohistochemical results showed pituitary carcinoma without hormone suppression as the final diagnosis, and the patient was then referred for complementary treatment with radiotherapy, since it was not possible to perform a complete resection of the tumor due to invasion of parasellar structures. Systemic screening for metastasis at the time was negative.

Outpatient follow-up with the neurosurgical team continued for another 2 years, with sella MRI demonstrating an excellent response to radiotherapy and stability of the condition during the follow-up period (Figure 2).

In the third year of follow-up, the patient was hospitalized again due to subacute paraparesis and urinary retention. On admission, she already had paraplegia with a week of evolution. Spine MRI identified two intradural and extramedullary expansive lesions at the level of T10-T11 and L2-L3, causing medullary and radicular compression (Figure 3). New systemic screening with computed tomography (CT) scans of the chest and abdomen was negative for other metastatic lesions.

Given the time course of the neurological symptoms and the lack of new benefits from surgical therapy, the patient was referred to the oncology team, where she underwent a new radiotherapy cycle, supplemented with temozolamide.

Figure 1. Expansive lesion with solid aspect and necrotic component, intra and suprasellar, which extends to the middle fossa and involves the cavernous segments of the internal carotid arteries and presents homogeneous enhancement after the use of gadolinium.
DISCUSSION

Pituitary tumors are neoplasms that originate from the adenohypophysis, represented mostly by adenomas. Despite having the potential to invade bone, meningeal and neural structures adjacent to the sella, it is considered a benign disease.

Unlike pituitary adenomas, pituitary carcinoma is defined as a malignant neoplasm of the adenohypophysis, being mandatory...
for the presence of systemic dissemination of the disease or dissemination in the central nervous system for its characterization. It is a rare disease, representing 0.5% of pituitary tumors and 0.2% of sellar tumors, with its peak prevalence in the fourth and fifth decade of life, being an even rarer disease in the pediatric population.\(^2,3\)

In addition to the presence of metastases, other criteria are necessary to characterize a pituitary carcinoma, such as histology and markers compatible with pituitary tumors and the absence of another primary tumor, since other systemic neoplasms can affect the sellar area.\(^4\) Such criteria are necessary since it is impossible to distinguish adenoma from carcinoma based only on histological criteria, since hypercellularity, nuclear pleomorphism and necrosis may also be present in ACTH and GH producing adenomas.

Interestingly, systemic metastases are more common than dissemination in the central nervous system, with the liver, bones and lungs being the most common metastatic sites, and they occur due to hematogenous and lymphatic dissemination of the tumor.\(^5\) In the central nervous system, dissemination occurs via CSF, and affects mainly the brain, cerebellum and region of the cerebellopontine angle. In the case presented above, the patient had only intraspinal metastases, with no evidence of systemic disease or disease elsewhere in the nervous system.

In four cases of pituitary carcinoma described by Alshaikh et al.,\(^2\) three of them presented dissemination to the spine, but with no description of intraspinal involvement.\(^2\) In the case described by Guzel et al.,\(^3\) the patient had intramedullary lesions, in addition to concomitant intracranial involvement.\(^3\) Despite the possibility of CSF dissemination in this type of tumor, neuraxial examination is not routinely performed, being guided by the clinical symptoms of the patient.\(^6\) Intraspinal metastatic spread of pituitary carcinoma represents an exceptionally rare disease, with few cases reported so far, and there is usually a long interval between the diagnosis of the primary tumor and the clinical presentation of the intraspinal metastasis.\(^7,8\)

Treatment of pituitary carcinoma involves surgical resection, radiotherapy and adjuvant chemotherapy. Radiotherapy has been shown to be effective in the local control of the disease, without showing any benefit in terms of increased survival, and is also used in the management of bone and visceral metastases for temporary control and pain management.\(^4,6\)

Several different chemotherapy schemes have been used as adjuvant therapy, with good response, however with short and temporary duration. Based on currently available data from ACTH and prolactin-secreting carcinomas, temozolamide has been shown to be an effective therapy.\(^6,9,10\)

Despite advances in treatment, it remains an extremely aggressive disease, with average survival of 12 months after diagnosis. The obstacles to the advancement of studies on the subject are due to the rarity of the disease, making it difficult to carry out large-scale studies.\(^6\)

**CONCLUSION**

Pituitary carcinoma is a rare and aggressive neoplasm. To be considered a pituitary carcinoma, it is mandatory that there is dissemination of the disease, either systemic or in the central nervous system. The treatment for this disease is multidisciplinary and involves surgical resection of the tumor, complemented with radiotherapy and adjuvant chemotherapy. Currently, it is difficult to distinguish which pituitary neoplasms can evolve into carcinoma, when based only on their histological characteristics.

**REFERENCES**


CORRESPONDING AUTHOR

João Pedro Einsfeld Britz, MD
Conceição Hospitalar Group
Cristo Redentor Hospital
Department of Neurosurgery
Porto Alegre, Rio Grande do Sul, Brazil
E-mail: joaopedrobritz@hotmail.com

Funding: nothing to disclose
Conflicts of interest: nothing to disclose
Institution: Cristo Redentor Hospital.