Intraventricular Squamous Papillary Craniopharyngioma

Report of a Case with Intraoperative Imprint Cytology

Manoharan Madhavan M.B.B.S., M.D.(Path), M.I.A.C., Jain George P., M.B.B.S., M.S., M.Ch. (Neurosurg), J. Abdullah Jafri, M.D., Ph.D., and Zamzuri Idris, M.B.Bch. (M. Neurosurg)

Background

Squamous papillary craniopharyngioma is a distinct entity, and its cytologic features may be misleading. Because of the rarity of this tumor, this case is being reported with a note on the cytologic features

Case

A 56-year-old Malay man who had 1-month history of generalized lethargy was admitted for altered sensorium. On examination, he was found to have neck stiffness, bilateral papilledema and generalized atrophy of muscles, with reduced power in all limbs. Magnetic resonance imaging of the brain showed a solid mass in the third ventricle causing obstructive hydrocephalus. Intraoperative cytology of the mass diagnosed intraventricular meningioma. However, the final histopathologic examination revealed squamous papillary craniopharyngioma.

Cytology showed an abundant cellular yield composed essentially of sheets and groups of polygonal cells with a moderate amount of cytoplasm and bland, round nuclei.

Conclusion

Craniopharyngioma, squamous papillary type, is a rare entity and usually occurs in adults as an intraventricular solid tumor. Awareness of this entity will aid in arriving at the correct cytologic diagnosis. (Acta Cytol 2005;49:431–434)

Keywords: craniopharyngioma, papillary; cerebral ventricles, intraoperative period, imprint cytology.

Craniopharyngioma constitutes about 3% of brain tumors.[1] Most patients with craniopharyngioma are in the first or second decade of life. The location of the tumor is usually suprasellar, although it may occupy the sella as well. It is usually composed of adamantinomatous-type epithelium. In rare cases the tumor has a macroscopic papillary appearance, lined with pseudopapillary squa-

From the Department of Pathology and Neuroscience Unit, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia

Dr. Madhavan is Associate Professor, Department of Pathology.

Dr. George is Lecturer, Neuroscience Unit.

Dr. Jafri is Professor, Neuroscience Unit.

Dr. Idris is Trainee Lecturer, Neuroscience Unit.

Address reprint requests to: Manoharan Madhavan, M.B.B.S., M.D.(Path), M.I.A.C., Department of Pathology, School of Medical Sciences, Universiti Sains Malaysia, 16150, Kubang Kerian, Kelantan, Malaysia (madhavan@kb.usm.my)

Financial Disclosure: The authors have no connection to any companies or products mentioned in this article.

Received for publication April 2, 2004.

Accepted for publication May 29, 2004.

0001-5547/05/4904-0431/$19.00/0

mous epithelium, and is termed *squamous papillary type*. It is considered a distinct entity as it is usually solid, occurs exclusively in adults and has a better postoperative outcome. The intraoperative cytology may be misleading, as the features are different from those of the adamantinomatous type. This case is being reported because of its rarity, with a note on the cytologic features.

Case Report

A 56-year-old Malay man who had been treated for dengue fever in another local hospital was admitted to our neurosurgical unit following a 1-month history of generalized lethargy. He became bed bound and developed altered sensorium He appeared drowsy and did not respond to questions. This happened gradually and progressively. The patient had a history of a central headache prior to his deteriorating condition. There was no history to suggest endocrine or hypothalamic disturbances other than a previous history of intermittent fever that led to the incorrect diagnosis of dengue fever.

On the day of admission the patient was drowsy, nonresponsive and confused. His vital signs were normal. He had neck stiffness, and on cranial nerves examination there was bilateral papilledema with midsize pupils, 4 mm in diameter, which reacted sluggishly to direct light. Generalized atrophy of the muscles with reduced power in all limbs was noted. In addition, the left lower limb was slightly hypertonic, with slightly increased reflex. The Babinski sign was positive on the left side. All the basic blood investigations were normal.

Magnetic resonance imaging of the brain (Figure 1) showed a solid tumor measuring 2.9×3.4×3.5 cm situated exclusively inside the third ventricle at the level of the foramen of Monro, causing obstructive hydrocephalus. Total microsurgical excision of the tumor was performed through a pericoronal, interhemispheric, transcallosal approach. Intraoperatively the tumor appeared predominantly soft, with some firm areas and a moderate amount of vascularity, which could be easily aspirated.

Intraoperative imprint cytology smears, stained with hematoxylin-eosin, revealed a high cellular yield The cells were predominantly polygonal, with a moderate amount of cytoplasm and uniform, bland, round nuclei. They were arranged predominantly in syncytial groups, with some lying singly (Figure 2) No evidence of keratinization, calcified debris or cellular whorling was present. With the presence of a syncytial cell arrangement, a diagnosis of intraventricular meningioma was made. However, histopathologic examination of the tumor revealed a solid sheet of squamous cells punctuated by a fibrovascular core (Figure 3). No evidence of keratinization or basaloid-type squamous cells and calcified debris was present. Hence, a diagnosis of craniopharyngioma, squamous

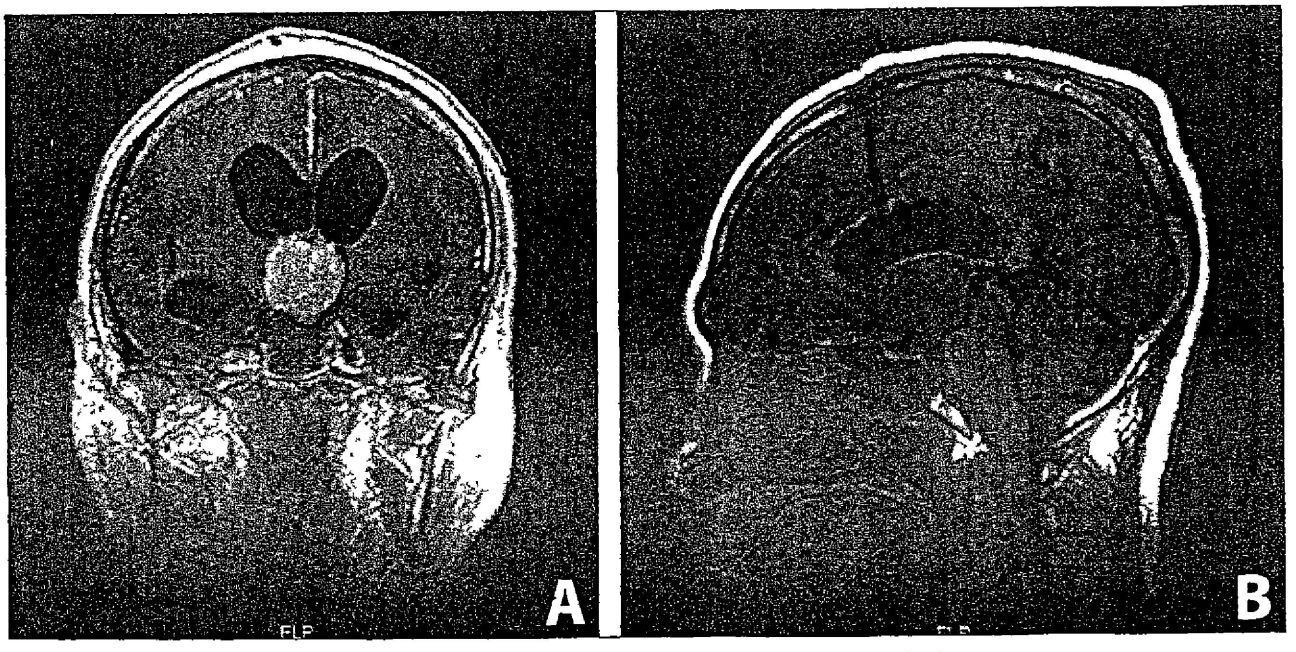

Figure 1 (A) Coronal magnetic resonance image of the brain, after gadolinium, showing an enhancing, rounded mass in the third ventricle obstructing the flow of cerebrospinal fluid at the foramen of Monro. (B) Sagittal T1-weighted image showing the mass of isointensity to the brain parenchyma occupying the anterior two-thirds of the third ventricle.

Figure 2 Imprint cytology showing polygonal cells arranged in syncytial groups (hematoxylin-eosin, ×100)

papillary type, was made.

The patient recovered uneventfully after the operation. However, at the time of discharge, on the 12th postoperative day, he died of acute myocardial infarction.

Discussion

There are 2 distinct clinicopathologic entities described as craniopharyngioma. The more common entity is the adamantinomatous type. It usually occurs during the first and second decades of life. Few cases have been observed in people in their 70s and 80s.[2] Craniopharyngiomas, 10–20% are squamous papillary type,[2,3] and it typically affects adults, as in our case. A case affecting a 10-year-old boy has been reported.[3]

The adamantinomatous type usually involves the suprasellar region and extends into the adjacent areas, including the third ventricle. A few cases have been seen in the sella and infrasellar region.[4,5] In the present case the tumor was situated in the third ventricle. Craniopharyngioma involving the third ventricle exclusively is typically the squamous papillary type, although it may affect other regions as well.

Craniopharyngioma of the adamantinomatous type is usually cystic but is sometimes solid or combined solid and cystic. In contrast, the papillary type is frequently solid.[6] A few reported cases were both cystic and solid or entirely cystic.[2]

Microscopically the squamous papillary type is composed of papillary structures and mature squamous epithelium in solid areas and attenuated cells in cystic areas.[4] The present case was entirely solid and composed of solid sheets of mature squamous cells punctuated by a fibrovascular core. Stellate reticulum, microcysts, and flaky and wet keratin were conspicuously absent. Hence, a diagnosis of squamous papillary type was made. Occasionally both types of epithelium are present together. Calcification, typical of the adamantinomatous type, is extremely rare in the squamous papillary type. If present, it occurs as microcalcification.[4]

The cytologic features have been described only for the adamantinomatous type[7] and are composed of a mixture of numerous keratinized cells, characteristic squamous cells of basal type, ghost cells, macrophages and calcified debris. In our case an intraoperative cytologic diagnosis was sought. Cytology showed an abundant cellular yield composed essentially of sheets and groups of polygonal cells with a moderate amount of cytoplasm and bland, round nuclei. In view of the syncytial arrangement of cells and small clustering of cells resembling cell whorling, a diagnosis of intraventricular meningioma was suggested. No evidence of keratinization was present. Even if it were present, it might have suggested another diagnosis, such as epidermoid cyst or Rathke's cleft cyst with squamous metaplasia.[8] The solid nature of the lesion and awareness of it might suggest the diagnosis.

Craniopharyngiomas are usually treated with gross total resection. Radiotherapy may be given if residual tumor is present after surgical excision. Between the 2 histologic types of this tumor, no difference in resectability, efficacy of radiotherapy and overall survival has been documented.[4,9] Adamson et al[6] showed that the postoperative outcome was good in squamous papillary type, as there was clear demarcation between the tumor and adjacent brain. However, a case of meningeal seeding from surgical manipulation has been reported.[10]

Figure 3 The tumor was composed of a solid sheet of squamous cells punctuated by a fibrovascular core (hematoxylin-eosin, ×25).

References

1 Lederman GS, Recht A, Loeffler JS, Dubuisson D, Kleefield J, Schnitt SJ Craniopharyngioma in an elderly patient. Cancer 1987,60 1077–1080

2 Crotty TB, Scheithauer BW, Young WF Jr, Davis DH, Shaw EG, Miller GM, Burger PC Papillary craniopharyngioma A clinicopathological study of 48 cases J Neurosurg 1995,83 206–214

3 Giangaspero F, Burger PC, Osborne DR, Stein RB Suprasellar papillary squamous epithelioma ("papillary craniopharyngioma") Am J Surg Pathol 1984,8 57–64

4 Petito CK, DeGirolami U, Earle KM Craniopharyngiomas A clinical and pathological review. Cancer 1976,37.1944–1952

5. Kachhara RR, Nair SS, Gupta AK, Radhakrishnan VV, Bhattacharya RN Infrasellar craniopharyngioma mimicking a clival chordoma A case report. Neurol India 2002,50 198–200

6 Adamson TE, Wiestler OD, Kleihues P, Yasargil MG. Correlation of clinical and pathological features in surgically treated craniopharyngiomas J Neurosurg 1990;73.12–17

7. Smith AR, Elsheikh TM, Silverman JF Intraoperative cytologic diagnosis of suprasellar and sellar cystic lesions Diagn Cytopathol 1999,20 137–147

8 Parwani AV, Taylor DC, Burger PC, Erozan YS, Olivi A, Ali SZ Keratinized squamous cells in fine needle aspiration of the brain. Cytopathologic correlates and differential diagnosis Acta Cytol 2003,47:325–331

9 Honegger J, Grabenbauer GG, Paulus W, Fahlbusch R. Regression of a large solid papillary craniopharyngioma following fractionated external radiotherapy J Neurooncol 1999,41:261–266

10 Elmaci L, Kurtkaya-Yapicier O, Ekinci G, Sav A, Pamir MN, Vidal S, Kovacs K, Scheithauer BW· Metastatic papillary craniopharyngioma Case study and study of tumor angiogenesis Neurooncol 2002,4 123–128