Extensive Ossification in a Craniopharyngioma

SAMRUAY SHUANGSHOTI, M.D.*,
SHANOP SHUANGSHOTI Jr, M.D.*

Abstract

A rare extensive ossification occurred in a suprasellar craniopharyngioma of a man who died at 21 years of age. The tumor produced headache, retarded physical and mental development, visual disturbances, and increased intracranial pressure. The neoplasm recurred after surgical and roentgenological treatments. Differentiation of multipotential mesenchymal cells or mesenchymal type cells within the tumor has been suggested as the mode of occurrence of bone in this craniopharyngioma.

Key word : Craniopharyngioma, Ossification, Brain Tumor

SHUANGSHOTI S, SHUANGSHOTI S, Jr
J Med Assoc Thai 2000; 83: 1424-1429

We do not know how many cases of ossification affecting the craniopharyngioma have been reported. However, we have found in our review of the literature a small number of authors who stated briefly about ossification in craniopharyngiomas in their experiences[1-5]. For example, Petito *et al* asserted that 6 of 192 histopathologically verified craniopharyhgiomas of their series were ossified, a prevalence of 3 per cent[6]. These authors, however, gave no details of their patients[1-6]. Herein, we fully present our experience of extensive ossification occurring in a craniopharyngioma.

PATIENT

A 21-year-old man had had intermittent headache and blurred vision since he was 6 years old. He, moreover, was retarded in physical and mental developments in comparison to children of the same age. The headache was aggravated by learning, and he stopped attending school at 10 years of age.

He was first hospitalized at 18 years of age in order to investigate the headache and intermittent coma. He weighed 32 Kg. His vision was only finger count bilaterally. Ocular movement was full bilate-

* Department of Pathology, Faculty of Medicine, Chulalongkorn University, Bangkok 10330, Thailand.

rally. Pupils, 5 mm across, were reactive to light. The pale optic discs showed papilledema. There was bilateral extensor plantar responses.

A computerized tomogram brain scan exhibited an inhomogeneous hypodensity lesion with cystic and solid components and calcification in the suprasellar region. Five days after hospital admission, the lesion was removed as much as possible via bifrontal craniotomy and proved to be a craniopharyngioma. The patient further received radiotherapy of 4,900 rads within 38 days. Additionally treatment for panhypopituitarism was given.

Three years later, he was rehospitalized because of headache, fever and cough, stuporose, and bilateral papilledema. His blood pressure was low and fluctuated. He died 2 hours and 45 minutes after hospital admission.

Pathological examination

The specimen removed surgically when the patient was 18 years old consisted of multiple fragments of gray and firm tissue, 0.5 to 1.5 cm in greatest dimension. Some pieces were focally calcified.

The present general postmortem findings included widespread thrombi in the lungs with a recent hemorrhagic infarct (8 x 7 x 5 cm) of the right lower lobe, acute congestion and edema of lungs, advanced fatty liver, and acute tubular necrosis of the kidneys.

The 1200-g brain had a nodular and hard mass, 3 x 3 x 2 cm, at the hypothalamic region; it protruded into the third ventricle. There was moderate hydrocephalus of the lateral ventricles (Fig. 1A). After decalcification in 5 per cent nitric acid for 2 weeks, the lump was still sectioned with difficulty, and found to be gritty and solid. The coarsely granular and chalky cut surfaces were studded with yellow and rust-colored foci (Fig. 1B). The rest of the brain was severely congested and edematous. There was a prominent pressure groove of the cerebellar tonsils.

Microscopically, the surgical and postmortem materials were strikingly similar, and will be described together. The mass was composed of epithelial and stromal components. The epithelial element consisted of stratified keratinized squamous cells with underlying cuboidal cells at the base to resemble epidermis of the skin. The squamous epithelial cells often formed keratinized whorls (or pearls) (Fig. 2A, arrowheads). Moreover, squamous epithelial cells were frequently arranged in balls (Fig. 2B). The intercellular bridges were occasionally noted (Fig. 2B, arrowhead).

Cuboidal epithelial cells were also detected. They often formed a row around aggregates of stellate-shaped cells which showed an occasional central loose (cystic) zone (Fig. 2C). Calcification was observed intraepithelially (Fig. 3A). Some calcific

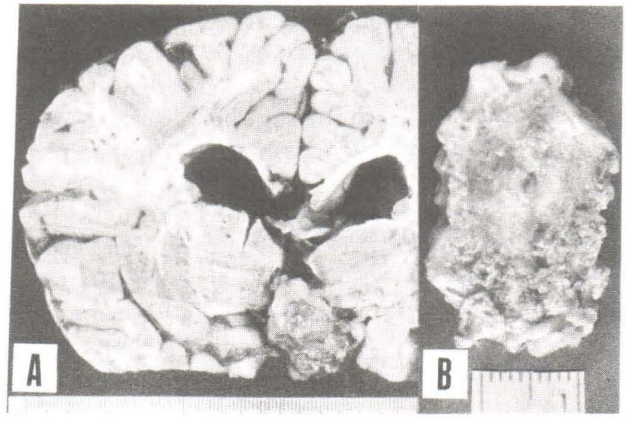

Fig. 1. Gross features of recurrent craniopharyngioma. The scale is in centimeter.
A. Nodular tumor at the hypothalamic region partially protrudes into the third ventricle. Note moderate hydrocephalus of the lateral ventricles.
B. The cut surface of the lesion is rough and solid.

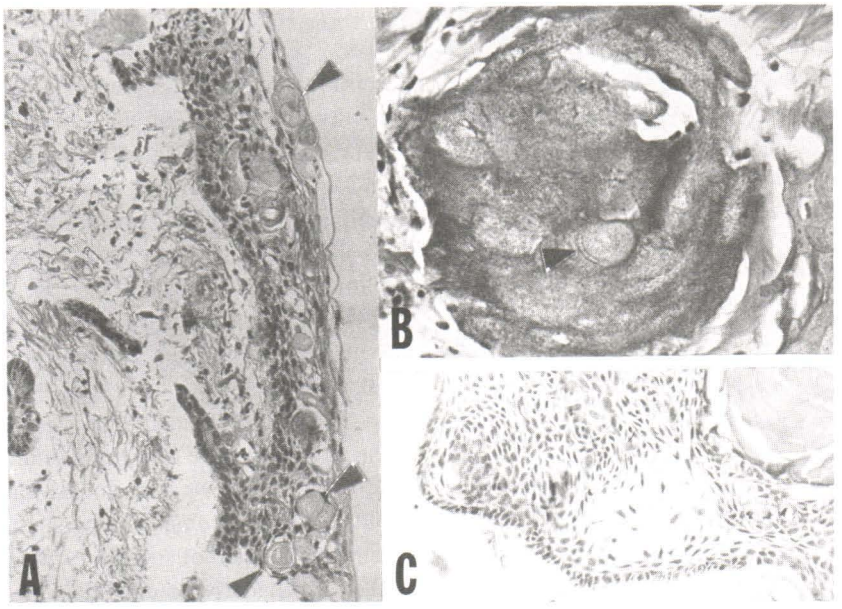

Fig. 2. Microscopic features of the craniopharyngioma.

A. Stratified squamous epithelium with basal cuboidal cells and subepithelial stroma with sparse chronic inflammatory infiltrates are exhibited. Masses of "wet" keratin are scattered intra-epithelially; some are indicated by the arrowheads (H & E, x 100).

B. Concentric nest of keratinized squamous epithelial cells is demonstrated The arrowhead points toward a prickle cell with intercellular bridges (H & E, x 400).

C. Layer of cuboidal epithelial cells covers aggregate of stellate-shaped cells. Note a mass of "wet" keratin (H & E, x 100).

foci had larmellar appearance to resemble calcified epithelial whorls (Fig. 3B).

The stroma of the tumor consisted of fine connective tissue fibers (Fig. 2A). However, hyalinized and coarse connective tissue fibers mixed with fibroblasts as well as fibrotic zones were also perceived as were calcific and ossified foci. In the latter, both incompletely-formed and well-formed bones were present (Fig. 4). Moreover, foci of xanthogranulomatous reaction were detected (Fig. 5).

The pathological diagnosis, then, was a craniopharyngioma showing severe ossification and calcification.

DISCUSSION

The craniopharyngioma demonstrates the epithelial part to be composed of keratinized squamous cells and cuboidal or columnar cells without glandular structures and hair follicle[1-7]. Nodules of "wet" keratin are also common and significant in histopathologic diagnosis of this tumor which were also detected in our case[7]. Moreover, the young age of our patient, visual difficulties, retardation of physical and mental development, and suprasellar location of the lesion are also characteristic clinical features of the craniopharyngioma[1]. Hence, the diagnosis of craniopharyngioma is beyond doubt in this instance.

Banna found in reviewing 10 series of craniopharyngiomas that the range of calcification was within 53 to 100 per cent; the average was 68 per cent[2]. By contrast, Petito *et al* encountered only 6 of 192 histopathologically verified craniopharyngiomas to be ossified, a prevalence of 3 per cent[6]. We are unable to explain as to why ossification is so rare compared to the commonness of calcification in craniopharyngiomas.

The mode of occurrence of bone in craniopharyngioma as well as in other nonosseous tumors remains speculative. However, metaplasia and dif-

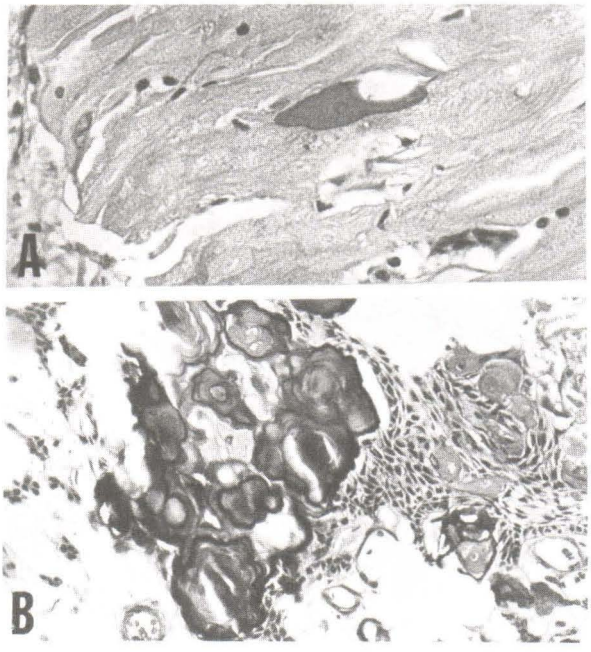

Fig. 3. Epithelium and calcification.
A. A calcified epithelial cell lies within lamella of cornified epithelial cells (H & E, x 400).
B. Calcified concentric corpuscles are demonstrated (H & E, x 100).

ferentiation should play a role. Willis used the term metaplasia to mean the transformation of an adult or fully differentiated tissue of one kind into a differentiated tissue of another kind in response to abnormal circumstances[8]. According to Parsons, metaplasia should be applied to the change that occurs in nonneoplastic condition[9]. Thus, metaplasia is the most suitable term to be utilized when osseous tissue is encountered in relation to extensive fibrosis of the stroma of the choroid plexus and lung[10,11]. Nevertheless, we have also observed the term metaplasia that has been employed in association with the formation of bone in some neoplasms such as in meningioma[12]. We, however, feel that the term differentiation would be more applicable in such a neoplastic condition. Based on our previous studies on the development of bones in a parapharyngeal meningioma, a subcutaneous neurilemmoma, and a congenital osteolipoma, we suggest that the pluripotential mesenchymal cells or mesenchymal type cells *viz* the stromal elements of the current craniopharyngioma, have differentiated into bone within this neoplastic lesion[13-15].

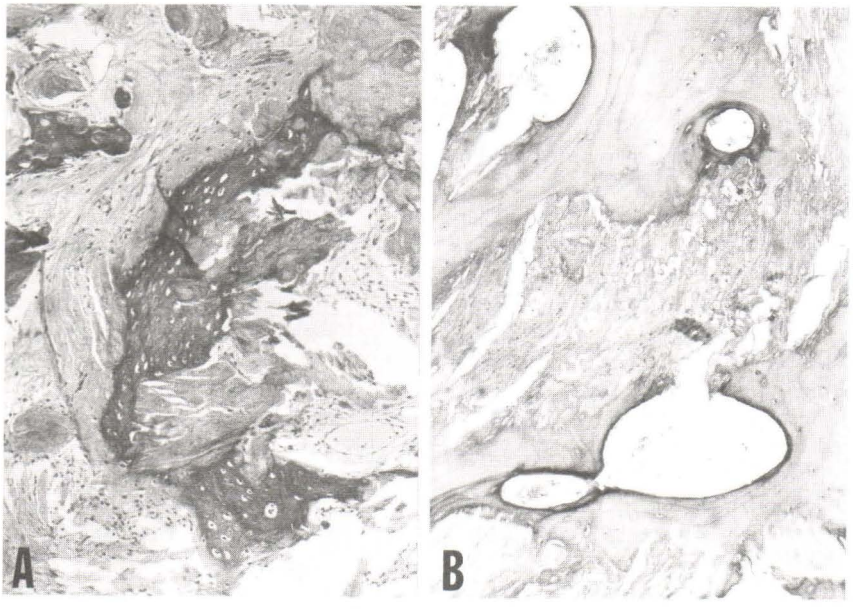

Fig. 4. Ossification in craniopharyngioma.
A. Irregular islands of incompletely-formed bone are surrounded by fibrotic stroma of the tumor (H & E x 100).
B. Fibrotic interstitium of the neoplasms is bordered by well-formed bone (H & E, x 100).

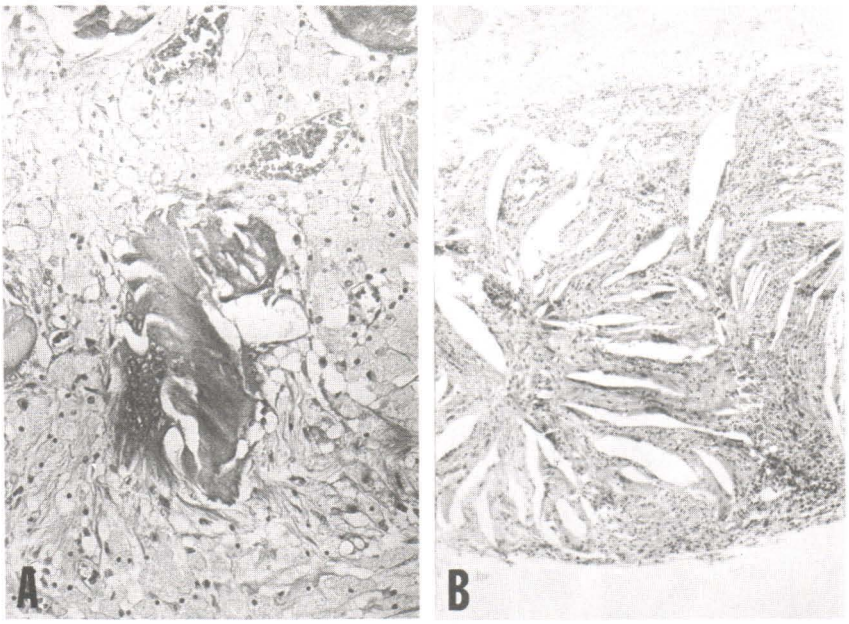

Fig. 5. Xanthogranulomatous reaction in craniopharyngioma.

A. Xanthogranulomatous reaction consisting of foreign bodies surrounded by multinucleated foreign body giant cells and foamy (xanthoma) cells is shown in the matrix of the craniopharyngioma (H & E, x 100).

B. Numerous clefts of cholesterol crystals, lymphocytes, and foreign body giant cells are mingled in the fibrotic stroma of the craniopharyngioma (H & E x 50).

(Received for publication on October 29, 1998)

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กระดูกเกิดอย่างกว้างขวางในเครนิโอฟาริงจิโอมา

สำรวย ช่วงโชติ, พ.ด., ชนพ ช่วงโชติ, พ.บ.**

ผู้ป่วยชายไทยเสียชีวิตเมื่ออายุ 21 ปี ด้วยเครนิโอฟาริงจิโอมา ที่ทำให้เขามีร่างกายและสติปัญญาเจริญไม่สมอายุ สายตาไม่ปกติ และความดันเพิ่มในโพรงกะโหลกศีรษะ เนื้องอกมีหย่อมกระดูกเกิดมากภายในก้อน ซึ่งพบปรากฏการณ์เช่นนี้น้อยมากในเครนิโอฟาริงจิโอมา ได้ชี้แนะว่ากระดูกนี้อาจเกิดจากการแตกตัวของเมเซนคัยมัลเซลล์หรือของเซลล์ประเภทเมเซนคัยม์ ภายในก้อนเนื้องอกไปเป็นกระดูก

คำสำคัญ : เครนิโอฟาริงจิโอมา, การเกิดกระดูก, เนื้องอกสมอง

สำรวย ช่วงโชติ, ชนพ ช่วงโชติ
จดหมายเหตุทางแพทย์ ฯ 2543; 83: 1424–1429

* ภาควิชาพยาธิวิทยา, คณะแพทยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย, กรุงเทพฯ 10330