CASE REPORT / OLGU SUNUMU

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Three Rare Localizations of Intracranial Dermoid Tumors

İntrakraniyal Dermoid Tümörlerin Üç Nadir Yerleşim Yeri

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Abstract

Intracranial dermoid tumors are extracerebral congenital cysts. Dermoid cysts have two types as intradural and extradural. Intradural dermoid cysts are originated from the intracranial cerebrospinal fluid space. However, calvarium-originated extradural lesions increase in size with a slow growth in cyst volume, but the cause of active growth is unknown. Dermoid cysts act like slow-growing cerebral tumors. Differential diagnosis includes other cysts and cystic tumors. Their radiologic appearance looks like polycystic lesions which is associated with a wide expansion in the cerebrospinal fluid areas and cause brain translocation. In the cranial tomography and magnetic resonance imaging assessment, high-cholesterol-containing dermoids demonstrate cerebrospinal fluid characteristics. The present study based on the review of central nervous system patients with highly rare dermoid localizations such as calvarial bone, temporal lobe and lateral ventricle.
Key Words: Dermoid Tumor, Cyst, Atypic, Extradural-Intradural Lesion

Öz

İntrakraniyal dermoid tümörler ekstraserebral konjenial kistlerdir. Dermoid kistler intradural ve ekstradural olarak iki tipte incelenir. İntradural dermoid kistler intrakraniyal beyin-omurilik sıvısı boşluğundan köken alırlar. Ancak, kalvaryum kaynaklı ekstradural lezyonlar kistik hacim olarak yavaş büyürler, fakat bunun nedeni bilinmemektedir. Dermoid kistler yavaş büyüyen beyin tümörleri gibi davranırlar. Ayırıcı tanıda diğer kist ve kistik tümörler bulunur. Radyolojik olarak polikistik lezyonlara benzer, beyin translokasyonuna neden olan beyin omurilik sıvı alanlarında genişlemelerle ilişkilidir. Bilgisayar tomografi ve manyetik rezonans değerlendirmede, yüksek kolesterol içeren dermoidler serebrospinal sıvı karakteristiği gösterir. Bu çalışmada kalvarial kemik, temporal lob ve lateral ventrikül gibi dermoid lokalizasyonları için oldukça nadir olan santral sinir sistemi hastaları değerlendirilmiştir.
Anahtar Kelimeler: Dermoid Tümör, Kist, Atipik, Ekstradural-İntradural Lezyon

Introduction

Intracranial dermoid cysts are rare tumors accounting for 0.04-0.7% of intracranial tumors (1). Intracranial dermoid cysts typically occur in the midline and 67% of these cysts are located in posterior fossa (2,3). Dermoid is often present in parasellar and frontobasal regions in the midline, particularly in the intradural parts of these regions (1). The dermoid cysts may be present in several different locations, these locations were reported to be frontal lobe, frontal base, parasellar, suprasellar, middle base, Sylvian fissure, hypothalamus, temporal lobe, pineal, posterior fossa, cerebellopontine angle, fourth ventricle and clivus (5). However, it is sometimes extradural-located and there are rare cases reported up to the present. Extradural-located dermoid cysts are intradiploic in approximately 25% of the dermoid cyst cases. Supratentorial localization is quite often. However, cysts can occur in different locations. When occurs in posterior fossa, dermoid cysts may expand and push intracranial nerves or result in hydrocephalus through pressure on the fourth ventricle. The current article presents three dermoid tumor cases with different and rare localizations as accompanied with the literature.

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Case Reports

Case 1

A 38-year-old male patient presented at our clinic with headache, nausea and vomiting. Neurological examination did not reveal any deficits. Cranial magnetic resonance imaging (MRI) revealed a mass in size of 4.2*4.2*5.2 cm in right temporal lobe, with low and equal intensity in T2 sequence but without gadolinium retention in T1 sequence. There was no spread in the cerebrospinal fluid (CSF) spaces around the tumor (Figure 1a, b). Diffusion MRI demonstrated limited diffusion intensity (Figure 2). Cranial computer tomography (CT) showed bone destruction (Figure 3). Right parietotemporal craniotomy was performed and although the mass seemed intradural radiologically, it was extradural intraoperatively and originated from diploic space. The mass was totally removed. There was no postoperative additional neurological deficit. Pathology report was consistent with dermoid tumor (Figure 4).

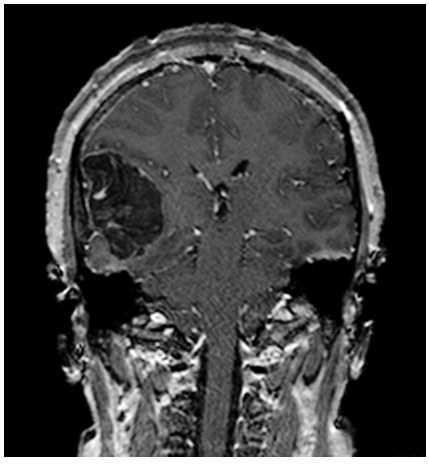

Figure 1a: T1 sequence high signal intensity was found with no gadolinium enhancement

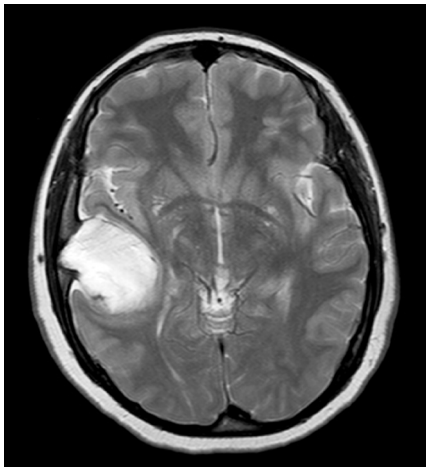

Figure 1b: Axial magnetic resonance imaging, hyperintense signal on T2-weighted sequence was seen

Case 2

A 18-year-old female patient presented at our clinic with headache, nausea and vomiting. Cranial MRI of the patient who was under follow-up at an external center for 6 years with the diagnosis of hydrocephalus revealed hydrocephalus and a lesion located in the frontal horn of the left lateral ventricle, which was hypointense in T1 sequence (Figure 5), hyperintense in T2 sequence (Figure 6) and hypointense in fluid-attenuated inversion recovery sequence. The ventricle was accessed via a neuroendoscope using a left frontal burr hole. Third ventriculostomy was performed. Meanwhile, the entire ventricle surface was observed to be covered with hair and epithelial tissue (Figure 7). No postoperative additional neurological deficit was observed.

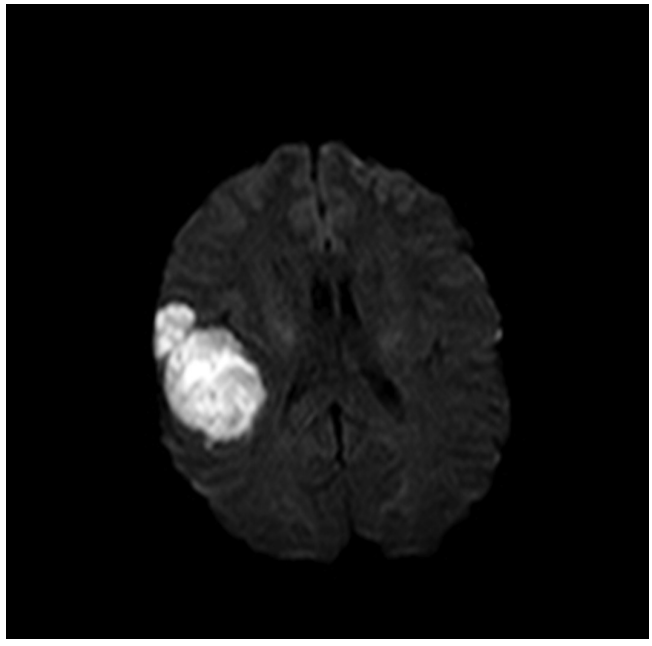

Figure 2: Diffusion restriction on diffusion magnetic resonance imaging

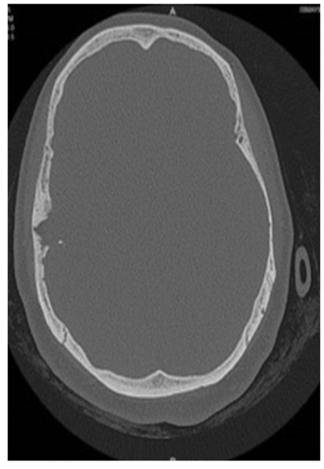

Figure 3: Temporal bone destruction on cranial tomography

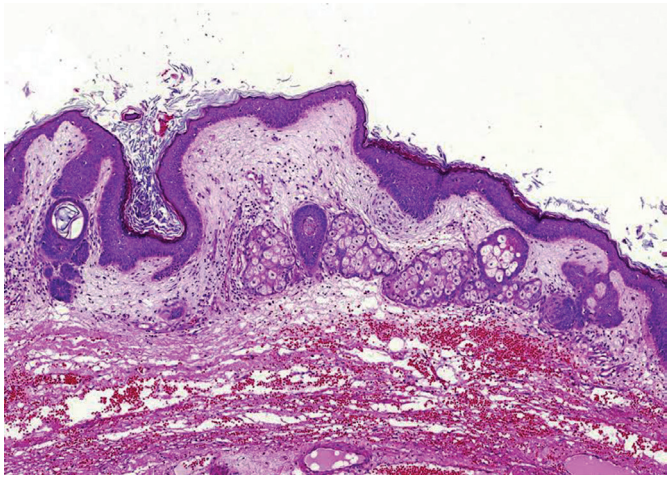

Figure 4: Pathological preparat include squamous epithelial and keratinous cells

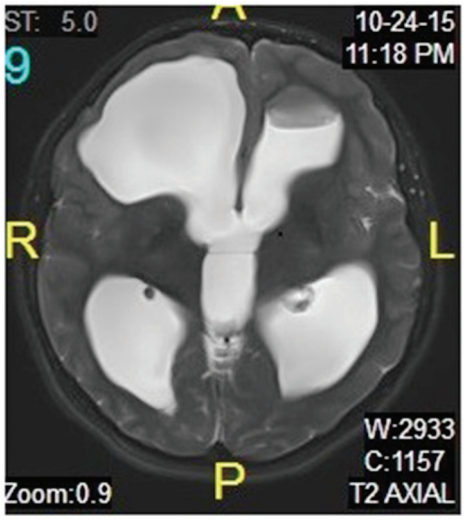

Figure 5: Lesion located in the frontal horn of the left lateral ventricle, which was hyperintense on T1 weighted magnetic resonance imaging

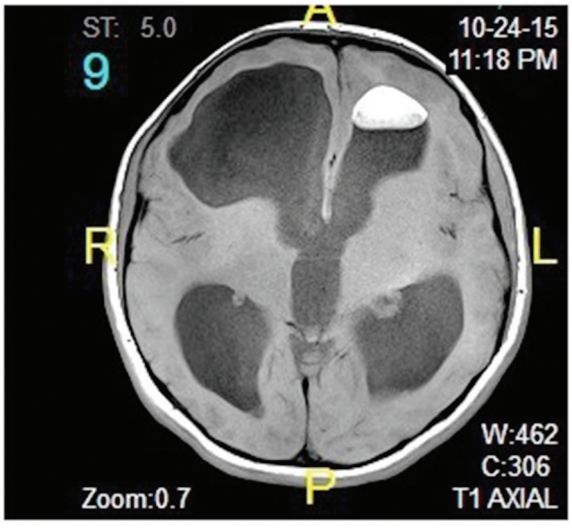

Figure 6: Lesion located in the frontal horn of the left lateral ventricle, which was hypointense on T2 weighted magnetic resonance imaging

Case 3

A 23-year-old female patient presented at our clinic with headache and seizure for 4 days. The patient did not have any neurological deficit. Cranial MRI revealed a lipid-containing mass consistent with ruptured dermoid cyst, which was located in the right temporal lobe (Figure 8) and extending over the subarachnoid space through the temporal lobe. Right frontotemporal craniotomy and total tumor excision were performed. Without any neurological deficit, the patient was discharged at postoperative day 4. Histopathological assessment reported that it was a lesion consistent with dermoid cyst characterized by keratin mass and cystic degeneration.

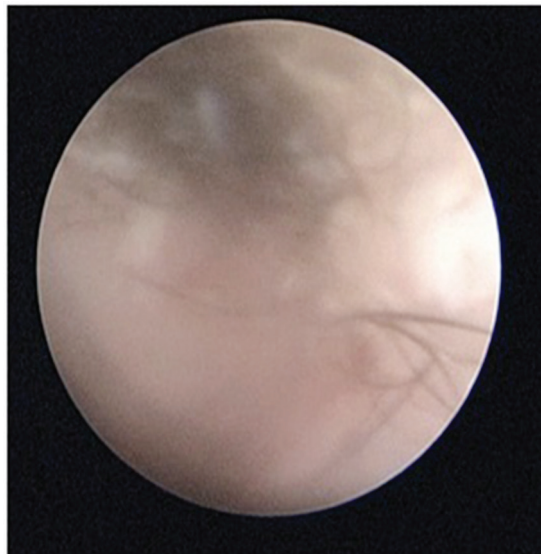

Figure 7: Hairs and epithelial tissues in the layer of lateral ventricle by endoscopic view

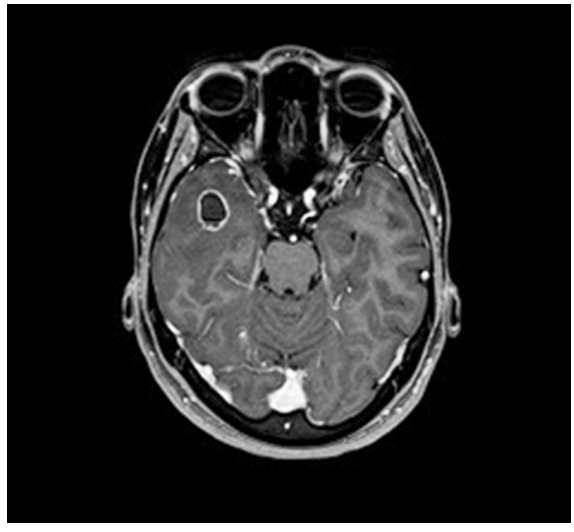

Figure 8: A mass containing lipid fragmant with peripheral gadolinium enhanced on T1 weighted magnetic resonance imaging

Discussion

Dermoid tumors are originated from embryonic ectodermal cells. The primary neurulation process during the neural tube closure involves these embryonic ectodermal cells (2,4,6). Therefore, dermoid cysts contain dermis elements such as apocrine glands, hair, hair follicles, sweat glands or sebaceous glands. In the present cases, there were keratin materials in the histopathological specimen taken during the hair and tumor resection. Cranial dermoid cysts are relatively rare (0.3-1.8% within all surgically resected craniocerebral tumors), benign and mostly intradural lesions. Extradural dermoid cysts account for 25% of the cranial dermoid cysts and affect calvarium (8). Dermoid cysts mainly occurs in the intradural areas such as frontobasal, suprasellar, parasellar regions and posterior fossa midline. Additionally, interdural dermoid cysts have been reported rarely (8). Additionally, although posterior fossa is the dominant location of dermoid cysts, a few intradural dermoid cysts originating from the temporal bone base have been reported (1). Extradural dermoid cysts occurs in posterior fossa, anterior fontanel and orbita regions in children (1,6,11). Extradural dermoid tumor, which is located in the paramedian petrous apex and leads to bone erosion, has been reported only in one case in adults (5). But in our study all cases were atypic localizated such as calvarium, lateral ventricle and temporal lobe. Lateral temporal bone extradural dermoid tumor has been rarely reported up to the present. The extradural origin despite the presence in the temporal bone in the present case is a rare condition. Martinez-Lage et al. (6) suggest that posterior fossa extradural dermoids and midline dermoid cysts may be originated from the cutaneous ectoderm that invaginates during dural invagination through the development of falx cerebri and tentorium cerebelli. There are some challenges to apply this hypothesis for the origin of an extradural dermoid cyst. For instance, there is no dural invagination in the lateral temporal region. One of these theories is that the dermoids may be originated from the multipotent embryonic cells. Second one is that it may be originated from the epithelial cells migrated from the otic vesicles with neurovascularity that is translocated or developed (4). Although there is no multipotent stem cell in the dura, the proliferation of these cells is likely to play a role in extradural dermoid cyst formation. Besides, epithelial cells may migrate during the course of neurovascular extradural region formation.

The basis of dermoid cysts is unknown because there is not enough case regarding intradural dermoid cysts in which epithelial cell migration or multipotent stem cell proliferation is observed (2,3). The epidermoid cyst includes an epithelial layer, an outer capsule and in some occasions an inner cystic fluid (9). In the first case, dermoids were hardly but totally dissected from the dura. CT imaging showed an association with parietotemporal bone destruction; however, there was no association with scalp here. Cranium is derived from the peripheral mesenchyme during brain development. Although this time period involves both mesodermal and ectodermal cells, the cartilaginous unit of the cranium is formed by the fusion of a few cartilages originating from the ectodermal cells (7). Nevertheless, the possibility for an extradural and lateral dermoid to be derived from the residues of ectodermal stem cells should be developed carefully. Specific to second case, due to high damage risk to neural structures, dissection of tumor from ventricle epandym was not performed according to our intraoperative observation. In consequence of invasiveness and adhesiveness of tumor, there was a high surgical risk and potential damage to venous structures which may result in poor patient surgical outcome.

For the third case, sudden termination of long term constant headache is not an usual complaint in neurosurgical practice. The physiopathological mechanism of the relationship between the termination of the headache and new onset of seizure simultaneously can be described by rupture of the dermoid cyst which caused a sudden decrease of intratumoral pressure and releasing of epileptogenic content into the temporal parenchyma.

Dermoids are often hypodense in CT imaging due to the lipid content (1,10). Additionally, there may be capsular calcification and there is not contrast enhancement in most dermoid tumors. Dermoids may appear with directly high-signal density in T1 MRI sequence and low or high signal density in T2 MRI sequence due to the existing lipid. Sometimes, it is difficult to specify a dermoid when it is intradural or extradural. In extradural dermoids, skeletal abnormalities such as bone erosion may present as a characteristic sign (10). Besides, expansion of the fluid space around the mass cannot be detected on in MRI. In patients with bone erosion and fluid space expansion, extradural dermoid cysts may be considered in the differential diagnosis. In our first case bone destruction was revealed around tumour tissue on the CT. Dermoid cysts on the CT scans, may have mixed densities, and uncommonly enhance following contrast administration. Disseminated fat droplets and intracystic fat appears hypodense. However calcification in the wall is hyperdense. Fat-fluid level and hydrocephalus may be present after rupture of dermoids into the ventricular system. On MRI, usually dermoid cysts are variable on T2-weighted sequences and hyperintense on T1-weighted sequences. But the presence of cholesterol may usually appear hypointense on T2-weighted sequence (12). Conventional MRI is not so useful in distinguishing other cystic pathologies of brain such as arachnoid cysts from dermoids cysts. On diffusion-weighted imaging (DWI), the dermoid tumors are hyperintense to parenchyma of brain. However dermoid

tumors are similarly on apparent diffusion coefficient (ADC) to that of CSF and parenchyma. Arachnoid cysts have an opposite pattern (elevated ADC, but low DWI) (13). Despite the imaging appearance of dermoids is typical, some other intracranial lesions such as craniopharyngiomas, lipomas, teratomas, and sometimes arachnoid cysts should be regarded in the differential diagnosis. An analysis of all available MRI and CT images usually allows the radiologist for a right preoperative diagnosis. The present article reports three cases with highly rare dermoid localizations such as calvarial bone, temporal lobe and lateral ventricle. Dermoid cysts are generally intracranial midline-located and intradural lesions; however, they should be kept in mind in the differential diagnosis in rare cases. Treatment strategy should be created according to intraoperative observations. While in some cases tumor allows total removal, in the others it may not be possible, in spite of its benign characteristic, due to its stick attachment to the surround structures. Forcing the total removal may lead unwanted patient outcomes and intraoperative complications. Surgical management of dermoid tumors should be rearranged and revised in the light of intraoperative observations and surgical state of affairs. Thus, operative plan and staging applied to dermoid tumors should be pathology-specific including location, adhesiveness and clinical manifestations.

Ethics

Informed Consent: Consent was obtained from all patients.

Authorship Contributions

Surgical and Medical Practices: A.Ü., İ.D., O.Ö., Ü.E., O.M., Concept: İ.D., O.Ö., Design: O.Ö., O.M., Data Collection or Processing: İ.D., O.Ö., O.M., Analysis or Interpretation: O.Ö., Ü.E., O.M., Literature Search: İ.D., O.Ö., Ü.E., O.M. Writing: A.Ü., İ.D., O.Ö., Ü.E., O.M.

Conflict of Interest: No conflict of interest was declared by the authors.

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