Craniopharyngioma (CP)

A craniopharyngioma (CP) is an embryonic malformation of the sellar region and parasellar region.

Its relation to Rathke's cleft cyst (RCC) is controversial, and both lesions have been hypothesized to lie on a continuum of ectodermal cystic lesions of the sellar region.

Craniopharyngiomas frequently grow from remnants of the Rathke pouch, which is located on the cisternal surface of the hypothalamic region. These lesions can also extend elsewhere in the infundibulohypophyseal axis.

These tumors can also grow from the infundibulum or tuber cinereum on the floor of the third ventricle, developing exclusively into the third ventricle.

Jakob Erdheim (1874-1937) was a Viennese pathologist who identified and defined a category of pituitary tumors known as craniopharyngiomas. He named these lesions “hypophyseal duct tumors” (Hypophysenganggeschwülste), a term denoting their presumed origin from cell remnants of the hypophyseal duct, the embryological structure through which Rathke's pouch migrates to form part of the pituitary gland. He described the two histological varieties of these lesions as the adamantinomatous and the squamous-papillary types. He also classified the different topographies of craniopharyngiomas along the hypothalamus-pituitary axis. Finally, he provided the first substantial evidence for the functional role of the hypothalamus in the regulation of metabolism and sexual functions. Erdheim's monograph on hypophyseal duct tumors elicited interest in the clinical effects and diagnosis of pituitary tumors. It certainly contributed to the development of pituitary surgery and neuroendocrinology. Erdheim's work was greatly influenced by the philosophy and methods of research introduced to the Medical School of Vienna by the prominent pathologist Carl Rokitansky. Routine practice of autopsies in all patients dying at the Vienna Municipal Hospital (Allgemeines Krankenhaus), as well as the preservation of rare pathological specimens in a huge collection stored at the Pathological-Anatomical Museum, represented decisive policies for Erdheim's definition of a new category of epithelial hypophyseal growths. Because of the generalized use of the term craniopharyngioma, which replaced Erdheim's original denomination, his seminal work on hypophyseal duct tumors is only referenced in passing in most articles and monographs on this tumor.

Jakob Erdheim should be recognized as the true father of craniopharyngiomas [1)].

Epidemiology

Incidence of 0.13 cases per 100,000 people every year. They represent approximately 2%–5% of all primary brain tumors and have a bimodal presentation in children aged 5–14 and adults aged 55–65 years

The incidence of craniopharyngioma in the United States was estimated from two population-based cancer registries that include brain tumors of benign and borderline malignancy: the Central Brain Tumor Registry of the United States (CBTRUS) and the Los Angeles county Cancer Surveillance Program. Information on additional pediatric tumors was available from the Greater Delaware Valley Pediatric Tumor Registry (GDVPTR). The overall incidence of craniopharyngioma was 0.13 per 100,000 person years and did not vary by gender or race. A bimodal distribution by age was noted with peak incidence rates in children (aged 5-14 years) and among older adults (aged 65-74 years in CBTRUS and 50-74 years in Los Angeles county). Survival information was available from GDVPTR and the National Cancer Data Base (NCDB), a hospital-based reporting system. In the NCDB, the 5-year

survival rate was 80% and decreased with older age at diagnosis. Survival is higher among children and has improved in recent years.

Craniopharyngioma is a rare brain tumor of uncertain behavior that occurs at a rate of 1.3 per million person years. Approximately 338 cases of this disease are expected to occur annually in the United States, with 96 occurring in children from 0 to 14 years of age [2].

The Childhood Cancer Registry of Piedmont, Italy, estimates an incidence of 1.4 cases per million children per year. Similar data are provided by other registries in Western countries, while higher rates have been observed in Asia and Africa. There are no known specific environmental risk factors for craniopharyngioma, and genetic predisposition is not demonstrated [3].

Zacharia et al., used the surveillance, epidemiology and end results program (SEER) database to identify patients who received a diagnosis of craniopharyngioma during 2004-2008. They analyzed clinical and demographic information, including age, race, sex, tumor histology, and treatment. Age-adjusted incidence rates and age, sex, and race-adjusted expected survival rates were calculated. They used Cox proportional hazards models to determine the association between covariates and overall survival. We identified 644 patients with a diagnosis of craniopharyngioma. Black race was associated with an age-adjusted relative risk for craniopharyngioma of 1.26 (95% confidence interval [CI], 0.98-1.59), compared with white race. One- and 3-year survival rates of 91.5% (95% CI, 88.9%-93.5%), and 86.2% (95% CI, 82.7%-89.0%) were observed for the cohort; relative survival rates were 92.1% (95% CI, 89.5%-94.0%) and 87.6% (95% CI, 84.1%-90.4%) for 1- and 3-years, respectively. In the multivariable model, factors associated with prolonged survival included younger age, smaller tumor size, subtotal resection, and radiation therapy. Black race, on the other hand, was associated with worse overall survival in the final model. They demonstrated that >85% of patients survived 3 years after diagnosis and that subtotal resection and radiation therapy were associated with prolonged survival. They also noted a higher incidence rate and worse 1- and 3-year survival rates in the black population. Future investigations should examine these racial disparities and focus on evaluating the efficacy of emerging treatment paradigms [4].

Origin

Its relation to Rathke's cleft cyst (RCC) is controversial, and both lesions have been hypothesized to lie on a continuum of cystic ectodermal lesions of the sellar region.

It grows close to the optic nerve, hypothalamus and pituitary gland.

Classification

Craniopharyngioma Classification.

Craniopharyngioma Natural History

Craniopharyngioma natural history.

Clinical Features

see Craniopharyngioma Clinical Features.

Diagnosis

see Craniopharyngioma Diagnosis.

Differential diagnosis

Rathke's cleft cyst.

Compared with craniopharyngiomas, sellar gliomas presented with a significantly lower ratio of visual disturbances, growth hormone deficiencies, lesion cystic changes, and calcification. Sellar gliomas had significantly greater effects on the patients' mentality and anatomical brain stem involvement [5].

Co-occurrence

Simultaneous sellar-suprasellar craniopharyngioma and intramural clival chordoma, successfully treated by a single staged, extended, fully endoscopic endonasal approach, which required no following adjuvant therapy is reported [6].

Treatment

see Craniopharyngioma treatment

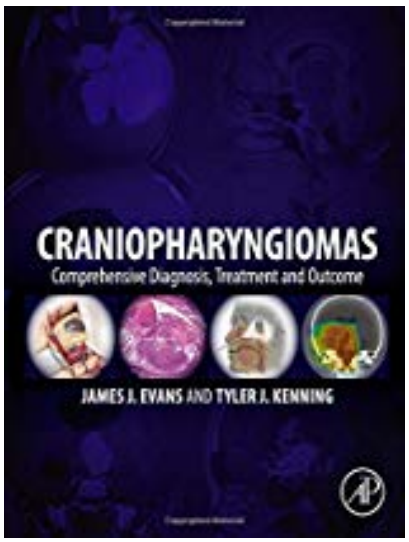
Outcome

see Craniopharyngioma outcome

Recurrence

see Craniopharyngioma recurrence.

Books



Craniopharyngioma: Surgical Treatment.

10 Selected Works

Craniopharyngioma Selected Works.

Case series

see Craniopharyngioma case series.

Case reports

see Craniopharyngioma case reports.

1)
Pascual JM, Rosdolsky M, Prieto R, Strauβ S, Winter E, Ulrich W. Jakob Erdheim (1874-1937): father of hypophyseal-duct tumors (craniopharyngiomas). Virchows Arch. 2015 Jun 19. [Epub ahead of print] PubMed PMID: 26089144.
2)
Bunin GR, Surawicz TS, Witman PA, Preston-Martin S, Davis F, Bruner JM. The descriptive epidemiology of craniopharyngioma. J Neurosurg. 1998 Oct;89(4):547-51. PubMed PMID: 9761047.
3)
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4)
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6)
Iacoangeli M, Rienzo AD, Colasanti R, Scarpelli M, Gladi M, Alvaro L, Nocchi N, Scerrati M. A rare case of chordoma and craniopharyngioma treated by an endoscopic endonasal, transtubercular transclival approach. Turk Neurosurg.2014;24(1):86-9. doi: 10.5137/1019-5149.JTN.7237-12.0. PubMed PMID: 24535799.