Neurosurgical forum
Letters to the editor

Craniopharyngioma Classification

TO THE EDITOR: We read with great interest the article by Kassam et al. (Kassam AB, Gardner PA, Snyderman CH, et al: Expanded endonasal approach, a fully endoscopic transnasal approach for the resection of midline suprasellar craniopharyngiomas: a new classification based on the infundibulum. *J Neurosurg 108:*715–728, April, 2008). The authors showed the unparalleled possibilities provided by the expanded transsphenoidal approach in combination with endoscope-assisted technology in achieving total and safe resection of craniopharyngiomas that have a suprasellar and/or intraventricular location. The expanded endonasal approach (EEA) variants described by this group of experts in endoscope-assisted microneurosurgical techniques may introduce a deep change in the old paradigm that considered midline lesions above the diaphragma sellae to be unapproachable via a transsphenoidal route. We are impressed with the technical possibilities of the excision of retroinfundibular craniopharyngiomas as shown in their article, and we congratulate the authors on the innovative and elegant procedures presented, such as the transposition of the pituitary gland, which opens a free corridor to lesions with a suprasellar retrostalk position.

In their paper Kassam and colleagues introduced a new topographical classification of craniopharyngiomas that is based on the relationships of the tumor to the infundibulum and pituitary stalk, as observed in the surgical field by endoscopic viewing. This scheme was helpful in selecting the type of EEA required for the most adequate exposition and safest excision of a lesion. We think this classification highlights important concepts regarding the true anatomical relationships of a tumor to vital neurovascular structures situated at the basal brain surface, especially the undersurface of the optic chiasm and third ventricle floor, which are usually hidden from direct view when using a transcranial approach. Nonetheless, their new system shares with previous schemes the drawback of being based on the particular viewpoint provided by a specific surgical approach, and hence it focuses on a limited description of the anatomical relationships that should be considered preoperatively when deciding on the best approach to such a complex lesion as the craniopharyngioma. The lack of a unified topographical classification scheme capable of describing in every case the accurate surgical relationships between a tumor and the type of distortion it causes to surrounding neurovascular structures is probably one of the reasons the craniopharyngioma is, in the words of Harvey Cushing, the "most baffling problem which confronts the neurosurgeon." Nevertheless, the surgical outcome in a patient harboring a craniopharyngioma is known to be basically influenced by 2 factors: the degree of excision achieved and the damage caused to the hypothalamus during dissection and removal of the tumor. The hypothalamic nuclei are embedded within the third ventricle walls, in close relation to the infundibulum and tuber cinereum, the components of the third ventricle floor that become distorted with the expansion of a craniopharyngioma. The type of craniopharyngioma-induced deformation to the third ventricle boundaries—for example, progressive invagination caused by tumoral compression, invasion of the third ventricle by a mass breaking through the floor, or even the replacement of these structures by a lesion growing within the hypothalamus itself—will dictate the final position and functional state of the hypothalamus. Consequently, we think that the usefulness of a topographical classification of craniopharyngiomas in terms of both planning the surgical approach and predicting the potential risks of injury associated with the lesion excision should be based on an accurate description of the tumor relationships to the third ventricle and the diencephalic structures bordering it.

In the early 1960s French authors initiated theoretical discussions about the different possible relationships between craniopharyngiomas occupying the third ventricle and the distorted ventricle boundaries.[8] Given the chance of approaching an intraventricular lesion via a transcortical-transventricular approach, the position and functional state of the third ventricle walls, containing the hypothalamic nuclei, became of paramount importance. The absence of reliable neuroradiographic methods of diagnosis at that time, together with the usual fatal outcome following attempted excision of a tumor via a frontal transventricular approach, led Pertuiset and colleagues[8] to introduce the topographical concept of a pseudointraventricular location for craniopharyngiomas. This topography would be assigned to any tumor seeming to have a third ventricle location but in fact originating from a suprasellar extraaxial position, which would cause invagination and upward stretching of the third ventricle floor during lesion growth. The upward position of the intact third ventricle floor as a thinned membrane capping the tumor would suppose a high risk of hypothalamic injury in the event that a transventricular approach was attempted through these structures to the lesion. Nonetheless, Van Den Bergh and Brucher[15] favored the transventricular approach as a safe route for the removal of intraventricular craniopharyngiomas, even in cases of pseudointraventricular lesions capped by remnants of the third ventricle floor, because they observed that those remnants become atrophic and nonfunctional due to prolonged tumoral compression. They noted instead that the viable hypothalamic tissue remained in a lower position, attached to the basal tumoral pole, which was the area most vulnerable to surgical manipulation.

The article by Kassam et al. includes pre- and postoperative MR images as well as beautiful intraoperative endoscopic photographs of a solid craniopharyngioma occupying the third ventricle; the basal portion of the lesion expands the third ventricle floor and is classified as a Type III retroinfundibular craniopharyngioma (Figs. 14–16 in their article). After tumor excision, the widely opened third ventricle floor, showing a residual circumferential rim of subependymal bleeding along the area of tumor attachment to the third ventricle walls, could be photographed through an endoscope. Similar intraoperative photographs of intraventricular craniopharyngiomas have been taken from a

basal endoscopic view by Laufer et al.[5] and de Divitiis et al.[2] and also showed the basal opening of the third ventricle floor and subependymal blood marks at the inner inferior edges of the third ventricle walls, which were related to the tumor attachments to the hypothalamic area. All of these images beautifully illustrate the tumor and third ventricle relationships of the not-strictly-intraventricular type of craniopharyngioma (Fig. 1A and B), as described in our classification scheme based on an extensive review of well-described cases of craniopharyngiomas with an intraventricular location.[7] The main characteristic of this type of lesion is its primary development neither in the third ventricle nor along the suprasellar course of the pituitary stalk, but instead within the thin neural layer of the infundibulum and/or tuber cinereum, the main constituents of the third ventricle floor. Following the embryonic theory for the origin of craniopharyngiomas, this infundibulotuberal variant develops from epithelial remnants of the craniopharyngeal duct that are included within the neural tissue when the pars tuberalis comes into contact with the diencephalic infundibular evagination, before the pia mater covers the basal brain surface.[1] With its progressive growth, the lesion will cause expansion of the floor, breaking through the ependymal layer and occupying the third ventricle and thus simulating a true, or strictly, intraventricular lesion (Fig. 1C–F). Preoperative MR imaging identification of the remnants of the third ventricle floor cannot be made for this topography; however, once a tumor is totally excised, postoperative MR images usually show the anatomical defect at the third ventricle floor.

The identification of a craniopharyngioma occupying a tuberal position and with a cystic extension into the third ventricle was originally made by Norman M. Dott[3] in 1938. In his masterpiece, "Surgical aspects of the hypothalamus," Dott illustrated the detailed relationships of this topographical variant of craniopharyngioma and pointed out the tight adhesions of the inferior tumor capsule to the hypothalamic walls. He claimed the need for a staged combined transventricular and pterional basal approach for these lesions and successfully removed some hypothalamus-centered lesions several decades before the application of similar approaches via microsurgical techniques.[16] Note, however, that the term "tuberal" was not used as a formal category in the topographical classification of craniopharyngiomas until 2 decades later when another british author, Douglas W. C. Northfield,[6] observed a relocation of the third ventricle floor by a tumor mass in 20 of 49 surgically treated cases, with postoperative necropsy confirmation of such a location in many of these cases. Similar findings were observed in 23 of 40 cases surgically treated by William Sweet,[13] one of the neurosurgeons who pioneered the use of the translamina terminalis approach to remove intraventricular craniopharyngiomas. Sweet also histologically analyzed the tight adherences between the basal tumor surface and the third ventricle walls, providing evidence of a nonfunctional gliotic cleavage plane that separated the tumor capsule from the hypothalamic nuclei. He argued that safe tumor dissection from the hypothalamus was possible through this gliotic tissue. A modern topographical classification system of craniopharyngiomas based on tumoral relationships as displayed on preoperative MR imaging studies was applied by Raybaud et al.[9] in 1991, who considered the infundibulotuberal topography as the most frequently observed type in patients younger than 18 years of age (10 of 23 cases).

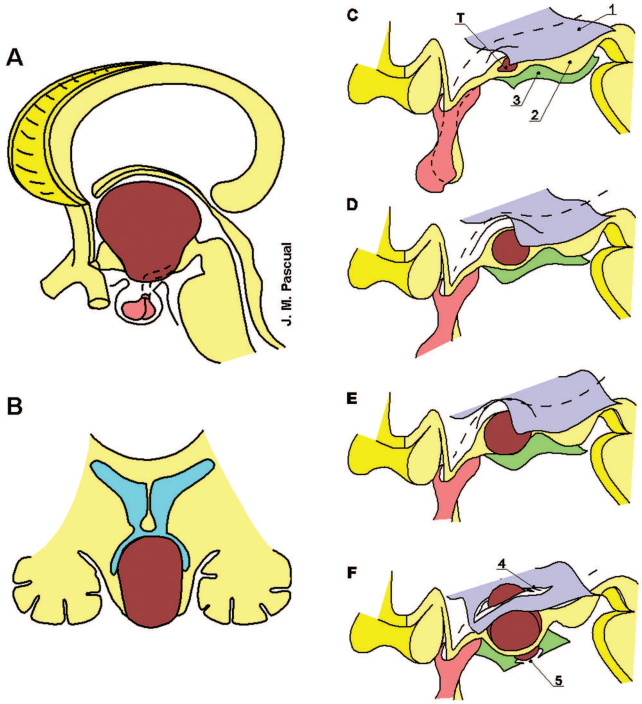

FIG. 1. Schematics illustrating the topographical relationships of the not-strictly-intraventricular type of craniopharyngioma, which corresponds to Type III retroinfundibular in the new classification scheme by Kassam et al. Sagittal (A) and coronal (B) views showing occupation and expansion of the third ventricle floor as caused by the basal part of the lesion. Origin of the lesion from epithelial remnants of the craniopharyngeal duct (C) and progressive phases of its growth at the third ventricle floor (D–F), following the embryonic theory by Ciric and Cozzens. T = tumor; 1 = ependymal layer; 2 = nervous tissue layer; 3 = pia mater; 4 = upper tumoral pole breaking through the ependymal layer; 5 = lower tumoral pole breaking through the pia mater.

In 1985 Juraj Steno[11] challenged the classic topographical concept of craniopharyngiomas—that is, these lesions predominantly occupy an extracerebral suprasellar position—by showing from among the autopsy specimens of 30 nonsurgically treated patients 14 examples of extraventricular lesions, whose equator showed a tight circumferential attachment to the third ventricle floor, and 8 cases of purely intraventricular tumors. These striking figures were confirmed in a recent MR imaging investigation by the same author in a surgical series of craniopharyngiomas, with the extraventricular type occurring in 25 of 44 supradiaphragmatic lesions in children and adults.[12] Once these tumors had been removed, a wide defect in the third ventricle floor could be observed in the surgical field and on postoperative MR images. The MR imaging study of craniopharyngiomas by Eldevik et al.[4] in 1996 showed the occupation of the third ventricle in 10 of 13 children and in 7 of 14 adults. Strictly or mostly intraventricular tumors have also been found in > 50% of the cases in a large series studied by Shi et al.[10] and in a series of children evaluated by Tomita and Bowman.[14] In all of these cases the tumor

capsule adhered to the inferior third ventricle walls (hypothalamus).

Kassam et al. have demonstrated that with the EEA, debulking and sharp dissection of a craniopharyngioma expanding at the infundibulotuberal area can be safely achieved by avoiding blind pulls of the tumor capsule attached to the hypothalamus. In this respect, we share the opinion of Shi et al.[10] that the prevention of surgical damage to the tiny perforating vessels that supply the hypothalamic nuclei is one of the fundamental factors related to a good long-term outcome following excision of a craniopharyngioma. In a comparative review of the surgical results achieved with the excision of intraventricular craniopharyngiomas through different approaches, we found that both the transfrontal transventricular and transcallosal approaches were associated with a significant, higher morbimortality rate than was the translamina terminalis approach.[7] These results were independent of the degree of excision attempted, suggesting that some anatomical factor related to the route of access might have influenced the poorer outcome associated with the transventricular or transcallosal approaches. The performance of surgical maneuvers from a longer working distance together with a worse view of the surgical field provided by both approaches would prevent safe control of the basal adherences to hypothalamic walls in the not-strictly-intraventricular type of craniopharyngioma and might explain the less favorable prognosis; however, we think that ischemic injury to the hypothalamus caused by unnoticed damage to the perforating vessels may also be a factor. Perforating vessels are hidden from the view and control of the surgeon during the initial phases of debulking and dissection when the mass is approached through the foramen of Monro or the corpus callosum. Kassam and colleagues warned of the existence of tumoral engulfing of the posterior perforating vessels in some cases of retroinfundibular lesions, a situation that can only be anticipated with direct visual exploration of the area via an inferior approach, such as the EEA, before any undue manipulation of the tumor margins.

Despite the successful results presented in this work and ignoring for the present moment the difficulties that EEA entails in getting a watertight closure, most of the illustrative cases presented in the work by Kassam et al. involve lesions of a moderate size that are centered at the level of the third ventricle floor. These craniopharyngiomas may be the type of infundibulotuberal lesions more suitable to an excision attempt via the EEA. The safest and most appropriate approach to the largest tumors occupying the entire third ventricle and causing maximum distortions to vital neurovascular structures remains to be established. Validation of the results obtained by using the EEA will be impossible until an analysis of a larger series with a longer follow-up is performed. Meanwhile, this approach may be a useful tool in the study of the true anatomical and histological relationships that craniopharyngiomas have with surrounding neurovascular structures.

Jose M. Pascual, M.D., Ph.D.
Rodrigo Carrasco, M.D.
La Princesa University Hospital
Ruth Prieto, M.D.
Clinico San Carlos University Hospital
Francisco Gonzalez-Llanos, M.D.
Toledo Hospital Center
Fernando Alvarez, M.D.
Jose M. Roda, M.D., Ph.D.
La Paz University Hospital
Madrid, Spain

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