Craniopharyngioma Clinical Features

Typical manifestations at diagnosis are some combination of headache, visual impairment, Polyuria/Polydipsia SIAD, diabetes insipidus, growth retardation, and significant weight gain.

The presence of hydrocephalus, distortion of the circle of Willis, and large tumor volume were associated with headaches, and the last 2 variables were also associated with more severe and frequent headaches. Radiation treatment and insertion of the Ommaya reservoir were associated with reduced headache frequency. In conclusion, headaches are common in patients with craniopharyngioma and are likely related to tumor size and volume. In most patients, headaches improve with successful tumor treatment [1)]

CP is frequently diagnosed after long duration of history (DOH), especially in older children. However, DOH was not associated with tumor size, hypothalamic involvement (HI), survival, or functional capacity (FC) [2)].

Visual and neurological deficits necessitate rapid diagnostic workup.

Infundibulo tuberal syndrome

see Hydrocephalus in craniopharyngioma

A significant number of patients with craniopharyngioma are GH deficient. The safety of GH replacement in these subjects has not been established.

Obesity

Craniopharyngiomas are tumors located in the hypothalamic region which leads to obesity in about 50% of cases. Long-term efficacy and safety of bariatric surgery are lacking in this peculiar population. The aim of this study is to determine the 5-year weight loss and resolution of type 2 diabetes (T2D) after bariatric surgery in patients operated on craniopharyngioma who had developed hypothalamic obesity.

Materials and methods: This is a multicenter french retrospective case-control study. Subjects with craniopharyngioma (n = 23) who underwent sleeve gastrectomy (SG) (n = 9) or Roux-en-Y gastric bypass (RYGB) (n = 14) (median age 35 years [25;43] and BMI 44.2 kg/m2 [40.7; 51.0]; 8/23 with T2D) were individually matched to 2 subjects with common obesity for age, gender, preoperative body mass index, T2D, and type of surgery.

Results: TWL% after 1 and 5 years was lower in the craniopharyngioma group than in the control group: 23.1 [15.4; 31.1] (23/23) vs 31.4 [23.9; 35.3] at 1 year ($p = 0.008$) (46/46) and 17.8 [7.1; 21.9] (23/23) vs 26.2 [18.9; 33.9] at 5 years ($p = 0.003$) (46/46). After RYGB, TWL% was lower in the craniopharyngioma group compared to the control group ($p < 0.001$) and comparable after SG both at 1 and 5 years. No difference between the two groups was observed in T2D remission rate and in early and late adverse events. No hormonal deficiency-related acute disease was reported.

Conclusions: Bariatric surgery induced a significant weight loss in the craniopharyngioma group at 1 and 5 years, but less than in common obesity. SG may be more effective than RYGB but this remains to be demonstrated in a larger cohort [3].

Psychiatric disorders

Pascual et al. from the University Hospital La Princesa, Puerta de Hierro University Hospital, Ramón y Cajal University Hospital, Hospital del Sureste, Madrid, Spain. Department of Surgery and Experimental Medicine, University of Ferrara, Italy. Independent Medical Translator, Jenkintown, PA, US. School of Medicine, Technische Universität, Dresden,Germany. Statistics Department, Computing Center, C.S.I.C. Madrid, Spain. investigated a collection of 210 craniopharyngiomas published from 1823 to 2017 providing detailed clinical and pathological information about psychiatric disturbances, and compared the hypothalamic damage in this cohort with the present in a control cohort of 105 cases without psychiatric symptoms.

Psychiatric disorders occurred predominantly in patients with craniopharyngiomas developing primarily at the tuberoinfundibular region (45%) or entirely within the third ventricle (30%), mostly affecting adult patients (61%, p<0.001). Most tumors without psychic symptoms developed beneath the third ventricle floor (53.5%, p<0.001), in young patients (57%, p<0.001). Psychiatric disturbances were classified in six major categories: i) Korsakoff-like memory deficits, 66%; ii) behavior/personality changes, 48.5%; iii) impaired emotional expression/control, 42%; iv) cognitive impairments, 40%; v) mood alterations, 32%; and vi) psychotic symptoms, 22%. None of these was associated with hydrocephalus. Severe memory deficits occurred with damage of the mammillary body (p<0.001). Mood disorders occurred with compression/invasion of the third ventricle floor and/or walls (p<0.012). Coexistence of other hypothalamic symptoms such as temperature/metabolic dysregulation or sleepiness favored the emergence of psychotic disorders (p<0.008). Postoperative psychiatric outcome was better in strictly intraventricular craniopharyngiomas than in other topographies (p<0.001). A multivariate model including the hypothalamic structures involved, age, hydrocephalus and hypothalamic symptoms, predicts the appearance of psychiatric disorders in 81% of patients.

CPs primarily involving the hypothalamus represent a neurobiological model of psychiatric and behavioral disorders [4].

Autonomic dysfunction

Autonomic dysfunction in Craniopharyngioma.

Chemical meningitis

Chemical meningitis

1)
Khan RB, Merchant TE, Boop FA, Sanford RA, Ledet D, Onar-Thomas A, Kun LE. Headaches in children with craniopharyngioma. J Child Neurol. 2013 Dec;28(12):1622-5. doi: 10.1177/0883073812464817. Epub 2012 Nov 8. PubMed PMID: 23143722; PubMed Central PMCID: PMC4264380.

2)
Hoffmann A, Boekhoff S, Gebhardt U, Sterkenburg AS, Daubenbüchel AM, Eveslage M, Müller HL. History before diagnosis in childhood craniopharyngioma: associations with initial presentation and long-term prognosis. Eur J Endocrinol. 2015 Dec;173(6):853-62. doi: 10.1530/EJE-15-0709. Epub 2015 Sep 21. PubMed PMID: 26392473.
3)
Faucher P, Carette C, Jannot AS, Gatta-Cherifi B, Van Straaten A, Piquet MA, Raverot G, Alligier M, Batisse T, Ziegler O, Drui D, Bretault M, Farigon N, Slim K, Genser L, Poghosyan T, Vychnevskaia K, Blanchard C, Robert M, Gronnier C, Poitou C, Czernichow S. Five-Year Changes in Weight and Diabetes Status After Bariatric Surgery for Craniopharyngioma-Related Hypothalamic Obesity: a Case-Control Study. Obes Surg. 2022 May 6. doi: 10.1007/s11695-022-06079-9. Epub ahead of print. PMID: 35524022.
4)
Pascual JM, Prieto R, Castro-Dufourny I, Mongardi L, Rosdolsky M, Strauss S, Carrasco R, Barrios L. Craniopharyngiomas primarily involving the hypothalamus: a model of neurosurgical lesions to elucidate the neurobiological basis of psychiatric disorders. World Neurosurg. 2018 Sep 18. pii: S1878-8750(18)32099-0. doi: 10.1016/j.wneu.2018.09.053. [Epub ahead of print] PubMed PMID: 30240857.