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History of Neurology

The first sixty-five craniopharyngioma operations in France

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ABSTRACT

Craniopharyngiomas (CPs) are benign epithelial tumors that develop along the hypothalamus–hypophyseal axis and were first described by Jakob Erdheim in 1904. These tumors have represented a challenge for surgeons since the rise of modern neurosurgery. The study of CPs is linked to the development of this surgical discipline in parallel with neuroendocrinology within the French school of neurology, led by Joseph Babinski. For the present study, all CP cases published in the French scientific literature before the development of modern neuroradiology were gathered, and 65 cases that underwent surgical procedures between 1921 and 1973 were selected. From our analysis of them, useful information has been obtained that can be applied to the management of CPs today. Most tumors were adamantinomatous CPs (62 patients) with an infundibulo-tuberal location (40.6%). The most frequent surgical route employed was subfrontal (69%). Selection of the surgical approach and degree of removal did not appear to have been influenced by the presumed topography of the tumor, and resulted in a poor outcome in 47% of patients. However, the authors were able to recognize the presence of symptoms indicating that the tumor had caused hypothalamic and/or infundibular damage, such as seen in the infundibulo-tuberal syndrome, first described by Claude and Lhermitte in 1917. At present, the optimal surgical approach and degree of removal are still the subject of debate, although the presence of clinical signs pointing to hypothalamic involvement by CPs should always be preoperatively accurately assessed to improve surgical outcomes.

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''What do you think will remain of me after I die? [...] My biggest achievement will be having opened the way for Martel and Vincent''[1].
Joseph Babinski [1].

1. Introduction

The birth of neuroendocrinology can be dated to the beginning of the 20th century in France, when Joseph Babinski (1857–1932) reported, in 1900, the case of an obese young girl who was sexually underdeveloped in *Revue Neurologique* (Fig. 1) [2]. These clinical signs were caused by a tumor located in the infundibular region with microscopy characteristics that ''strangely resemble those of the tumors of the jaw'' described by Jacques Onanoff, one of Babinski's students, in his doctoral dissertation [2–6]. In 1901, the Viennese physician Alfred Fröhlich (1871–1932) reported on a case with similar clinical characteristics that was later operated on by Anton von Eisenberg [7–10]. This patient was ultimately diagnosed with the same kind of tumor. Nevertheless, these neoplasms were not fully recognized until 1904, when the Austrian pathologist Jakob Erdheim (1874–1937) grouped them under the heading of ''tumors of the pharyngo-hypophyseal tract'' [11], which would later be replaced by the term ''craniopharyngiomas'' (CPs) [12].

The parallel development of neuroendocrinology and neurosurgery within the French neurological school, mainly among the pupils and successors of Joseph Babinski, are unquestionably related to the treatment of this type of tumor [1]. Although CPs are benign epithelial tumors, their clinical symptoms and surgical outcomes are strongly influenced by their topography to the point that infundibular CPs are nowadays considered prototypical lesions associated with infundibulo-hypothalamic injury [5,13]. This fact highlights the importance of a thorough study and correlation of clinical features with pathological findings in each CP case to optimalize the therapeutic strategy. As the clinical data are meticulously described and illustrated in early papers dealing with this kind of neoplasm, an historical review should prove useful even today (Figs. 2 et 3).

The present report has gathered together all CP cases described in the French medical literature before the advent of modern neuroradiology (computed tomography [CT] and magnetic resonance imaging [MRI]) [5], and assessed the influence that clinical and pathological CP features had on patient outcomes among the first cases operated on in France.

2. Materials and methods

2.1. Database generation and case-selection criteria

A thorough search was performed for well-detailed CP cases reported in the French medical literature from 1700 to 1973. The search involved all cases verifying the diagnosis of CP with surgical, neuroradiological and/or necropsy (autopsy) findings in official medical journals, as well as in specialized texts and monographs, written in French. Our search comprised articles appearing in the PubMed, Medline and Scopus databases after introducing the keywords ''craniopharyngioma'' and ''craniopharyngiome''. The reference lists of the articles obtained were also reviewed to identify relevant non-indexed manuscripts. A total of 128 cases were collected [5], involving 71 patients who underwent surgical treatment. However, of these, only the 65 cases with a complete description of the surgical procedure were ultimately selected for the current analysis (Table 1) [14–35].

2.2. Categorization of variables

The data collected from each case are summarized in Table 1, and include epidemiological data (gender, age), pathological features of the neoplasm (size, shape, histological type and topography), surgical variables (type of approach and degree of removal) and outcome and follow-up of each patient. Postoperative outcomes were classified into four grades: (1) good (no permanent disabling neurological or neuropsychological deficits); (2) fair (permanent endocrine, neurological or neuropsychological disturbances, but allowing independent living); (3) poor (disabling sequelae requiring continuous support); and (4) postoperative death. Any ancillary diagnostic tests used by each author and the examinations that allowed a precise histological and topographical diagnosis to be made are also described.

2.3. Topographical classification of CPs

Tumors were classified into five groups according to their relationship to the floor of the third ventricle [36–38]:

- sellar/suprasellar: tumor circumscribed to the sellar and/or suprasellar area;
- pseudointraventricular: suprasellar neoplasm pushing against an intact third-ventricle floor, mimicking an intraventricular tumor;
- secondary intraventricular: suprasellar CP invading the third-ventricle cavity after breaking through the floor;
- strictly intraventricular: CPs located entirely within the third-ventricle cavity with an intact floor;
- and non-strictly intraventricular or infundibulo-tuberal: tumor originating within the third-ventricle floor and occupying the infundibulo-tuberal area while growing into the third-ventricle cavity.

2.4. Statistical analysis

These analyses were all performed using SPSS Version 21 software (IBM Corp., Armonk, NY, USA). Frequencies and descriptive statistics for the various categorical variables under study were obtained. Bilateral correlation between pairs of categorical variables were tested using the asymptotic Chi^2, or the Monte Carlo or exact test for Chi^2 in the case of frequency tables containing cells with either zero elements or

[1] ''Que pensez-vous qu'il restera de moi après ma mort? [...] mon meilleur titre de gloire sera d'avoir ouvert la voie à Martel et à Vincent''.