

# A Large Choroid Plexus Papilloma Removed by the Cerebellomedullary Fissure Approach. Case Report and Review of the Literature

## Vaka Sunumu ve Literatürün Gözden Geçirilmesi: Serebellomedüller Fissür Yaklaşımıyla Koroid Pleksusa Yerleşmiş Papillomun Çıkarılması

### ABSTRACT

We report a case of large choroid plexus papilloma of the fourth ventricle in a 23-year-old woman. She presented with severe headache, dysphagia, and gait disturbances. Horizontal nystagmus, ataxic gait and quadriparesis were detected on initial examination. Imaging studies showed a large mass in the left side of brain stem and a marked hydrocephalus. The tumour was removed by microsurgical dissection of the cerebellomedullary fissure. We have discussed the effectiveness of this approach for removal of bulky tumors of the fourth ventricle and reviewed the literature about its benefits and potential hazards.

**KEY WORDS:** Cerebellomedullary Fissure, Choroid plexus papilloma, Fourth ventricle, Hydrocephalus, Microsurgery

### ÖZ

Bu yazıda, 23 yaşında kadın hastada 4. Ventrikül yerleşimli koroid pleksus papilloma vakası sunulmaktadır. Hastada şiddetli baş ağrısı, yutma güçlüğü ve yürümede dengesizlik yakınmaları vardır. Nörolojik muayenede: Horizontal nistagmus, ataksik yürüme ve tetraparezi tesbit edildi. Görüntüleme çalışmalarında; beyin sapının sol tarafında geniş bir kitle lezyonu yanısıra hidrosefalide tesbit edildi. Serebellomedüller fissür mikrocerrahi olarak diseke edilerek tümör çıkarıldı. Bu yazıda cerebellomedüller fissürün mikrodiseksiyon ile açılarak büyük kitlelerin beyin sapından çıkarılmasının etkinliği ve potansiyel komplikasyonları mevcut literature ve sunulan vaka eşliğinde tartışıldı.

**ANAHTAR SÖZCÜKLER:** Serebellomedüller Fissür, Koroid pleksus papilloma, 4. ventrikül, Hidrosefali, Mikrocerrahi

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## INTRODUCTION

Tumours that are located in the third and the fourth ventricles are always worrisome for the neurosurgeon, for access to those areas is a challenging work and post-operative complications are usually inevitable. Conventional route of access to the fourth ventricle is through a section in the vermis which may result in many complications such as ataxia (10, 15). One alternative way is to use cerebellomedullary fissure as a corridor to reach the fourth ventricle that was first described by Matsushima et al (9).

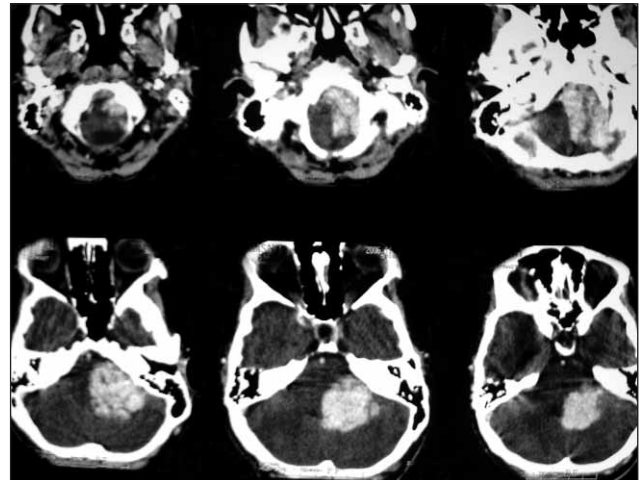
The cerebellomedullary fissure is located between cerebellar tonsils and medulla oblongata. Lateral and rostral extensions of this fissure are cerebellomedullary and cerebellopontine cisterns, and fastigium respectively. Tela choroidea and inferior medullary vellum compose the floor of the fissure. The posterior inferior cerebellar artery runs through the fissure and the vein of the cerebellomedullary fissure runs in the fissure toward the cerebellopontine angle and eventually drains into the superior petrosal sinus. The floor of the cerebellomedullary fissure composes the roof of the fourth ventricle (6). We used the cerebellomedullary fissure approach to remove a large papilloma of the fourth ventricle.

## CASE REPORT

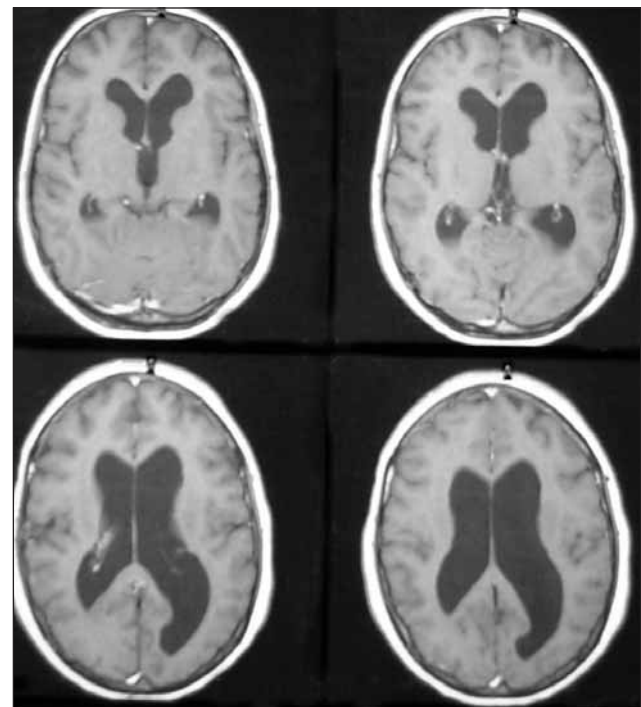
A 23-year-old woman presented with gait disturbances, severe headache and dysphagia developed in the last five months. Horizontal nystagmus, a weak gag reflex, ataxic gait and quadriparesis with an upward plantar reflex were detected on examination. Imaging studies showed a large mass in the left side of the brain stem which was enhanced with radiocontrast. Moreover, a marked hydrocephalus, more prominent in the left side, was present (Figure 1 –D).

### Surgical technique

The patient was positioned between lateral decubitus and prone while the head was flexed. A midline incision was made frominion to the spinous process of C3. After a far lateral suboccipital craniotomy, the dura was opened and a large, light brown, shiny, lobulated gritty tumour was seen. Cerebellar tonsils were retracted and, with the use of operating microscope, piecemeal removal of the tumour performed until near-total resection. Bleeding persisted from a small remnant



*Figure 1A: Contrast-enhanced CT scan showing the tumor location in the posterior fossa.*



*Figure 1B: Axial MR Image demonstrates the marked bilateral hydrocephalus.*

anteromedially along the basilar artery that was terminated by changing the view of the microscope and slopping the operating table by which total removal of the tumour was achieved. The tumour's abundant feeding arteries were derived from the vertebral artery and posterior inferior cerebellar artery, but the most important feeders which had to be occluded to attain a good haemostasis originated from the basilar artery. A ventriculoperitoneal shunt was then placed. Post-operative imaging proved total resection of the tumour (Figure 2 A,B,C).

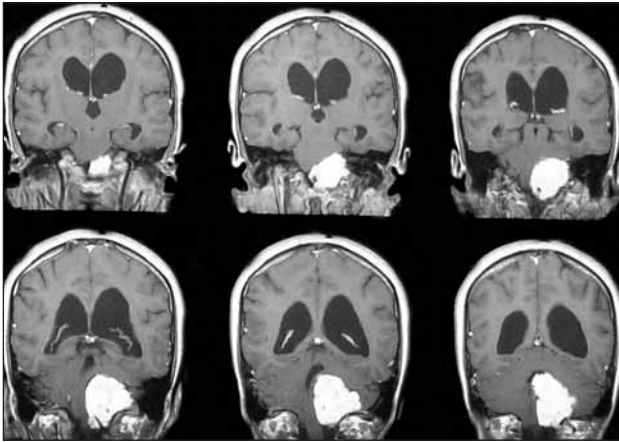


Figure 1C: Coronal MR Image depicts how the tumor creates pressure on the brain stem.

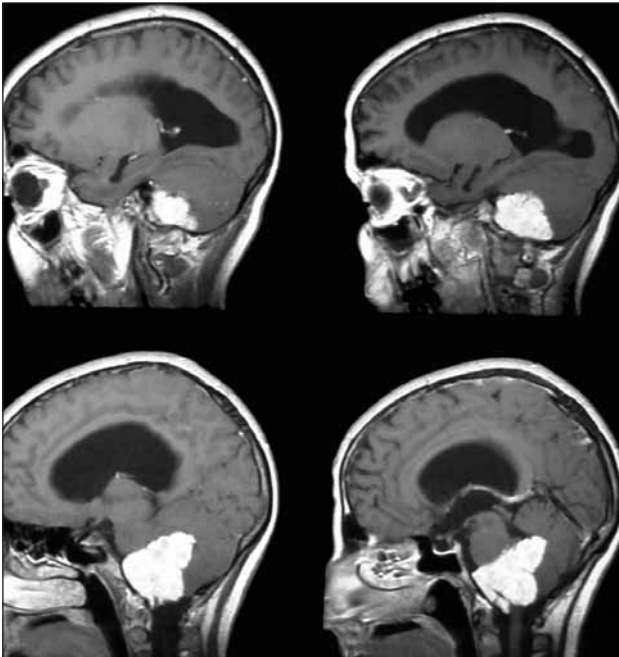


Figure 1D: Sagittal views of the same tumor.

In histopathological studies, pinkish friable highly vascular fragments of tissues with villi-form surface were observed on gross examination. A complex array of delicate branching fibrovascular connective tissue covered by a single layer of uniform cuboidal to columnar epithelial cells, with round to oval, basally situated, relatively monomorphic nuclei, with no mitotic activity, all together confirmed papilloma as the first diagnosis (Figure 3 A,B).

### DISCUSSION

Here, we reported a large papilloma of the fourth ventricle which was removed using the

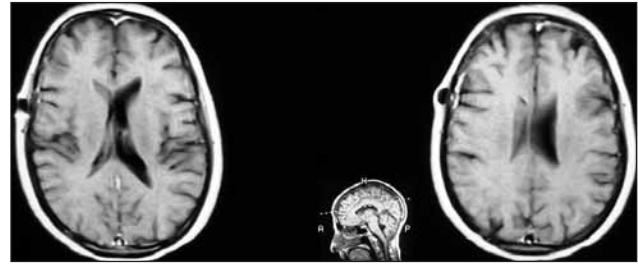


Figure 2A: Post-operative axial MR image 6 months after surgery shows the resolution of the hydrocephalus and complete removal of the tumor.

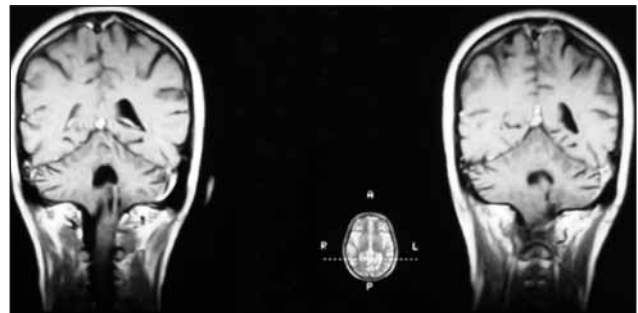


Figure 2B: Coronal view.

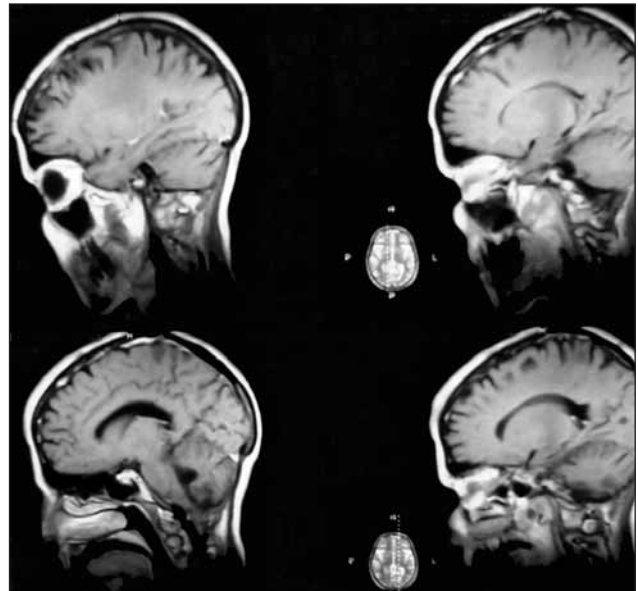
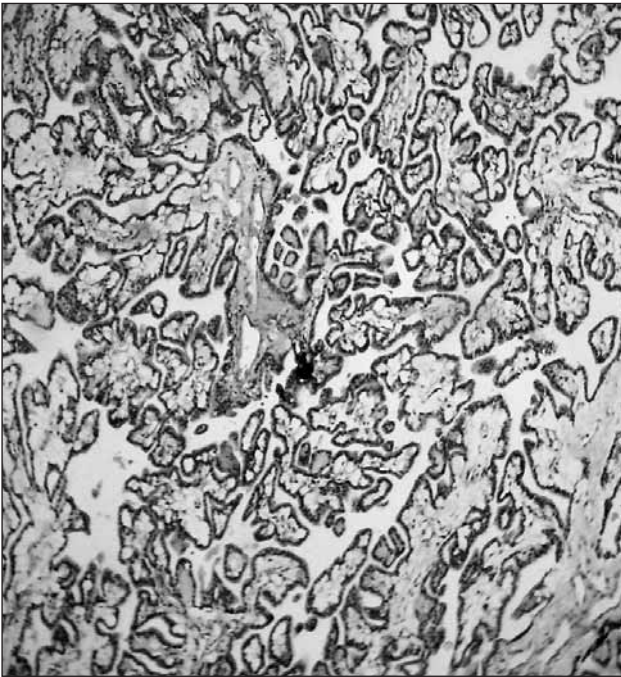


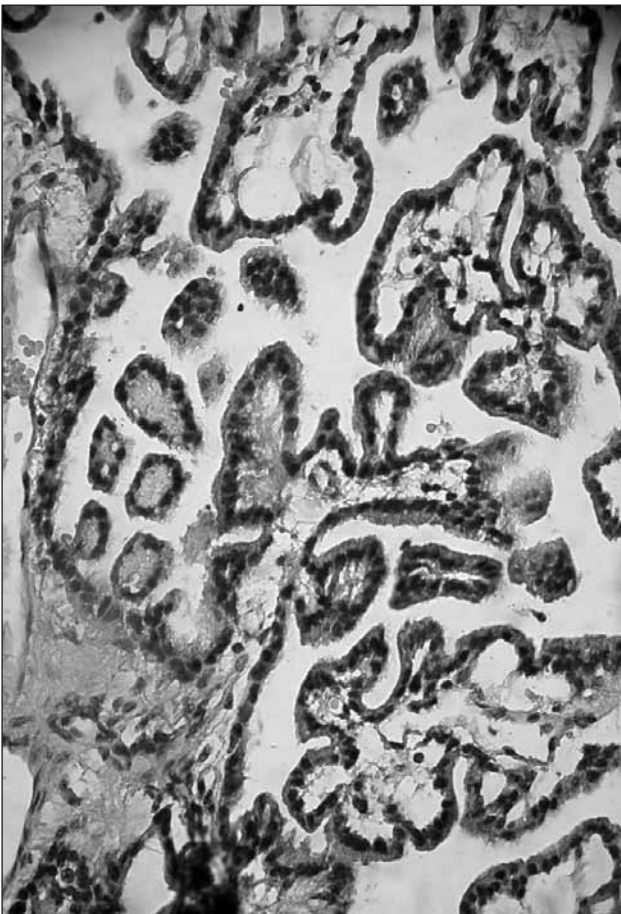
Figure 2C: Sagittal view.

cerebellomedullary fissure approach. Choroid plexus papilloma is a benign neoplasm that arises from the epithelium of the choroid plexus. This tumour is rare in both adults and children, comprising less than 1% of all adult intracranial neoplasms and 4 to 5% of all pediatric intracranial neoplasms (1,5). The most common site of the choroid plexus papilloma is the fourth ventricle in





**Figure 3A:** Light microscopic findings of papilloma of the fourth ventricle in tissue section H&E, x100.



**Figure 3B:** Light microscopic findings view of the same section, x400.

adults and the lateral ventricles in pediatric patients (2,8).

Tumours of the fourth ventricle, are generally resected by splitting of the vermis, a well-established approach, which may result in some complications such as cerebellar mutism (especially in children), ataxia, staggering gait and oscillation of the trunk and neck. It is also necessary to retract the cerebellar hemispheres to get access to a deep-located tumour and this may cause some injuries (10,15). To avoid such complications, the cerebellomedullary fissure provides a good corridor to the fourth ventricle. Matsushima was the first to make use of the microsurgical anatomy of the cerebellomedullary fissure and used this approach to get satisfactory results in nine patients (9). Subsequently, Kellogg and Piatt (7) and then Ziyal et al (15) reported successful application of this approach for tumours of the fourth ventricle. Since then, some other studies reported the successful use of this approach for different tumours of the fourth ventricle namely: epidermoid and dermoid cysts, astrocytomas, medulloblastomas, ependymomas, meningiomas, hemangioblastoma, choroid plexus carcinoma, choroid plexus papilloma, adenocarcinoma in the pons, cavernoma in the medulla, (3,4) papillary thyroid carcinoma metastasis and a distal posterior inferior cerebellar artery (PICA) aneurysm (15). The surgical outcome had been favorable in different case series. Total removal was achieved in roughly 82% of cases (3,4,14). Total removal is not always achievable in tumours focally attached to critical areas in the fourth ventricle (3). This approach has also been used as so-called uvulotonsillar approach to remove an AVM of the inferior medullary velum (13).

The most important feeder artery was the vertebral artery in the reported patients, in contrast to the normal feeders of choroid plexus reported previously which are the anterior inferior cerebellar artery, posterior inferior cerebellar artery and superior cerebellar artery (12). A study reports that combination of the infratentorial supracerebellar approach and the cerebellomedullary fissure approach may cause cerebellar venous insufficiency with venous congestion and possible venous infarction and these two approaches should therefore not be used together (11).

We again insist on the applicability of the subtonsillar-transcerebellomedullary approach in diverse pathologies which may be reached through

the cerebellomedullary fissure and the lateral aspect of the fourth ventricle and proved that this approach is suitable even for bulky tumours of this area with minimal destruction of fine neural tissues. It is therefore important for neurosurgeons to know its exact anatomy and the clinical situations to which it can be applied.

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