Pituicytoma

Pituicytoma is a rare glial neoplasm from pituicytes of the neurohypophysis or infundibulum. It occurs in the sella and suprasellar area, and it is extremely uncommon to observe intraventricular pituicytoma without affecting the infundibulum or infundibular recess.

Posterior lobe tumors, the family of pituicyte tumors, include the traditional pituicytoma, the oncocytic form (spindle cell oncocytoma), the granular cell form (granular cell tumor), and the ependymal type (sellar ependymoma). Although these historical terms are entrenched in the literature, they are nonspecific and confusing, such that oncocytic pituicytoma, granular cell pituicytoma, and ependymal pituicytoma are now proposed as more accurate [1)]

In 2017, changes in the classification of non-neuroendocrine tumors are also proposed, in particular those tumors arising in the posterior pituitary including pituicytoma [2)].

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 [3) 4)].

Pituicytoma is considered to be a distinct Grade I neoplasm [5)].

Although usually intra-sellar, pituicytomas can have suprasellar extension; however, purely suprasellar examples although rare have been reported [6)].

Epidemiology

PTs had a higher prevalence in the fifth and sixth decades of life, with a slight male predominance. [7)]. [8)]

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 [9)].

Diagnosis

Pituicytoma Diagnosis

Subtypes

TTF-1 Expressing Sellar Neoplasm with Ependymal Rosettes and Oncocytic Change: Mixed Ependymal and Oncocytic Variant [10].

Differential Diagnosis

When small, and clearly localized to the infundibulum the main differential includes:

granular cell tumors of the pituitary region

craniopharyngioma

When larger, then it is difficult to anticipate the diagnosis with other diagnoses being far more common, including:

pituitary macroadenoma

meningioma

pituitary metastasis

pituitary infiltration

lymphocytic hypophysitis

neurosarcoidosis

optic pathway glioma

Absent Rosenthal fibers and eosinophilic granular bodies, usually help to distinguish between pituicytomas and pilocytic astrocytomas [11].

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 [12].

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 [13].

Treatment

see Pituicytoma treatment.

Review

Less than 50 cases have been reported in the world literature till 2013 [14)].

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 [15)].

Case series

Pituicytoma Case Series.

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