Symptomatic Subependymoma: Cases Presenting with Different Clinicopathological Features

Semptomatik Subependimoma: Değişik Klinikopatolojik Özelliklere Sahip Olgular

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Abstract: We report three examples of subependymoma presented as different clinicopathological aspects. One has an ependymomatous component, the second one is associated with craniopharyngioma. In third case, there is massive hemorrhage into the large subependymoma. The clinical and histopathological findings and the prognosis of these tumors are discussed.
Key words: Craniopharyngioma, ependymoma, subependymoma

Özet: Farklı klinikopatolojik özelliklere sahip üç subependimom olgusu sunulmaktadır. Olgulardan biri ependimom alanlarına sahiptir, diğer olgu kraniofaringiom ile birliktelik göstermektedir. Üçüncü olguda ise büyük bir subependimom içinde yoğun kanama izlenmiştir. Olguların klinik ve histopatolojik özellikleri ile prognozları tartışılmıştır.
Anahtar Sözcükler: Ependimoma, kraniofaringioma, subependimoma

INTRODUCTION

The subependymoma was recognized as a distinct histopathological entity by Scheinker (11). They generally project into the ventricular lumen rather into the brain paranchyma. Many are asymptomatic, found incidentally at autopsy. More recently, the clinical series and case reports of symptomatic examples appear in the literature (1,5,6,7,13). Symptoms are most often produced by large tumors, particularly those arising from the septum pellucidum, the floor of the fourth ventricle, and the lateral ventricular walls (1,9,10). About a fourth of the symptomatic subependymomas have mixed tumor cell populations, and consist of a mixture of ependymoma and subependymoma. The prognosis in such instances is much less favorable (9). Although the subependymoma is generally considered to be a neoplasm, the possibility that these tumors may in some instances be reactive has been suggested by Russel and Rubinstein (10).

In this report, we describe three cases of subependymoma presented as different clinicopathological aspects.

REPORT OF CASES

Case 1: A 3 year old girl presented with ataxia in the past 4 months. Neurologic examination revealed a truncal ataxia that kept her from walking. Fundoscopic examination was unremarkable. Computed tomographic (CT) scans showed a tumor filling the fourth ventricle and containing the areas of calcification, and an obstructive hydrocephalus. The tumor was nonhomogeneously enhanced with contrast injection (Figure1,a). The tumor extending to the right cerebello.pontine angle was removed totally (Figure 1,b). Laryngeal spasm due to intubation and dysphagia developed in the early postoperative period. Dysphagia improved 3 weeks after surgery and she was discharged from the hospital with truncal ataxia. Pathological diagnosis was subependymoma and ependymoma. Postoperative radiation therapy was planned, but the parents of the patient refused any adjunctive therapy.

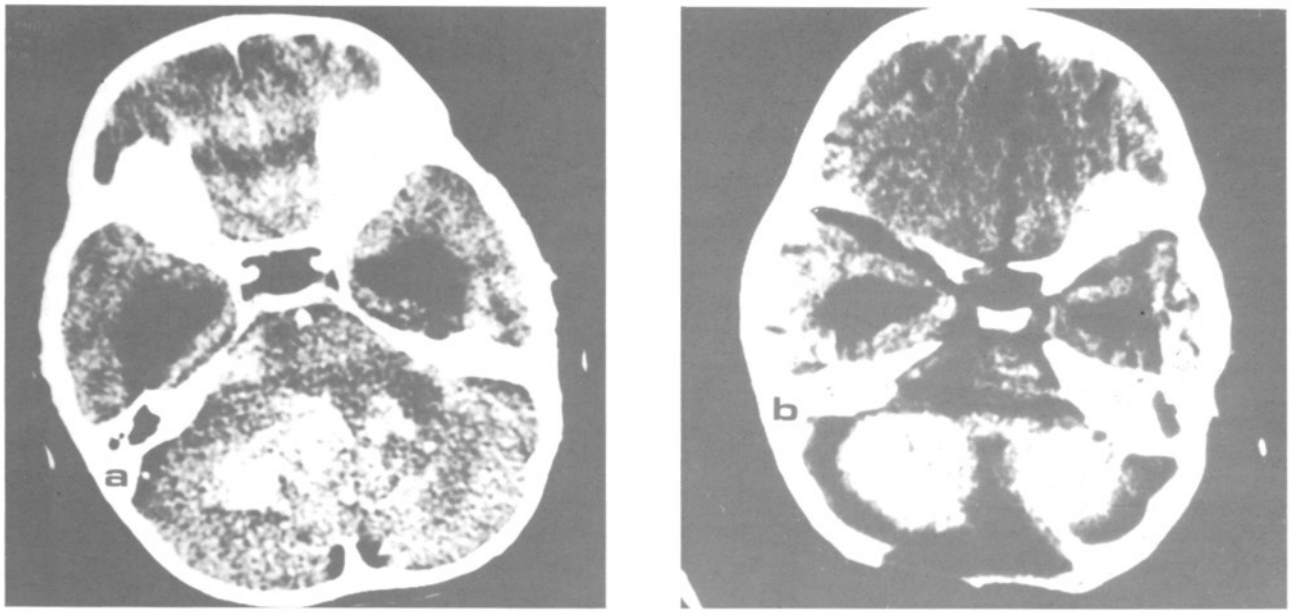

Figure 1: (a) CT scan showing nonhomogeneously enhanced tumor filling the fourth ventricle, (b) note the total tumor removal on postoperative CT scan.

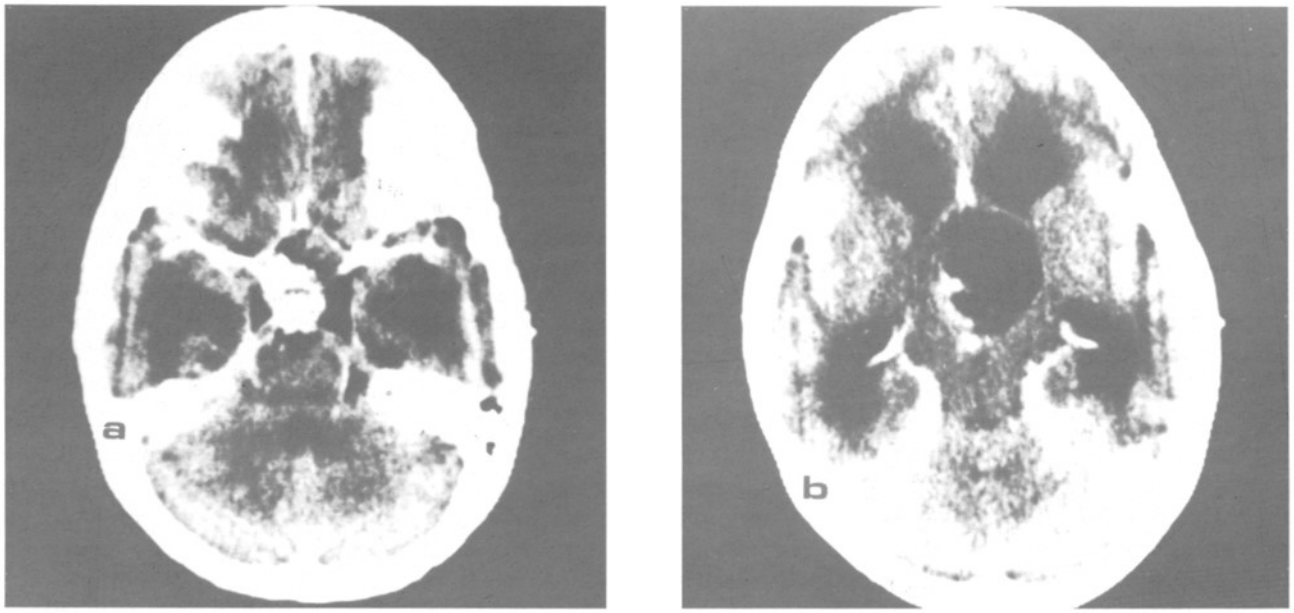

Figure 2: CT scans showing supra sellar cystic tumor, containing calcification (a) and extending into the third ventricle (b).

The patient was neurologically intact 18 months after surgery.

Case 2: A 12 year old girl presented with headache and vomiting in past 2 months. The patient also complained that she had occasionally been having headache since the age of 11 years. She was obese, however neurologically intact except for the papilledema. Computed tomographic (CT) scans showed a suprasellar cystic lesion with a component of calcification (Figure 2). Optic chiasm was seen to be swollen at surgery and biopsied. The tumor was considered as optic glioma and the radiation therapy was planned. Headache and vomiting recurred 4 months after surgery in spite of radiation therapy. Magnetic resonance imaging (MRI) scans revealed no change in tumor volume and findings was consisted with craniopharyngioma (Figure 3). A second surgery was performed 9 months after the first surgery. The cystic tumor, localized beneath the

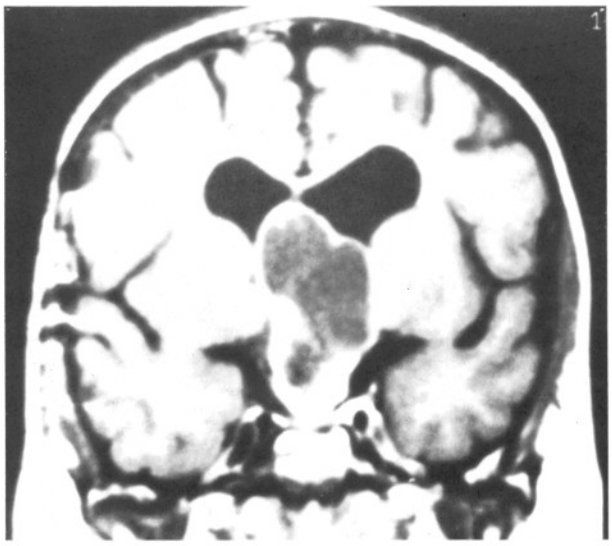

Figure 3: Sagittal view of MRI scan showing suprasellar cystic tumor.

optic chiasm and extending to the third ventricle was removed subtotally (Figure 4). Pathological diagnosis was craniopharyngioma and subependymoma. The patient was stuporous and had diabetes insipidus postoperatively. She was transferred to the pediatric intensive care unit for treatment of hyponatremia. Left subdural collection was evacuated via two burr holes. Meanwhile trombophlebitis developed in legs and the patient was heparinized. She was always stuporous. She deteriorated and the CT scan revealed left sided subdural hematoma due to the hemorrhagic diathesis. She died 7 weeks after second surgery despite the evacuation of subdural hematoma.

Case 3: A 76 year old woman presented with drop attacks, dizziness and left sided weakness that had arisen in the past weeks. Physical examination was unremarkable except for thoracic kyphosis and arthritic deformities in knees. The patient was alert and oriented. Neurologic examination revealed right abducens nerve paralysis and left hemiparesis. CT scan disclosed a round, enhanced mass in the right lateral ventricle (Figure 5). A right parietal craniotomy was performed and the tumor was excised in toto via transcortical approach. The pathological diagnosis was subependymoma and hemorrhage. The postoperative course was uneventful except for the confusional state. The patient is neurologically intact 6 months after surgery.

Pathological Findings

In case 1, the tumor was composed of nests of small glial cells separated by broad bands of

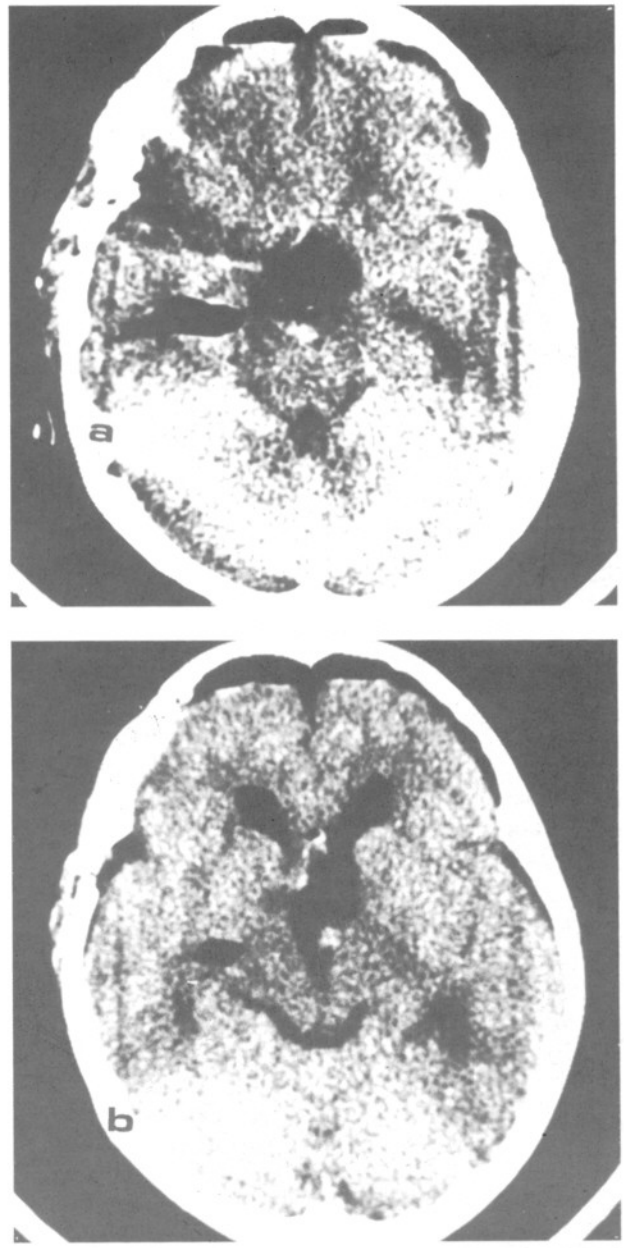

Figure 4 a,b: Postoperative CT scans showing almost total tumor removal.

neuroglial fibers, which imparted a low power lobularity (Figure 6). The cells contained round to oval nuclei in shape, usually with a delicate punctuate chromatin network. Some cells had vesicular and more irregular larger nuclei, more typical of astrocytes. Ependymatous area showed prominent ependymal rosettes and canals, abundant vascularity and endothelial proliferation (Figure 7). Mitoses and necroses were not seen. Increased cellularity, poorly formed perivascular pseudorosettes and endothelial proliferation were seen in some areas surrounding the ependymatous

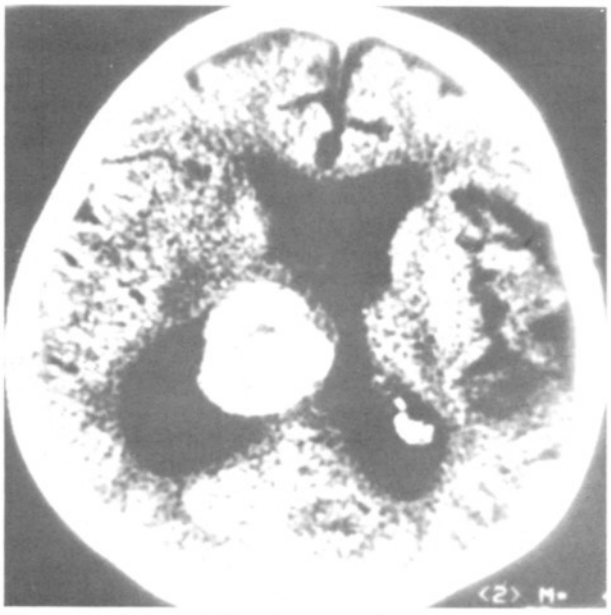

Figure 5: CT scan showing an enhanced tumor in the right lateral ventricle.

Figure 7: Case 1. Ependymal rosettes and tubules in ependymatous area. (H&E, X480)

portion of tumor. Rosenthal fiber formation was observed in the subependymoma component. Calcification and cystic spaces were often found. The fibers between the cells were positive for glial fibrillary acidic protein.

In case 2, the tumor appeared similar to case 1 except for the ependymatous area . In addition, there were the nests of craniopharyngioma (Fig. 8). These nests were composed of "ghost cells" of craniophryngioma. The prominent craniophryngioma nests were observed in new sections. The cellularity increased around these nests. The lesion was different from reactive astrocytosis or astrocytoma because of lobularity, cellular features and distribution.

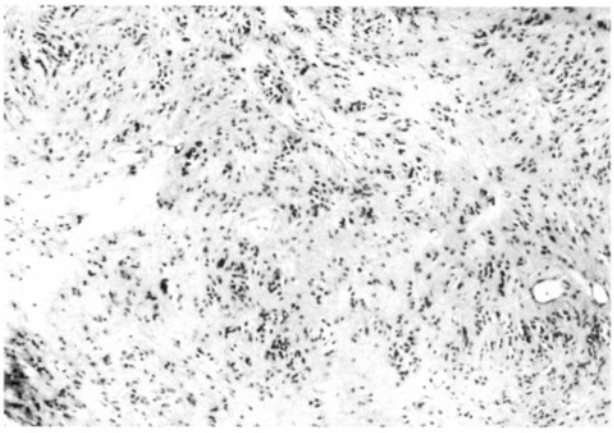

Figure 6: Case 1. Microscopic appearance of the tumor by nests of isomorphic cells with round and oval regular nuclei, typical lobular pattern. (H&E, X120)

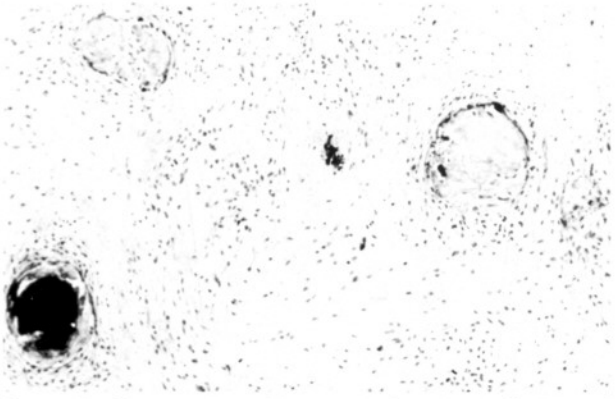

Figure 8: Case 2. Note "ghost cells" of craniopharyngioma in subependymoma. (H&E,X120)

In the last case, there was massive hemorrhaging into the large subependymoma. The intact tumor area was confined to the ventricle wall. The vascular sclerosis, vascular thrombosis, calcification and reactive gliosis were observed in the other areas.

DISCUSSION

Present cases call attention to different clinicopathological aspects. In the first case, the neoplastic proliferation is apparent and there is an ependymatous component. We suspect that the nature of the case is associated with craniopharyngioma, whether reactive or neoplastic. The possibility that subependymoma may in some instances be reactive to some other process (10). Scheithauer (12) reported the two cases with

granular ependymitis and leptomeningeal melanomatosis. Russell and Rubinstein (10) considered the reactive nature of a case that is associated with craniopharyngioma. It was known that reactive astrogliosis are observed in craniopharyngioma. In our second case, the lesion is different from reactive astrocytosis or astrocytoma because of lobularity, cellular distribution and nuclear features. On the other hand, there is typical feature of subependymoma in all fields. This feature is very coarse and masks the nests of craniopharyngioma. This course is considered to be a tumoral process.

The massive hemorrhage in subependymoma was reported (12,14). Third case indicated that the subependymoma might also be considered for intraventricular hemorrhage mass.

Subependymomas have a distinctive microscopic appearance, but their origin has been a matter of some controversy (2,8,10). Some authors suggest that it arises from subependymal astrocytes (3). The others consider it a variant of ependymoma (4,10). Several authors reported that subependymoma is composed of cells having the cytoplasmic features of ependyma cells, astrocytes and transitional cells at the light microscopic and ultrastructural level (2,8). These studies and light microscopic features of our cases that have a mixed cell population expressed this opinion. These mixed tumors possibly take origin from ependymoglial cells normally present in adult subependymal layer (2,8).

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