Pituicytoma

In 2017, changes in the classification of non-neuroendocrine tumors are also proposed, in particular those tumors arising in the posterior pituitary including pituicytoma ¹.

Pituicytoma is a rare glial sellar/suprasellar neoplasm arising in the neurohypophysis with a possible origin from the folliculostellate cells of the adenohypophysis which are non-endocrine spindled cells expressing S-100 and Bcl-2 ² ³.

Pituicytoma is considered to be a distinct Grade I neoplasm ⁴.

Although usually intra-sellar, pituicytomomas can have suprasellar extension; however, purely suprasellar examples although rare have been reported ⁵.

Epidemiology

PTs had a higher prevalence in the fifth and sixth decades of life, with a slight male predominance. ⁶.

Clinical features

The presenting symptoms are due to the mass effect of the tumor and include visual disturbances caused by direct compression on the optic chiasm, headaches, endocrinological symptoms and rarely diabetes insipidus ⁷.

Diagnosis

Radiologically, PTs were found anywhere along the hypothalamic-pituitary axis mimicking other, more frequent tumors growing in this anatomical region ⁸.

The MRI features are non-specific with most case reports showing a solid, homogenous mass, iso-intense on T1-weighted images and hyper-intense on T2-weighted images with homogenous contrast enhancement ⁹.

Amongst the various sellar tumors, pituicytoma and spindle cell oncocytoma (SCO) have considerable overlap in histological, Immunohistochemical (IHC) profile and can have extensive intraoperative bleeding making complete excision difficult with increased chances of recurrence. It is important to differentiate pituicytoma from SCO since the former is associated with a slightly better prognosis with recurrence being uncommon after complete surgical excision. Till 2013, out of 29 cases of pituicytoma with a detailed follow-up, recurrence was seen in six cases, all of which were found to have an incomplete resection during the first surgery ¹⁰.

SCO on the other hand have a tendency to recur even after complete excision. Hence, it is advocated to combine surgery with adjuvant radiotherapy in all cases of SCO to reduce the chances of recurrence. EMA is strongly positive in SCO, thus it can help to differentiate pituicytoma from SCO ¹¹.

Subtypes

TTF-1 Expressing Sellar Neoplasm with Ependymal Rosettes and Oncocytic Change: Mixed Ependymal and Oncocytic Variant ¹².
Treatmen

see Pituicytoma treatment.

Review

Less than 50 cases have been reported in the world literature till 2013.  

Salge-Arrieta et al., from the Ramón y Cajal University Hospital Madrid, Spain, published a retrospective review of case reports published in the scientific literature to 2018, including a new illustrative example treated.

116 cases were collected. PTs had a higher prevalence in the fifth and sixth decades of life, with a slight male predominance. Main symptoms, which tended to be progressive, included visual field defects and pituitary-hypothalamic dysfunction. Radiologically, PTs were found anywhere along the hypothalamic-pituitary axis mimicking other, more frequent tumors growing in this anatomical region. Surgical treatment included both transcranial or transsphenoidal approaches, and resulted in gross total resection and morbidity rates of 46.8 and 59%, respectively; the latter essentially consisted in anterior and posterior pituitary dysfunction, with limited impact on daily quality of life.

Due to both low frequency and the absence of pathognomonic clinical and/or radiological features, formulating a suspicion diagnosis of PT represents a considerable challenge even for experienced professionals. The indication for treatment should be made on an individual basis, but it is inescapable in the presence of a visual field defect. The surgical approach has to be tailored according to the topography of the tumor and preoperative symptoms; the greatest challenges in accomplishing a gross total removal are represented by the degree of adherence and vascularization of the PT.

Case series

Lefevre et al., from the Groupe Hospitalier Pitié-Salpêtrière, Paris, France published a retrospective multicenter study, reporting the clinical manifestations, radiological characteristics, histopathological features, treatment strategies and long-term outcomes of patients who have been treated for a Pituicytoma at various institutions in Paris, France over the past 10 years. In addition, they compared the results to the world literature in order to identify similarities concerning the radiographic diagnosis and the treatment strategies of these tumors.

Eight patients were operated on in four different hospitals. Misdiagnosis was constant before surgery, pituitary adenoma or craniopharyngioma being suspected. During surgery (transsphenoidal approach: six cases, transcranial approach: two cases) unusual tumors were noted, with important bleeding in most cases. Complete resection could be obtained in five patients. Pathological diagnosis was confirmed in all cases. During the follow up two recurrences occurred. One was subsequently treated with radiotherapy, the other underwent a second surgery.

Recent updates concerning the histological diagnosis of pituicytomas should be generalized to our practice in order to provide a better understanding of this rare pathology and its natural course.

References

Lopes MBS. The 2017 World Health Organization classification of tumors of the pituitary gland: a


