

A rare case of an aggressive macroprolactinoma invading the sphenoid bone, clivus and cavernous sinuses posing the differential diagnosis with chordoma

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Abstract

Aggressive macroprolactinomas represent 0.4 to 4% of pituitary adenomas, they are more frequent in men, the positive diagnosis is easy, the evaluation of aggressiveness is essentially based on MRI and The treatment with dopaminergic agonist in first intention is now well established and should not be deferred apart from neurological emergency situations.

Keywords: Aggressive Macroprolactinomas; Chordoma; Hypothalamic-Pituitary MRI; Knosp's Classification; Complex Management

1. Introduction

Prolactinomas are the most frequently encountered type of pituitary adenoma. It is often voluminous and invasive in men. Their diagnosis and management are often straightforward. However, some cases can be very challenging to treat [1]. We report the case of an aggressive macroprolactinoma invading the sphenoid bone, clivus and cavernous sinuses and which made the differential diagnosis with chordoma

2. Clinical case

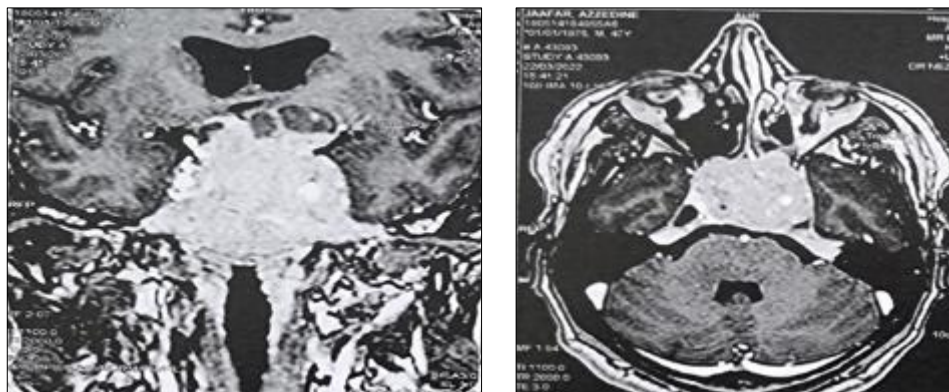


Figure 1 Hypothalamic-pituitary MRI

The patient is 47 years old, **On medical history:** diabetic for 3 months on metformin 500 mg/d Who presented with progressive headaches that were resistant to the usual analgesics and associated with a decrease in visual acuity, for which he consulted an ophthalmologist and then the neurosurgery department, with the following explorations:

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Hypothalamo-hypophyseal MRI: sellar and suprasellar lesional process measuring 40*55*52 mm with cystic and hemorrhagic zones invading the sphenoidal bone and the clivus, the cavernous sinus and the supra sellarcistern with vascular envelopment in favor of a chordoma or an invasive macroadenoma. The visual field: right eye: temporal hemianopia; left eye: profound lowering of retinal sensitivity in the entire visual field. The pre-op hypophysogram had objectified :TSH =1,10 mui /L. T4L=10.72 pmol /l; T3L=3.30 pmol/l; FSH=1.1ui/l; LH=1 ui/l; testosterone=untreated; 8 o'clock cortisol=17.8 ug/dl; prolactin>100 ng/ml. The patient was put on levothyrox 25 ug/d then operated by transphenoidal way after coverage of gesture by hydrocortisone hemiscucinate with at the anapath :morphological and immunohistochemical aspect of a pituitary adenoma expressing anti prolactin CA, proliferation index KI67% at 3%. Hypophysigram on day 3 post-op: prolactin: 4700 ng/ml; 8 o'clock cortisol: 8.44 ug/dl; T4: 10.2 pmol/l; T3L: 2.6 pmol/l. Then the patient was put on dostinex 0,5 mg : 4 cp/week with optimization of levothyrox at 50 ug/d

3. Discussion

Macroprolactinomas, especially in men, can sometimes present a very aggressive clinical course, as evidenced by progressive growth, bone invasion in the sphenoid sinus, cavernous sinus, suprasellar region or nasopharynx. Some may even evolve into pituitary carcinoma with craniospinal or systemic metastases [2]. The assessment of aggressiveness is mainly based on MRI, which remains the best technique to assess the extent of the tumor, CT scan provides better information on the interruption of the sellar floor and intratumoral calcification. Radiologically, the aggressiveness of these tumors is probably best reflected by invasion of the cavernous sinuses. [2]. Aggressiveness is defined as a radiologically invasive tumor with an unusually rapid tumor growth rate, or clinically relevant tumor growth despite optimal standard treatments (surgery, radiation therapy, and conventional medical treatments). There is also wide variation in the literature regarding the definition of invasiveness, based on imaging features, histologic evidence of sphenoid sinus mucosal invasion, and/or intraoperative findings [3]. In addition, the infiltrated anatomical structure is important because invasion of the clivus or sphenoid bone, unlike invasion of the cavernous sinus, is a more indicative feature of aggressive behavior [4]. In the Hardy classification, for example, only grade III (focal bone erosion) and grade IV (extensive bone erosion including the skull base) tumors are considered invasive (Figure 1) [5]. Furthermore, with regard to parasellar invasion of the cavernous sinuses according to Knosp's classification, only grade III and IV adenomas are considered truly invasive [6]. (Figure 2)

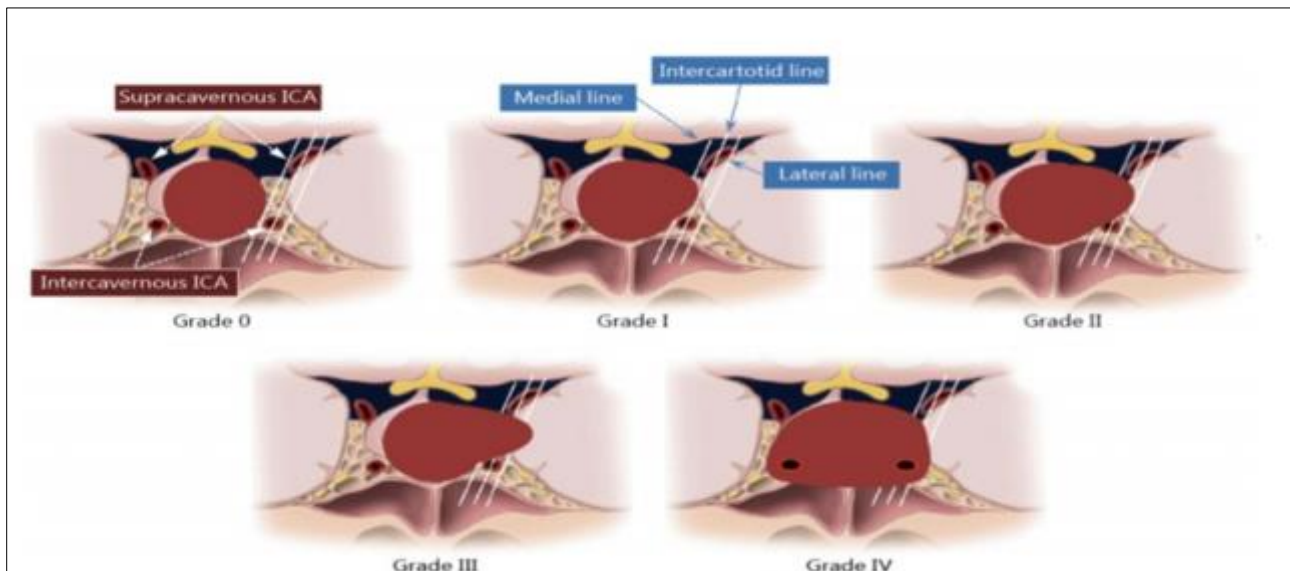


Figure 2 Hardy's Classification of pituitary tumours ; Upper part shows the classification of sphenoid bone invasion (grade 0: intact with normal contour; grade I: intact with bulging floor; grade II: intact, enlarged fossa; grade III: localized sellar destruction; grade IV: diffused destruction). Only Grade III and IV tumors are considered invasive. Lower part depicts a classification of suprasellar extension of an adenoma which may be symmetrical or asymmetrical (Grade A: suprasellar cistern only; grade B: recess of the third ventricle; grade C: whole anterior third ventricle; grade D: intracranial extradural; grade E: extracranial extradural [cavernous sinus])

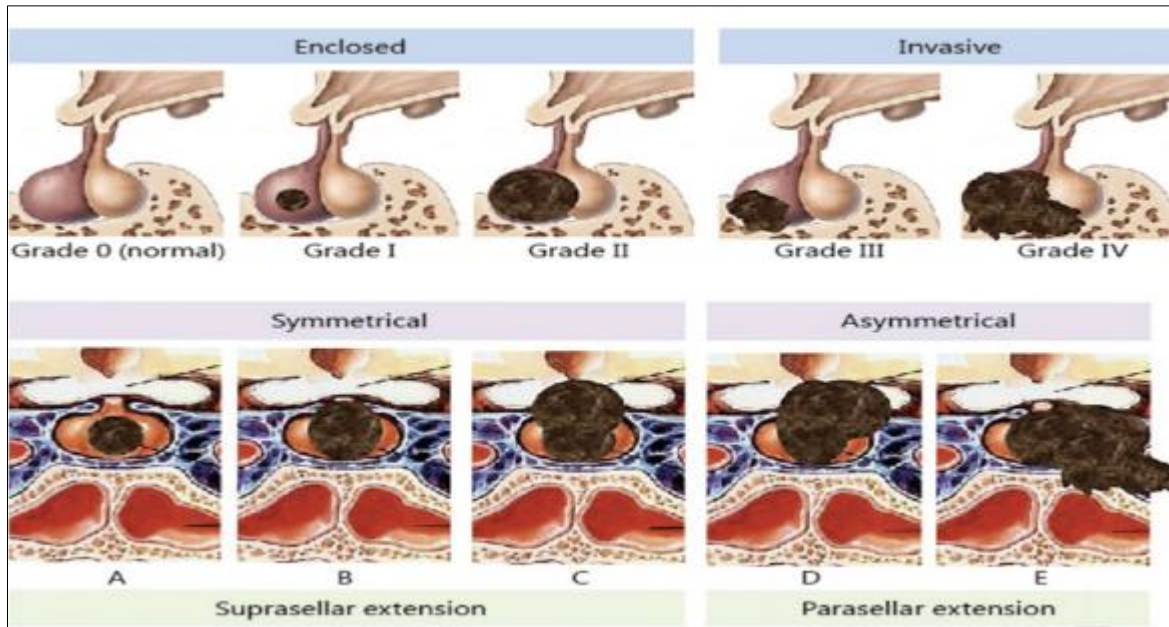


Figure 3 Knosp's classification of cavernous sinus invasion: Grade 0 - The adenoma does not pass the tangent of medial aspects of internal carotid artery (ICA). Grade I - The medial tangent is passed, but the extension does not go beyond the intercarotid line, which is the line drawn between the cross-sectional centers of the intraand supracavernous ICA. Grade II is characterized by the tumorextending beyond the intercarotid line, but not beyond or tangent to the lateral aspects of the intra-and supracavernous ICA. Grade III is characterized by the tumor extending laterally to the lateral tangent of the intra-and supracavernous ICA. Grade IV is characterized by total encasement of theintra-cavernous carotid artery (Based on [6])

Management should be multidisciplinary as most prolactinomas are benign and respond to medical treatment. However, in the case of aggressive prolactinomas, due to the risk of recurrence and local invasion of these tumors, several approaches such as medical treatment, surgery, radiotherapy and chemotherapeutic agents will be necessary. [7]. Temozolomide is an oral antineoplastic alkylating agent initially used for glioblastoma multiforme. Not long ago, it was shown to be effective in the treatment of refractory aggressive prolactinomas. It is an oral drug, relatively well tolerated, and has shown promising results [8].

4. Conclusion

Aggressive pituitary adenomas are of complex management with a very limited therapeutic arsenal at the present time.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

No conflict of interest.

Statement of ethical approval

The present research work does not contain any studies performed on animals/humans subjects by any of the authors.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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