



Pineal Epidermoid Tumors: Report of Five Cases

Pineal Epidermoid Tümörler: 5 Olgu Sunumu

Cem DINC, Ahmet Celal IPLIKCIOGLU, Erdinc OZEK

Ministry of Health Okmeydani Education&Research Hospital, Department of Neurosurgery, Istanbul, Turkey

Corresponding Author: Cem DINC / E-mail: drcemdinc@yahoo.com

ABSTRACT

AIM: The aim of this study was to retrospectively analyse the clinical, radiological features and surgical outcome of pineal epidermoid tumors treated at a single neurosurgical department.

MATERIAL and METHODS: We performed surgery on five patients with pineal region epidermoid tumors at a single neurosurgical department between the years 1998 and 2006. Headache, diplopia and ataxia were the most common presenting findings. Parinaud's syndrome was found in three patients. Hydrocephalus was demonstrated radiologically in two patients.

RESULTS: Two patients were operated on with the occipital-transtentorial approach, two were operated on with the infratentorial-supracerebellar approach and one was operated on with van Wagenen's approach. Recurrence of tumor was observed in one patient. One patient died at the first postoperative month due to ventriculitis.

CONCLUSION: Total removal of epidermoid tumors may provide good clinical recovery and may reduce the possibility of tumor recurrence and shunt placement.

KEYWORDS: Epidermoid tumor, Pineal region, Surgery

ÖZ

AMAÇ: Bu çalışmanın amacı, tek bir merkezde tedavi edilen pineal epidermoid tümörlerin, retrospektif olarak klinik, radyolojik özellikleri ve cerrahi sonuçlarını analiz etmektir.

YÖNTEM ve GEREÇLER: Pineal bölgede epidermoid tümörü olan beş hastaya, 1998 ve 2006 yılları arasında tek bir beyin cerrahisi bölümünde cerrahi girişim uygulandı. Baş ağrısı, diplopi ve ataksi en sık görülen bulgular idi. Parinaud sendromu, üç hastada tespit edildi. Radyolojik olarak iki hastada hidrosefali gösterildi.

BULGULAR: İki hasta oksipital-transtentoryal yaklaşımla, iki hasta infratentoryal-supraserebellar yaklaşımla ve bir hasta van Wagenen yaklaşımı ile ameliyat edildi. Tümör nüksü bir hastada görüldü. Bir hasta ventrikülit nedeniyle postoperatif birinci ayda kaybedildi.

SONUÇ: Epidermoid tümörlerin total çıkarılması klinik iyileşme sağlayabilir ve nüks ve şant yerleştirilmesi olasılığını azaltabilir.

ANAHTAR SÖZCÜKLER: Epidermoid tümör, Pineal bölge, Cerrahi

INTRODUCTION

Intracranial epidermoid tumors comprise about 0.2-1% of all intracranial tumors (23). These congenital lesions have a tendency to enlarge into cisterns and generally show lateral localization (4). However, pineal region localization of an epidermoid tumor is extremely rare (12,15,17,25). When the literature is reviewed, most of the cases presented are single case reports and yet a few studies deal with large series of intracranial epidermoid tumors (6,13). In our report, we present 5 cases of pineal epidermoid tumors treated surgically at our department in an eight-year period.

CLINICAL MATERIAL and METHODS

At our department, 5 cases with pineal epidermoids were treated surgically. The age of the patients at the time of the diagnosis ranged from 12 to 38 (mean 31) years. Three cases were male and two were female. Patient characteristics are summarized in the Table I. The duration of symptoms prior to diagnosis ranged from 4 to 18 months. Headache, visual

disturbance and papilledema were the most common symptoms and signs. Parinaud's syndrome, a typical sign of pineal region tumors, was found in three patients. Hydrocephalus was demonstrated radiologically in two cases. One patient underwent venticuloperitoneal shunt placement while he was first admitted emergently with a decreased level of consciousness. Computed tomography (CT) and magnetic resonance imaging (MRI) were performed on all cases in the preoperative and postoperative periods. We performed surgical treatment in all cases using three different approaches.

RESULTS

Four cases were admitted to our neurosurgery department with a complaint of headache and/or visual disturbances. One patient was admitted to the emergency department with decreased level of consciousness and CT scanning revealed a mass lesion in the quadrigeminal cistern with a density similar to the cerebrospinal fluid (CSF) and hydrocephalus. All

Table I: Characteristics in Five Patients with Pineal Epidermoid Tumors

Case no	Age/Sex	Operation	Recurrence	GOS
1	30 / M	Infratentorial - Supracerebellar	-	V
2	40 / F	van Wagenen	-	I
3	12 / M	Occipital - Transtentorial	6 years later	V
4	38 / M	Infratentorial - Supracerebellar	-	IV
5	26 / F	Occipital - Transtentorial	-	V

cases except for one revealed typical radiological findings on CT scans and MRI. In one case, we were not able to decide precisely even with a MR spectroscopy whether the lesion was an epidermoid tumor or arachnoid cyst and the final diagnosis was established by intraoperative findings and histopathological examination of the lesion.

Four patients underwent surgery in the sitting position. Despite some disadvantages, especially air-embolism and pneumocephalus, we believe that the sitting position provides

better exposure and anatomical orientation. In two cases, the supratentorial component of the tumor was significant and we therefore used the occipital-transtentorial approach. In two cases, we used the infratentorial-supracerebellar approach. The remaining patient was operated with van Wagenen's approach through the dilated right lateral ventricle in the prone position (Figure 1A-D). In one case, the tumor capsule was adherent to the venous structures and we had to leave in situ fragments. We encountered recurrence of tumor in this

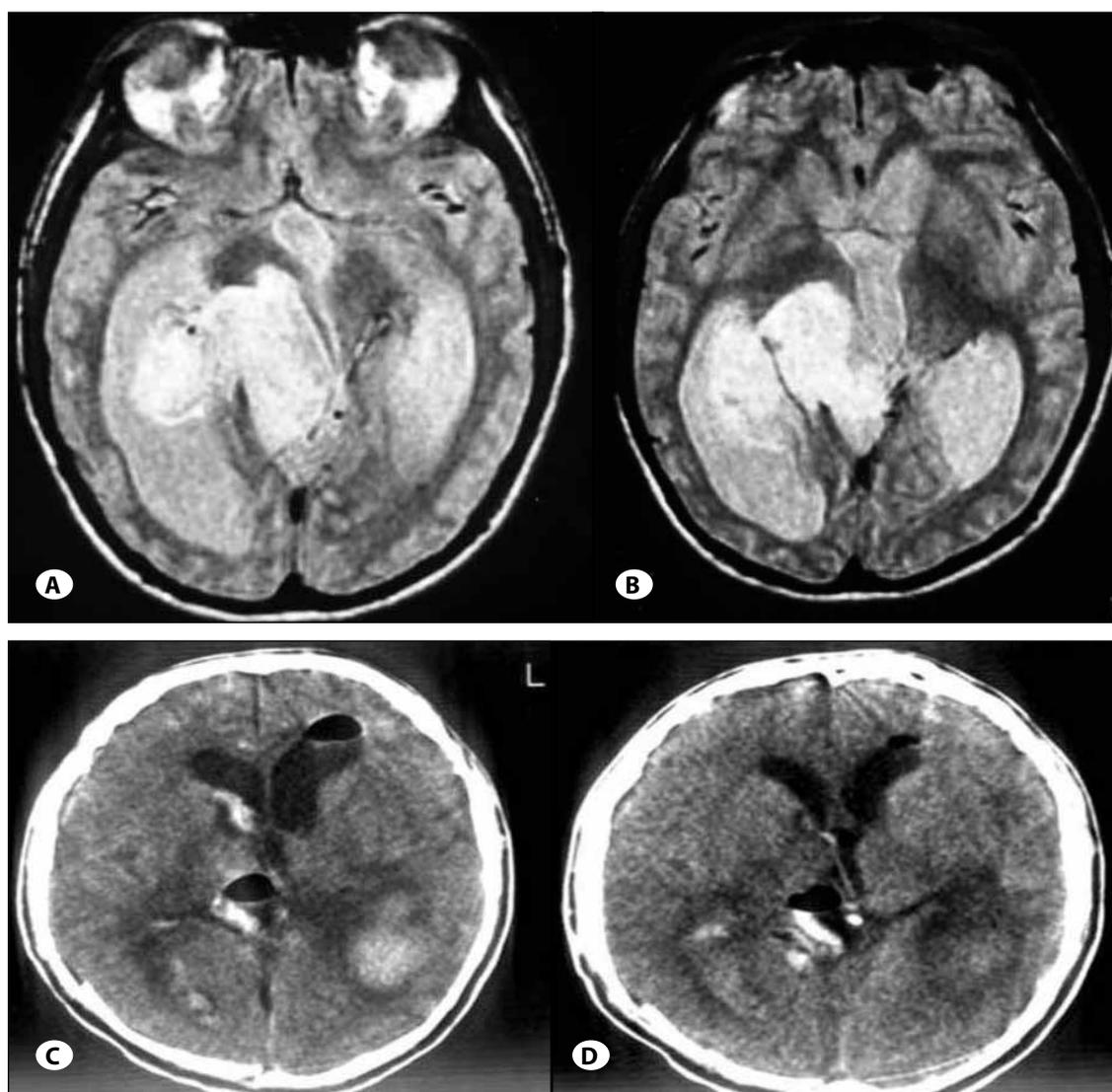


Figure 1: Case 2. Proton MRSI showing a mass in the pineal region enlarging to the right lateral ventricle and mild hydrocephalus (A,B). Postoperative control axial CT scans demonstrating total removal of tumor (C,D).

case six years after the first operation (Figure 2A-C). In this case, there was no tumor recurrence in the two years follow-up of the second operation. We inserted a ventricular external drainage system in the patient operated upon with the transventricular procedure and he died due to ventriculitis one month after the operation. The average duration of the follow-up was 5.6 years and there was no tumor recurrence in the other three patients.

DISCUSSION

Intracranial epidermoid tumors account for 0.2-1% of all intracranial tumors (23). The cerebellopontine and parasellar cisterns are the most common locations (4). However, the pineal region is an extremely rare site for epidermoid tumors to occur (11,12,15,17,25). The first case of a pineal region epidermoid tumor was reported by Cushing in 1928 (27). In 1970, Kirsch and Stears (12) reported that only 7 cases were present in the current literature of the time. Konavalov et al (13) reported a large series of pineal epidermoid tumors with six cases diagnosed between 1968 and 1999. In 1999 MacKay (14) also summarized 12 patients reported. Desai et al (6) reported a larger series of pineal epidermoids with 24 cases in 2006.

Epidermoid tumors are inclusion tumors of the central nervous system. They are thought to originate from ectodermal cells misplaced during the division of the neural and cutaneous ectoderm during the third or fourth week of intrauterine development (3,8,32). They consist of a capsule of stratified squamous epithelium containing desquamated epithelial cells, keratin and cholesterol. Unlike true neoplasms, which exhibit exponential growth kinetics, they show a linear growth pattern (4). However, in one of our cases, we observed a recurrence of the tumor six years after the first operation where the recurrent tumor reached it's original size. The patient was twelve years old when first operated. In contrast to the anticipated linear growth pattern, this tumor grew almost exponentially. We think that this unexpected growth could be related to puberty.

There is no gender dominance in previously reported series and as a rule pineal epidermoids are seen in relatively young patients (2,9,22,24,31). Headache, diplopia, vertigo and ataxia consist most common symptoms. Related to anatomic location of pineal region lesions, obstructive hydrocephalus and upward gaze palsy consist major clinical presentation (18,30). In fact, hydrocephalus with increased intracranial pressure causes a variety of symptoms and signs. Konavalov et al (13) reported that only one case presented with Parinaud's syndrome. We observed Parinaud's syndrome in three cases. Hydrocephalus on CT scanning was seen in two cases in our series.

CT scanning and MRI are the chief diagnostic tools. Angiography could demonstrate displaced deep venous structures but is not recommended routinely for pineal epidermoid tumors (15). CT scanning reveals nonenhancing, low density mass lesions without perifocal edema enlarging



Figure 2: Case 3. Sagittal T1-weighted MR imaging demonstrating a tumor in the pineal region iso-intense to the CSF (A). Early postoperative sagittal T1-weighted MR imaging showing complete resection of lesion (B). Sagittal T1-weighted MR imaging revealing tumor recurrence six years after the first operation (C).

into the quadrigeminal cistern and also hydrocephalus if it is present (7,10,21). MRI of the pineal epidermoid tumors demonstrates cauliflower-shaped masses that compress posterior part of the third ventricle or aqueduct. MRI signals of epidermoid cysts are similar to CSF; they present with low intensity in T1-weighted images and high signal intensity in T2-weighted images and do not enhance after administration of contrast agent (5,19,29). Despite these typical findings, some different radiological features have been reported. Differences in cholesterol and protein content and the presence or absence of haemorrhage have been postulated to account for the variable imaging appearances (19,28). We saw classical radiological findings in all present cases except for one.

The sitting position for neurosurgery is controversial. However, we generally prefer the sitting position for surgery of the pineal region tumors and other posterior fossa tumors. Although this approach brings some disadvantages such as air embolism and pneumocephalus, we believe that the sitting position provides better exposure and anatomical orientation. During the operation, the most important anatomical condition is the localization of the displaced deep venous system structures. Meguro et al (16) reported a case of a 74-year-old woman suffering thrombosis of the confluence of sinuses four days after the left occipital transtentorial removal of a pineal region epidermoid cyst. That is the reason why we have preferred different surgical approaches: in two cases, we performed the occipital-transtentorial approach where the tumor had a significant supratentorial component. In two cases, we performed the infratentorial-supracerebellar approach. In the remaining case, we performed van Wagenen's approach.

One case was re-admitted with recurrence of the tumor six years after the first surgery. The patient was initially operated via right occipital-transtentorial approach. In the second operation, we attempted the same approach to reach the tumor but dural adhesions did not allow us even to perform craniotomy and the operation was continued with the left occipital-transtentorial approach where the tumor was resected totally.

Although malignant degeneration and dysplastic changes of intracranial epidermoid tumors after subtotal resection are well-documented, the first and second histopathological examinations were the same in our patient who had recurrence (1,20,26).

CONCLUSION

Pineal epidermoid tumors are rare, benign congenital lesions. They cause variable gradual onset of symptoms according to their anatomic localization. We believe that a total surgical resection of the tumor with the correct surgical approach may prevent the recurrence, complications and further surgical interventions such as shunt placement.

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