SUPRASELLAR PAPILLARY SQUAMOUS EPITHELIOMA (PAPILLARY CRANIOPHARYNGIOMA)

Maj T CHATTERJEE ^{*}, Dr SANGEETA DESAI ⁺,
Lt Col R LAKHTAKIA [#], Lt Col SS GILL ^{**},
Lt Col S SATYANARAYANA ⁺⁺

MJAFI 2000; 56 : 158-160

KEY WORDS:Craniopharyngioma; Papillary squamous epithelioma.

Introduction

A case of a suprasellar papillary neoplasm is presented and the features of this lesion are contrasted with those of classical adamantinomatous craniopharyngioma. Characteristics distinguishing the former from the latter include a predominate, and perhaps exclusive occurrence in adulthood; frequent radiologic solidity and absence of calcification; macroscopic papillary nature; and microscopically, a well differentiated papillary squamous epithelium without calcification, palisaded cells, or Keratoid nodule. This papillary lesion is a clinicopathological entity [1] distinct from the classical adamantinomatous craniopharyngioma; also the papillary lesions can be completely excised whereas this generally eludes a surgeon dealing with adamantinomatous craniopharyngioma.

Case Report

A 33-year-old male was referred to a reference centre with complaints of lethargy, hypersomnolence and decreased vision in right eye. This was associated with headache more on the right side, as well as vomiting off and so. Symptoms were persistent for almost a year. Ophthalmological examination revealed right temporal hemianopsia. Examination of CNS revealed exaggerated DTJs in right upper and Lower limbs. There was no sensory deficit and no sign of cerebellar disease.

Relevant investigations were carried out. X-ray skull showed erosion of dorsum Sella. CT scan brain revealed a hypodense intracranial uncalcified suprasellar SOL involving left thalamus and 3rd ventricle (Fig-1).

MRI revealed a lobulated heterogenous mass in the suprasellar region measuring 4x4x3cms with a large cystic content. Mass was separate from the pituitary gland.

The patient underwent a right parietal craniotomy with near total excision of tumour. It could be removed in piecemeal. Pathological examination (Gross) revealed an externally smooth cyst filled with clear fluid. The multi-lobulated cavity was delineated by smooth walls thrown focally into small papillations.

Histopathologically the neoplasm consisted of cords of squamous epithelium embedded in loose connective tissue. The adjacent compressed nervous tissue was gliotic. The squamous epithelium showed progressive flattening from the basal layer. There was no evidence of Keratohyaline granules or nuclear squames. Papillae formations were excessive. Keratoid nodules, calcification and cholesterol clefts were absent. Basal layer showed no palisading (Fig-2 and 3). A diagnosis of Papillary Craniopharyngioma was given.

Discussion

There has been a tendency to consolidate histologically disparate suprasellar epithelial lesions under the term 'Craniopharyngioma' [1-3]. Some authors have included lesions which appear to be simple epidermoid cysts or Rathke's pouch cysts, while others have restricted the term craniopharyngioma to neoplasms

Fig. 1 : CT scan brain reveals a hypodense, intracranial uncalcified suprasellar space occupying lesion involving left thalamus and 3rd ventricle.

^{*} Graded Specialist (Pathology), Base Hospital, Delhi Cantt-10, ⁺ Assistant Pathologist, Tata Memorial Hospital, Mumbai, [#] Classified Specialist (Pathology), Department of Pathology, AIIMS, New Delhi. ^{**} Classified Specialist (Pathology), Command Hospital (SC), Pune 40, ⁺⁺ Classified Specialist (Pathology), Command Path Lab (WC), New Delhi-10.

Fig. 2 : Papillary craniopharyngioma - representative photograph of the surface lesion depicting papillae with finger - like projections.

with a more complicated histologic architecture. Within the latter group are two lesions (1) Classic adamantinomatous craniopharyngioma and (2) a Squamous papillary lesion that is the subject of this report. It is suggested that this squamous papillary lesion represents a clinicopathologic entity (1) distinct from the adamantinomatous craniopharyngioma. Macroscopically the papillary lesions are cysts containing variable number of papillations and therefore contrast with the fibrotic, often necrotic, non papillary adamantinomatous lesion filled with 'motor oil' fluid. The papillary nature and the absence of a pearly sheen and pilosebaceous contents differentiate the lesion from epidermoid and dermoid cysts [1].

Microscopically the papillary lesions are formed entirely of squamous epithelium in anastomosing trabeculae whose surface maturation and detachment produces prominent papillae. The calcified adamantinomatous lesion, in contrast, does not exhibit diffuse uniform surface maturation and contains prominent palisades of epithelial cells around cavities formed by degeneration of the fibrovascular stroma. The Keratohyoid nodule seen in classic calcified craniopharyngioma are not seen in this variant. Keratoid nodules [1] are bodies formed by cornified cells with copious pink cytoplasm that arise abruptly from surrounding neoplasm without a progressive squamous maturation. Most of the classic cases have a adamantinomatous pattern. No giant cell reaction or fibrosis is seen in the papillary variant in contrast to the classic one. The papillae, keratinised pearls and absence of anuclear squames or adenexal structures microscopically differentiate the papillary lesion from the epidermoid and dermoid cyst [4].

Clinically the papillary lesion has been described exclusively in adults [5-7]; the adamantinomatous

Fig. 3 : Papillary craniopharyngioma - deeper lesion photograph formed entirely of squamous epithelium in anastomosing trabeculae abutting on neural tissue surface papillae seen on left of photograph.

neoplasm occurs in both age groups and is clearly more common in children. There is predominance of dementia and signs of increased intracranial pressure in adults [11], whereas in children the predominant manifestation is of endocrine deficiency. Visual symptoms are common in both [7-10]. Prognosis was better in adults with this Squamous lesion in contrast to both age groups with adamantinomatous neoplasm [7]. Also the papillary lesion can be completely excised whereas this generally eludes a surgeon dealing with adamantinomatous craniopharyngioma. In another report the 5 year survival in adult patients without calcification was 61 percent as compared to 35 percent in adult patients with calcification [15]. Radiologically the Craniopharyngioma also appears to be heterogeneous and the reported data are consistent with the presence of two lesions : the usually cystic, calcified adamantinomatous neoplasm of children and in some, adults, and the non calcified solid papillary lesion described here [12-14].

The present case report describes the pathological, clinical and radiological features of a papillary lesion in an adult that contrasts with those of the classic adamantinomatous craniopharyngioma occurring in both age groups [5,7]. It is suggested that the papillary neoplasm be considered as a separate entity for which the term suprasellar papillary squamous epithelioma is descriptive.

REFERENCES

1. Giangaspero F, Osborne D, Burger PC, Stein RB. Suprasellar papillary squamous epithelioma. *The American Journal of Surgical Pathology* 1984;8:57-64.
2. Rubinstein LJ. Tumors of the Central nervous system. Armed Forces Institute of Pathology, Washington DC 1972.
3. Russell DS, Rubinstein LJ. Pathology of Tumours of the nerv-

- ous system, 4th ed, Baltimore, Williams and Wilkins, 1977;32-8.
4. Burger PC, Scheithaver BW. Tumors of the central nervous system. Baltimore, Armed Forces Institute of Pathology. Third series fascicle 1994;10:52-5.
 5. Crotty T, Scheithaver BW, Young WF, Davis D, Miller G, Burger PC. Papillary craniopharyngiomas. A morphological and clinical study of 46 cases. *Endocrine Pathol* 1992;3 (Supp 1): 56-8.
 6. Burger PC, Vogel FS. Surgical pathology of the nervous system and its covering, 2nd ed, New York, John Wiley and Sons 1982:511-25.
 7. Kahn EA, Goseh HH, Seeger JF, Hicks SP. Forty-five years experience with the craniopharyngiomas. *Surg Neurol* 1973;1:5-12.
 8. Banna M. Craniopharyngioma in adults. *Surg Neurol* 1973;1:202-4.
 9. Crane TB, Yee RD, Hopler RS and Hallinan JM. Clinical manifestations and radiologic findings in craniopharyngiomas in adults. *Am J Ophthalmol* 1982;94:220-8.
 10. Ivkov M, Ribaric I, Slavik E. Surgical treatment of craniopharyngiomas in adults. *Acta Neuro Surg* 1979;28:352-6.
 11. Russell RWR, Pennybacker JB. Craniopharyngioma in the elderly. *J Neurol Neurosurg Psychiatry* 1961;24:1-13.
 12. Braun IF, Pintio RS, Epstein F. Cystic Craniopharyngioma. *Am J Neuro Radiol* 1982;3:139-41.
 13. Cabezudo JM, Vaquere J. Computed Tomography with Craniopharyngioma: A review. *Surg Neurol* 1981;15:422-7.
 14. Flitz CR, Wortzman G. Computed tomography in Craniopharyngiomas. *Radiology* 1978;127:687-91.
 15. Petito CK. Craniopharyngioma A clinical and pathological review. *Cancer* 1976;37:1944-52.