

Olgu Sunumu

Epidermoid Tumor of the Fourth Ventricle with Accompanying Congenital Posterior Arch Defect of the Atlas

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Epidermoid tumors are rarely encountered within the fourth ventricle. There are slightly more than 100 cases reported in the literature. Congenital posterior arch defects of the atlas are also rare with less than 50 cases reported in the literature. The coexistence of these two pathologies in one patient has not been previously reported. We present a 51-year-old woman with an epidermoid tumor of the fourth ventricle and accompanying posterior arch defect of the atlas.

Keywords: Epidermoid tumor, fourth ventricle, atlas, posterior arch, congenital anomaly

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Dördüncü Ventrikül Epidermoid Tümörü ve Eşlik Eden Konjenital Atlas Posterior Ark Defekti

Epidermoid tümörler ender olarak dördüncü ventrikül içinde görülürler. Literatürde bu şekilde bildirilmiş 100 kadar olgu mevcuttur. Atlas'ın posterior ark defektleri de literatürde bildirilen 50'den az olgu ile oldukça enderdir. Bu iki patolojinin bir arada bulunması daha önce bildirilmemiştir. Bu çalışmada, 4. ventrikül epidermoid tümörü ve eşlik eden atlas posterior ark defektinin olduğu 51 yaşında bir kadın hasta sunulmuştur.

Anahtar kelimeler: Epidermoid tümör, dördüncü ventrikül, atlas, posterior ark, konjenital anatomi

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INTRODUCTION

Epidermoid tumors constitute approximately 1-2% of all intracranial tumors^(5,10). Most common site of their occurrence is the cerebellopontine angle (CPA) followed by sellar and parasellar regions^(1,5,9,10). Fourth ventricle is a relatively rare location for epi-

dermoid tumors with more than 100 reported cases^(4,10). Fourth ventricle epidermoid tumors account for 5-31.4% of all intracranial epidermoid tumors⁽⁵⁾. Herein we present a case of a fourth ventricle epidermoid tumor with accompanying congenital posterior arch defect of the atlas. To our knowledge, it is the first report to indicate such a coincidence.

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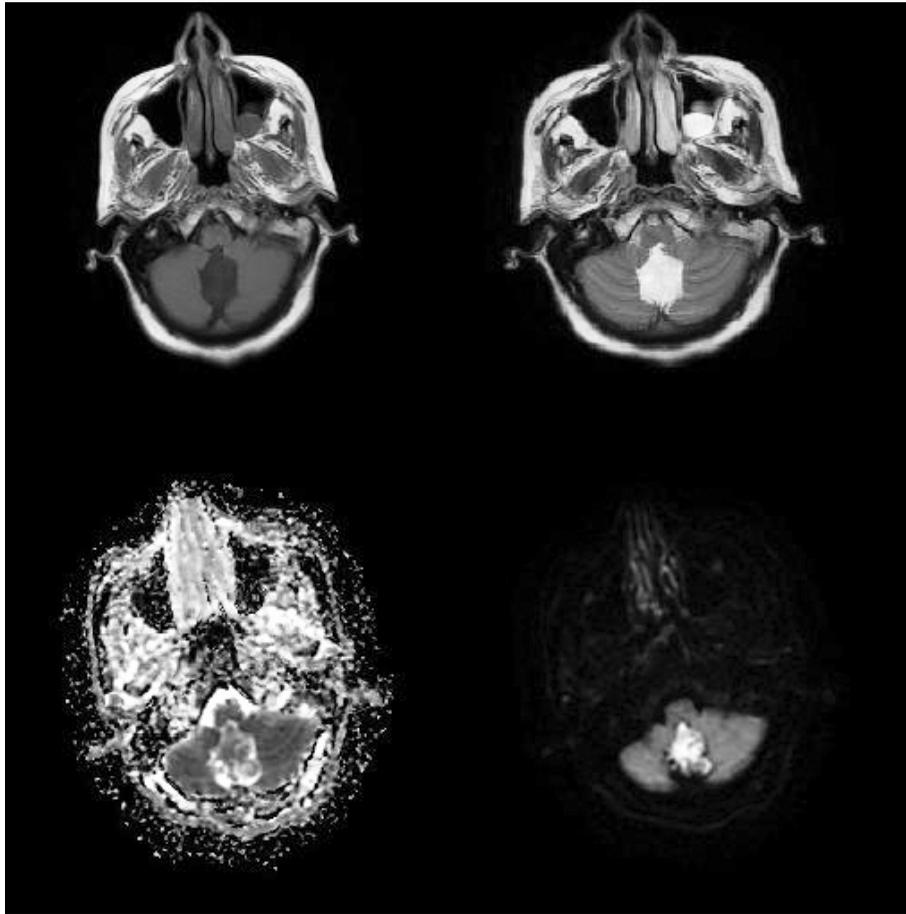


Figure 1. T1- (top row, left) and T2- weighted (top row, right) MRI showed an isointense lesion within fourth ventricle, with markedly restricted diffusion on DWI (bottom row).

CASE REPORT

A 51-year-old woman, with no other medical history was admitted with headache and nausea continuing for the last 3 years. One year ago, the patient was admitted to another institution with the same complaints. The computed tomography (CT) scan revealed a hypodense lesion, which was attributed to an arachnoid cyst or megacisterna magna. Symptomatic treatment was administered. No further investigation was made. The patient's current neurological examination revealed gait ataxia and nystagmus. A CT scan was obtained and a 3.3x2.3x2.2 cm hypodense lesion extending from the fourth ventricle to the cisterna magna was demonstrated. The CT slices were obtained just above the C1 level. There-

fore, preoperatively the defect on the posterior arch of the atlas was not seen. Cranial magnetic resonance imaging (MRI) revealed a lesion, that is isointense on T1- and T2- weighted images and slightly heterogenous and hyperintense on FLAIR images (Figure 1). Diffusion-weighted imaging (DWI) showed markedly restricted diffusion of the lesion with high hyperintensity, which is more likely in accordance with an epidermoid tumor (Figure 2).

The patient underwent surgery in prone position, and through a midline incision, suboccipital craniectomy and C1 posterior arcusotomy were performed. The midline dissection was made with no: 11 scalpel blade and muscle tissue was bluntly removed laterally. No monopolar cautery



Figure 2. Sagittal T1-weighted MRI demonstrated the extension of tumor below C1 level. The defect on the posterior arch of C1 can be seen (marked with the arrow). There is a small fibrous band instead of posterior arch on the midline sagittal MRI section.

was used and the posterior arch of the atlas was clearly exposed. A midline fusion defect on the posterior arch and on two hemiarches were seen. There was no separate ossicle in between the two hemiarches, instead a thick fibrous bridging was found. As the suboccipital craniectomy and C1 posterior arcusotomy was performed, the dura was opened in Y-fashion. The white glistening, pearl-like tumor was exposed underneath the arachnoid membrane. With no significant adherence to the adjacent brain tissue, the tumor was totally removed. Pathological diagnosis was in accordance with epidermoid tumor. With retrospective view the defect on the posterior arch of the atlas was seen in sagittal MRI (Figure 2). Early postoperative CT scan showed resection of the tumor. The postoperative course of the patient was uneventful and she was discharged from the hospital without any neurological deficit. Control MRI scan showed total resection of the tumor, without diffusion restriction on DWI.

DISCUSSION

Epidermoid tumors are maldevelopmental, slow growing tumors. They arise from displaced epithelial remnants during the neural tube closure or during the formation of cerebral vesicles, between the third and fifth weeks of gestation (1,4,10). Therefore, they are also named as “inclusion tumors”. The tumor has an outer capsule of connective tissue, which surrounds a layer of keratinized squamous epithelium and a cystic content of desquamated keratin, cellular debris and cholesterol crystals (4,10). Epidermoid tumors slowly grow within subarachnoid spaces and may remain asymptomatic until they reach a significant size. Usually symptoms are caused by compression to the cerebellum, brain stem or cranial nerves (9). Headache, gait ataxia and vertigo are common symptoms (5). Epidermoid tumors appear as irregular hypodense lesions on CT scans, whereas they are iso- or slightly hyperintense on T1 and T2-weighted images on MRI scans (1,4). Their radiological appearance may resemble arachnoid cysts, therefore DWI and FLAIR sequences are performed to make a differential diagnosis. Restriction of the diffusion on DWI and hyperintensity on FLAIR images are typical for epidermoid tumors (1,4,10). Surgical goal is total removal of these lesions, however due to severe adhesions to the floor of the fourth ventricle or adjacent neural elements this is not always possible. However, a subtotal resection with preservation of the neural function provides also a fairly good outcome, due to benign nature of these tumors (10). It is not wrong to claim that a total resection could be most possibly achieved in the initial surgery, in the absence of critical adhesions. Our case demonstrated similar radiological and clinical characteristics with the cases presented in the current literature.

Congenital defects of the posterior arch of the atlas are very rare. Less than 50 articles have been published about these defects so far (3,6,7). There

are three primary ossification centers in the C1 vertebra. One anterior center, that forms the anterior tubercle and two lateral centers, which are responsible for the formation of the lateral masses and the posterior arch ^(6,7). The lateral ossification centers appear at the seventh week of embryonic development and they extend dorsally to form the posterior arch ⁽⁷⁾. At birth, the two hemiarches are almost fused, excepting a gap of few millimeters ^(7,8). The fusion is completed between 4-9 years of life ^(3,7). Defects of the posterior arch are encountered in 3-5 % of the population ⁽⁷⁾. Although several theories have been postulated to explain the mechanism of the formation of these defects, their etiopathogenesis is still unclear. Posterior arch defects are considered as a developmental failure of chondrogenesis of the arch, rather than a defect in the subsequent ossification ^(3,6,7). Congenital defects of the C1 posterior arch are generally asymptomatic and diagnosed incidentally on neuroimaging studies ⁽⁸⁾. Currarino et al. proposed a classification for these defects ⁽²⁾: Type A (failure of posterior midline fusion of the two hemiarches with a small gap), Type B (unilateral defect ranging from small gap to complete absence of a hemiarch), Type C (bilateral defect with preservation of the most dorsal part), Type D (absence of posterior arch, with persistent posterior tubercle), Type E (absence of the entire arch, including the tubercle). Our case demonstrated a Type A defect.

The association of these defects with different genetic or congenital disorders, such as Arnold-Chiari malformation, Klippel-Feil syndrome, gonadal dysgenesis, Turner and Down syndromes is well-known ^(6,7). However, there is no known association between intracranial tumors and congenital posterior arch defects of the atlas. Dermoid tumors may have a dermal sinus tract or other type of accompanying midline defects, but such a condition is not known for the epidermoid tumors. The congenital failures that may lead to development of the epidermoid tumor and pos-

terior arch defect of the atlas occur on separate gestational weeks and the mechanisms of their occurrence are different. Although there was no clear association between these two pathologies, genetic basis of both pathologies should be further investigated with genetic studies, in order to establish a connection.

One important notice is the potential complications that may occur during surgery due to the defect in the posterior arch. In cases with incomplete preoperative documentation, such as in our case, where the posterior arch defect of the atlas was not seen preoperatively, there is a potential risk of iatrogenic injury with the use of monopolar cautery during muscle dissection. Our main preoperative focus was the tumor and our strategy was to achieve a total resection at the first operation. Although with retrospective view we could see the posterior arch defect on the MRI, we were unaware of the defect before surgery, but with careful and meticulous dissection any complication did not occur. Our suggestion is to avoid use of monopolar cautery, and instead prefer sharp dissection with blades to prevent potential injuries during the posterior approach to the craniocervical junction, especially in syndromes or diseases like Turner, Down and Klippel-Feil syndromes or Arnold-Chiari malformation, where there is a clear association with posterior arch defect of the atlas.

REFERENCES

1. Bohara M, Yonezawa H, Hanaya R, Takeshita S, Sumida M, Arita K. Posterior fossa epidermoid cysts presenting with unusual radiological appearances. *Neurol Med Chir (Tokyo)* 2011;51:85-8. <https://doi.org/10.2176/nmc.51.85>
2. Currarino G, Rollins N, Diehl JT. Congenital defects of the posterior arch of the atlas: a report of seven cases including an affected mother and son. *Am J Neuroradiol.* 1994;15:249-54.
3. Elmalky MM, Elsayed S, Arealis G, Mehdian H. Congenital C1 arch deficiency: Grand Round presentation. *Eur J Spine* 2013;22:1223-6. <https://doi.org/10.1007/s00586-013-2682-z>
4. Forghani R, Farb RI, Kiehl TR, Bernstein M. Fourth ventricle epidermoid tumor: Radiologic, intraoperative

- and pathologic findings. *Radiographics* 2007;27:1489-94.
<https://doi.org/10.1148/rg.275075011>
5. Kambe A, Anno Y, Oda N, Shiomi Y, Aoki H, Sasaki A. Reversible bilateral vestibular impairment caused by fourth ventricle epidermoid tumor. *Neurol Med Chir (Tokyo)* 2003;43:201-3.
<https://doi.org/10.2176/nmc.43.201>
 6. Kwon JK, Kim MS, Lee GJ. The incidence and clinical implications of congenital defects of the atlantal arch. *J Korean Neurosurg Soc.* 2009;46:522-7.
<https://doi.org/10.3340/jkns.2009.46.6.522>
 7. Sabuncuoglu H, Ozdogan S, Karadag D, Timurkaynak E. Congenital hypoplasia of the posterior arch of the atlas: Case report and extensive review of the literature. *Turk Neurosurg.* 2011;21:97-103.
 8. Sanchis-Gimeno JA, Aparicio L. Posterior arch defect in a dry atlas. *Eur Spine J.* 2011;20:1574-5.
<https://doi.org/10.1007/s00586-011-1715-8>
 9. Tamura Y, Ishikawa M, Iwasaki K, Kondo A. Epidermoid cyst of the cisterna magna presenting with cervicomedullary compression after trauma. *Acta Neurochir. (Wien)* 2002;144:947-8.
<https://doi.org/10.1007/s00701-002-0962-1>
 10. Tancredi A, Fiume D, Gazzeri G. Epidermoid cysts of the fourth ventricle: very long follow-up in 9 cases and review of the literature. *Acta Neurochir (Wien)* 2003;145:905-11.
<https://doi.org/10.1007/s00701-003-0008-3>