

# Characterization of an Intracranial Neurothekeoma: Case Report

## *Intrakranial Nörotekoma: Olgu Sunumu*

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### ABSTRACT

**AIM:** Neurothekeomas are benign tumors of presumed neural sheath origin. They are primarily found in superficial soft tissues, located in the upper portion of the body. Here, we report a case of intracranial neurothekeoma.

**RESULTS:** A 37-year-old female presented at our clinic with sudden-onset left hemifacial pain of varying duration. The physical and neurological examination findings were normal. The magnetic resonance imaging scan showed a mass compressing the pons and extending from the medial section of the left middle fossa to the posterior fossa. The patient was operated on using a left presigmoid transpetrosal approach and the mass was totally removed.

**CONCLUSION:** Neurothekeomas, also known as nerve sheath myxomas, are rare benign tumors. There have been two previous cases reported with an intracranial location. The information presented here now represents the third such case in the literature.

**KEYWORDS:** Brain tumor, Intracranial, Nerve sheath myxoma, Neurothekeoma

### ÖZ

**AMAÇ:** Nörotekomalar nöral kılıf orjinli iyi huylu tümörlerdir. Genellikle gövdenin üst kısımlarındaki yüzeysel dokularda rastlanır. Biz burada bir intrakranial nörotekoma vakasını sunuyoruz.

**BULGULAR:** 37 yaşında kadın hasta kliniğimize, yüzünün sol yarısında ani başlayan ağrı şikayeti ile başvurdu. Fizik ve nörolojik muayene bulguları normaldi. Kranial MR da sol orta kranial fossadan posterior fossaya doğru uzanan ponsa bası yapan kitle saptandı. Hasta sol presigmoid transpetrosal yaklaşımla opere edildi ve kitle tamamen çıkartıldı.

**SONUÇ:** Nörotekomalar, diğer bilinen adıyla sinir kılıfı miksomaları, iyi huylu tümörlerdir. Literatürde intrakranial lokalizasyonda bildirilen 2 vaka vardır ve biz burada 3. vakayı sunuyoruz.

**ANAHTAR SÖZCÜKLER:** Beyin tümörü, İntrakranial, Sinir kılıfı miksoması, Nörotekoma

### INTRODUCTION

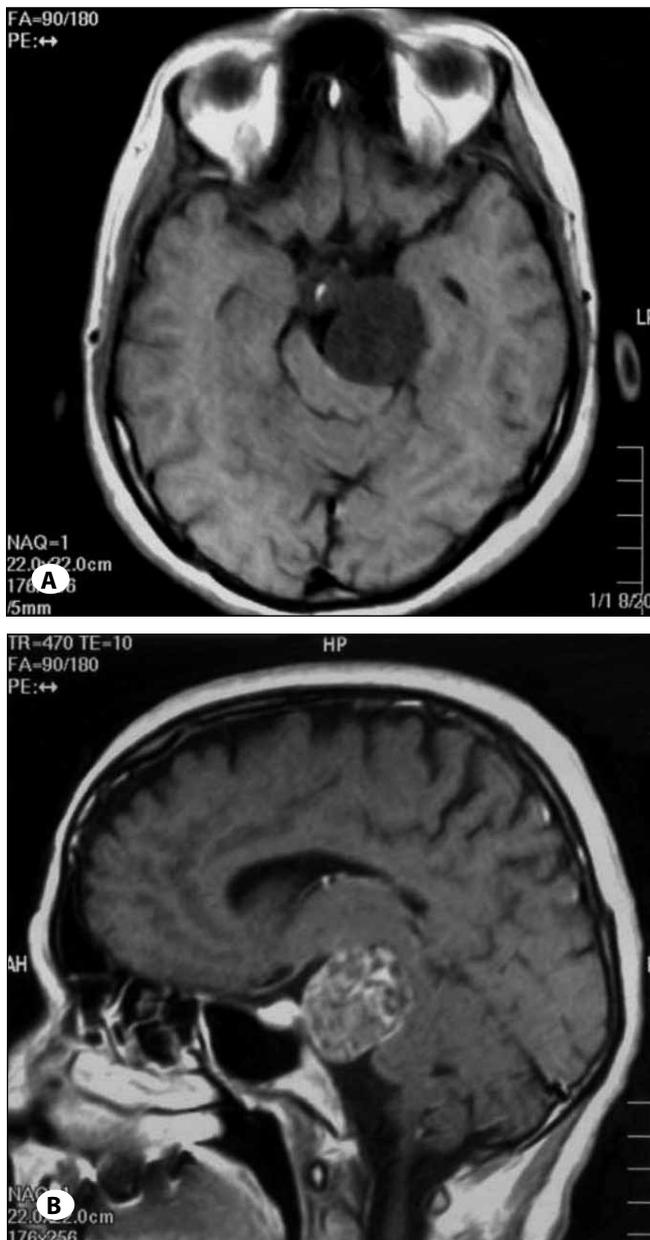
Neurothekeomas are benign tumors of probable nerve sheath origin. These lesions were first described in 1969 by Harkin and Reed who called them nerve sheath myxomas (6). Gallager and Helwig later re-named this tumor neurothekeoma (4). The dermal layer of the skin is the most common location for neurothekeomas and presentation is usually as a solitary nodule in the head, neck or upper limbs (1). The intracranial localization is quite rare and there have been only two previously reported cases (11,13). Here, we present a 37-year-old female with an intracranial neurothekeoma.

### CASE REPORT

A 37-year-old female presented at our clinic with left hemifacial pain. The patient complained of atypical facial pain that had started a year ago. The pain started suddenly and the duration varied. She had not benefited from any analgesics during this period. The clinical and neurological examination results were

normal. Hematological and biochemical investigations were also within normal limits. The cranial magnetic resonance imaging (MRI) scan showed a left prepontine extraaxial mass approximately 35 mm in diameter compressing the pons and mesencephalon. The mass was hypointense on T1-weighted images and hyperintense on T2weighted images with heterogeneous enhancement following contrast agent administration (Figure 1A,B).

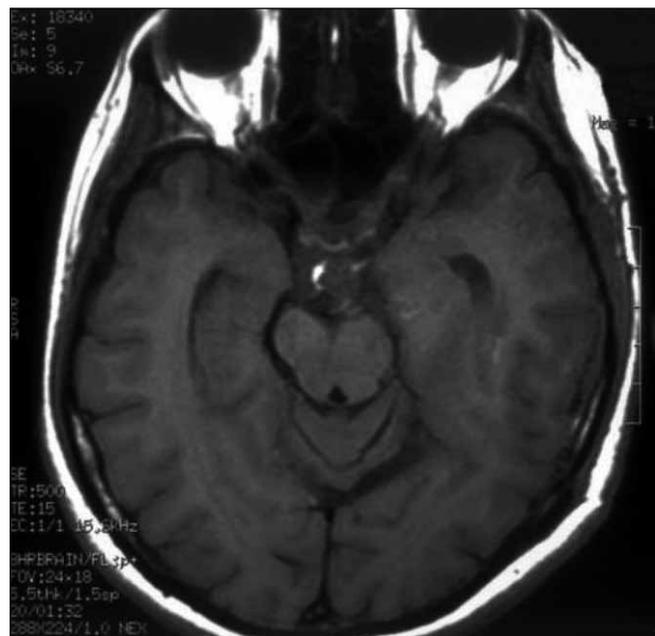
The patient was operated on using a left presigmoid transpetrosal approach. A soft, grey mass with little bleeding was totally removed. The mass was attached to the dura and no relation to the major nerves was observed. A paresis of down and in eye movements developed due to 4th nerve trauma postoperatively. This deficit resolved on the postoperative 8<sup>th</sup> day and the patient was discharged 2 days later. The cranial MRI performed one month after surgery did not show any residual tumor (Figure 2).



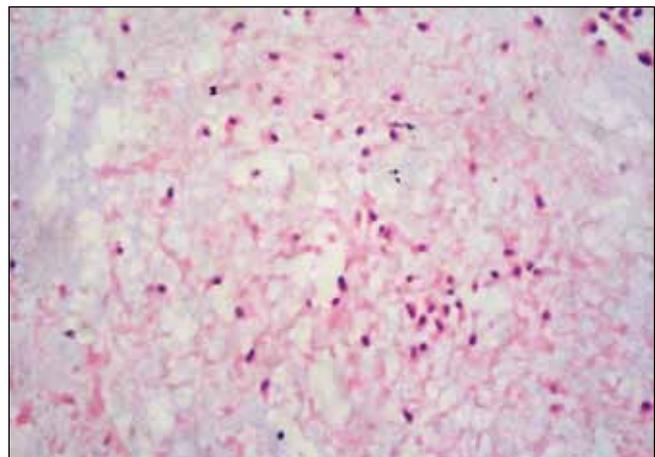
**Figure 1:** A) T1-weighted axial, B) T1weighted postcontrast sagittal magnetic resonance imaging (MRI) images. The mass was hypointense on T1-weighted MRI and hyperintense on T2-weighted images. Post-contrast study revealed nonhomogeneous enhancement.

**Neuropathological findings:**

In order to characterize the neurothekeoma, histological and immunohistochemical staining was performed. Histologically, cells with nuclei showing spindle-shaped extensions and large, pale cytoplasm placed in multilobulated groups in a myxoid stroma were observed. Mild nuclear pleomorphism was noted in places. No mitosis was seen. Regarding the immunohistochemical studies, diffuse positive staining of the cells with S-100 and vimentin was observed. Glial fibrillary



**Figure 2:** The postoperative axial MRI of the case.



**Figure 3:** Cells with nuclear extensions and clear, vacuolated cytoplasm showing mild nuclear pleomorphism in a myxoid stroma (H&E, x100).

acidic protein (GFAP), epithelial membrane antigen (EMA), E-cadherin, HBME-1, HMB-45, cytokeratin and desmin staining results were negative (data not shown). These histological and immunohistochemical findings are considered characteristic of neurothekeoma (Figure 3).

**DISCUSSION**

Neurothekeoma is a recently characterized benign cutaneous tumor usually arising on the upper trunk or head and neck of children and young adults. It was first described by Harkin and Reed as a nerve sheath myxoma, and various names have later been used such as pacinian neurofibroma, cutaneous lobular neurofibroma, perineural neurofibroma and neurothekeoma (4,6,7,14,16).

From the neurosurgical point of view, it is important to differentiate this tumor from schwannomas, sarcomas, meningiomas and gliomas with myxoid degeneration, as well as from cardiac myxomas metastasizing to the brain. There are two previous case reports that describe neurothekeomas with an intracranial location. The tumor was in the cerebral parenchyma in one of these cases and in the parasellar region in the other (11,13). In the case with the parasellar neurothekeoma, the authors did not see any relationship between the tumor and a major nerve, and therefore postulated that the intracranial variant of this tumor was derived from small nerve branches (13). In the case with the neurothekeoma in the middle cranial fossa brain parenchyma, there was also no tumor attachment to any nerve. The authors reported that the tumor could possibly have originated from the schwann cells innervating the blood vessels, while another possibility is divergent differentiation of the persistent pluripotent cell rests resting inside the brain parenchyma under suitable conditions (11). Consistent with these findings, we did not observe a relationship between the tumor and any nerve in this case report. We speculate that the tumor in our case may have derived from dural nerve branches as it was attached to the dura.

Intracranial schwannomas rarely originate from the oculomotor and trochlear nerves and they are usually seen in the oculomotor and the ambient cistern, respectively (15). Both of them can be manifested as a middle cranial fossa lesion. The differential diagnosis of these rare tumors can be made from neurothekeoma with careful evaluation of radiological and histopathological findings.

A few myxoid tumors arising within the spinal column, pituitary fossa or the posterior fossa have been reported (9,10,12). However, the pathological documentation was not sufficient to decide whether they were of schwann cell, glial or mesenchymal origin (13).

In our case, the neurothekeoma was hypointense in T1-weighted images and hyperintense in T2-weighted images using MRI. Furthermore, there was heterogeneous enhancement following contrast agent administration. These features are similar to the MRI findings of a previously published neurothekeoma case localized to the parasellar region (13).

Diagnosis of neurothekeoma is confirmed via histopathological evaluations and histochemical reactivity profiles. Specifically, neurothekeomas have characteristic light microscopic findings and can be subclassified into myxomatous and cellular variants (1,2). The myxomatous type generates large amounts of myxoid matrix and is typically immunoreactive to the S-100 protein (1-3,8). The cellular type contains little myxoid material and shows no immunoreactivity to the S-100 protein (5). Neurothekeoma cells are usually spindle-shaped and arranged in fascicular or whorled patterns. Epithelioid cells have been observed in these tumors. Neurothekeomas can also show immunoreactivity to vimentin.

Incomplete excision of neurothekeomas may lead to recurrence, therefore total excision is recommended. Aggressive recurrences or metastases have not been reported with cutaneous neurothekeomas. Similar findings hold true for intracranial neurothekeomas, where the previous two intracranial neurothekeomas cases (11,13), as well as the case presented here, underwent gross-total or total excision and no recurrences were reported. Because of this lesion is rarely seen in intracranial localization, we advise the longer follow up and close watch.

In conclusion, intracranial neurothekeomas are benign lesions similar to the cutaneous type and not recur following total excision. A definitive diagnosis of neurothekeoma can be made with immunohistochemical analysis.

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