21 Other Malformative Tumors and Tumor-Like Lesions

21.1 Craniopharyngiomas

Synonyms in the older literature: Tumors or cysts of the hypophyseal duct, cysts or tumors of Rathke’s pouch, tumors of Rathke’s cleft, cranopharyngeal pouch tumors, suprasellar cysts, pituitary stalk tumors, Erdheim’s tumors, adamantinomas or ameloblastomas of the pituitary region

Introduction

This well-known tumor was described very early and is listed in the WHO classification system as *craniopharyngioma*.

Historical Note and Definition

Some of the older descriptions of “Markschwämme” or medullary carcinomas of the pituitary gland were most probably true craniopharyngiomas. However, we are indebted to ERDHEIM (1904) for the first correct interpretation and adequate description of these tumors. STRADA (1911) and CRITCHLEY and IRONSIDE (1926) listed the early cases. Among subsequent contributions the following especially are worth mentioning: McLEAN (1930), FRAZIER and ALPERS (1931), WITTERMANN (1936), LOVE et al. (1939), MÜLLER and WOHLFAHRT (1950), MATSON (1964), SVIEN (1965), BANNA (1976).

Epidemiology (Age – Sex – Frequency – Site)

*Age:* The craniopharyngiomas can definitely be generally considered as tumors of childhood and adolescence, but they do also occur in adults (Fig. 9k). CUSHING (1932, 1935) himself saw two patients over 60 years of age; CAMPBELL and HUDSON (1960) had three patients over 60 years; KITANO et al. (1981), one of 3 years; and TABADDOR et al. (1974) reported one case of a neonatal craniopharyngioma. The peak incidence is between the ages of 10 and 25 (Fig. 9k). The oldest of our patients was 62, the youngest 5 years old. In childhood and adolescence craniopharyngiomas of the chiasmal region are one of the most common tumors. In FRAZIER and ALPERS’ series (1931) 70% of the patients were younger than 20 years; however, in our early series (1956) only 43.18% fell in this age group.

*Sex:* There is a certain male proponderance with a ratio of around 3:2 (116 male and 69 female patients in our series of 9000 cases). In MÜLLER and WOHLFART’s series (1950) it was only 5:4.
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Frequency: The craniopharyngiomas accounted for 1.2% of all tumors in our series of 9000 cases and for 4.6% in the series of CUSHING (1932, 1935). In the tumors collected by FRAZIER and ALPERS (1931, 1934), of 244 lesions around the sella only 14 were craniopharyngiomas (11 of these were histologically verified). In the publications of McLEAN (1930, 1936a) they made up 30% of the “hypophyseal tumors.” In the series of BENNET (1946) and of FRAZIER and ALPERS (1931) they accounted for 5.7% of the sellar and parasellar tumors. However, like the pineal tumors, their frequency is several times as high (SANO 1983a, 5.6%) in the Far East.

Site: Since craniopharyngiomas occur exclusively in the region of the sella, they are thought to originate from vestigial remnants of the craniopharyngeal duct (Rathke’s pouch). The only ways they vary in site are in their relation to the diaphragma sellae (Figs. 147, 148) and in the direction of extension from this point. Thus, both intrasellar and suprasellar craniopharyngiomas (Fig. 148; GP, Figs. 258, 259) occur, and also combined growth in both spaces.

Of these, the intrasellar types are initially separated from the brain by the dura and arachnoid. As they grow, however, they push the diaphragma upward – generally breaking through it – growing in the direction of the third ventricle, where they excavate a bed for themselves from below (GP, Fig. 258). The suprasellar (Fig. 149) type starts in the arachnoid of the basal cisterns and pushes directly against the third ventricle; the ventricular floor then becomes paper-thin and tears, so that the tumor capsule abuts directly onto the ventricular wall.