# TANYCYTIC EPENDYMOMA OF THE FILUM TERMINALE REGION; A CASE REPORT

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# TANICITIČNI EPENDIMOM FILUM TERMINALE REGIONA; PRIKAZ SLUČAJA

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## ABSTRACT

*Tanycytic ependymoma is a very rare spindle-cell variant of ependymoma derived from tanycytes, which are part of the primitive nervous system. This paper is presenting 48-year old woman who presented with low back and right-sided leg pain of moderate intensity. MRI showed spinal intradural tumor at the level of the L1 vertebral body. Right-sided L1 hemilaminectomy and en bloc tumor resection were performed. Neuroradiological and intraoperative diagnosis of schwannoma was revised to tanycytic ependymoma after careful immunohistochemical analysis. Six months post-operativly, MRI did not show tumor recurrence. Tanycytic ependymoma at the region of filum terminale is extremely uncommon and only three cases have been described in the literature. The low incidence of this tumor and atypical histological image, which is distinct from the typical features of commonly encountered ependymomas, can present a challenge in terms of making an accurate diagnosis. Awareness of this transitional form of ependymoma among neurosurgeons and pathologists may avoid incorrect surgical approaches and postoperative treatment course.*

**Key words:** *tanycytes, ependymoma, filum terminale*

## SAŽETAK

*Tanicitični ependimom je vrlo retka forma ependimoma koja vodi poreklo od tanicita, ćelija koje su deo primitivnog nervnog sistema. Ovde je prikazan klinički slučaj pacijentkinje stare 48 godina, kod koje je bolest počela umerenim bolovima u donjem delu leđa i desnoj nozi. MR pregled je ukazao na spinalni intraduralni tumor u nivou L1 prršljenskog tela. Nakon hemilaminektomije na L1 nivou sa desne strane, tumor je uklonjen u celosti. Nakon pažljive analize imunohistohemijskih preparata, neuroradiološka i intropeorativna dijagnoza švanoma je revidirana u tanicitični ependimom. MR pregled nakon 6 meseci od operacije nije ukazao na sigurne znake recidiva tumora. Tanicitični ependimom u filum terminale regionu je izuzetno redak i do sada je u literaturi opisano 3 slučaja. Niska inidenca ovog tumora i atipična histološka slika, koja se razlikuje od drugih čestih ependimoma u ovoj regiji, može biti veliki izazov pri pokušaju donošenja precizne dijagnoze. Podizanjem svesti o postojanju ove forme ependimoma među neurohirurzima i patolozima mogu se izbeći pogrešni hirurški pristupi, kao postoperativna evaluacija i tok lečenja.*

**Ključne reči:** *taniciti, ependimom, terminalni filum*

## ABBREVIATIONS

**MRI** - magnetic resonance imaging
**L** – lumbar
**HE** – hematoxylin-eosin

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## INTRODUCTION

Ependymomas are tumors of neuroectodermal origin which usually arise from the ependymal cells in the central canal of the spinal cord, the filum terminale region, choroid plexus or white matter adjacent to the ventricular surface of the brain (1). The annual incidence rate of all ependymomas in Europe is around 2 cases per million, occurring more often in men than women (2), and approximately 15% of all patients are children younger than 5 years (3). Spinal cord and filum terminale lesions are typically associated with back pain of long duration, and motor or sensory deficits of lower and upper extremities. Tanycytic ependymoma is an even more rare spindle-cell variant of ependymoma derived from tanycytes, which are part of the primitive nervous system. By reviewing the scientific papers that have been published so far, it is possible to find three similarly described cases of tanycytic ependymoma occurring at the region of filum terminale (4-6).

In the present paper we report a rare case of a tumor of the cauda equina region in a 48-year-old woman in whom the intraoperative diagnosis of schwannoma was revised to tanycytic ependymoma after the application of immunohistochemical stains and careful interpretation. The identification of ependymoma is of a particular significance not only in this case but in the similar cases as well because of the postoperative treatment course of patients and further evaluation.

## CASE REPORT

In this paper, we present the case of a 48-year-old female patient, who was admitted to the Centre for Neurosurgery, Clinical Centre "Kragujevac", Kragujevac, because of the spinal intradural tumor at the level of the L1 vertebral body, that is the region of filum terminale, diagnosed by means of a lumbosacral spine MRI. The lesion showed isointensity on the T1-weighted image and slightly higher signal intensity than the spinal cord on the T2-weighted image with minimal enhancement after gadolinium administration.

The patient's discomfort in terms of low back pain of moderate intensity had lasted for a couple of years before she felt the pain in her right leg five months prior to the hospitalization.There was no presence of a neurological deficit in the patient verified on hospital admission and there was no bowel or bladder dysfunction either.

After preoperative preparation the patient underwent surgery on the sixth day of hospitalization. We performed L1 right-sided hemilaminectomy, after which the strictly restricted intradural tumor was removed. The tumor was friable, its colour was gray-white and it was adherent to the filum terminale and spinal nerve. The entire tumor was removed under operative magnification with surgical microscope, the spinal nerves remained undamaged and the resection of the filum terminale was not performed.

The early postoperative course was uneventful. The patient had neither motor nor sensory deficits nor sphincter disturbances. Sutures were removed on the eighth postoperative day, after which liquorrhea occurred in the cranial part of the postoperative wound. The liquorrhea was managed by means of one secondary suture and the restriction of fluid intake.

Pathohistological analysis (HE staining and immunohistochemistry) indicated the presence of the moderate cellular tumor tissue of glial origin and solid and fascicular structure. Spindle-shaped cells formed perivascular rosettes and they were characterized by round to oval, moderately pleomorphic nuclei and grainy chromatin. In the pathohistological sample the cells showed diffuse immunoexpression of glial fibrillary acidic protein (GFAP) and individual expression of S100 protein. The lesion was characterized by the pathologists as a grade II tanycytic ependymoma according to the classification of World Health Organization (WHO) (7).

In the further course of treatment the patient was referred to physical therapy. No adjuvant radiotherapy was offered to the patient. In order to exclude the possibility that the removed ependymoma had occurred due to the liquor dissemination – MRI of the endocranium was performed during a postoperative period, showing no sure signs of expansive lesions.

Three months after the surgery, at the first control examination the patient did not claim to feel any level of pain and discomfort. Six months after the surgery a control MRI of the lumbosacral spine was performed, showing no signs of the recurrence of tumor which had previously been operated on. A control MRI is planned to be performed again in a one-year period.

## DISCUSSION

Tanycytes are specialized ependymal cells which line the floor of the third ventricle and provide structural and functional links between cerebrospinal fluid and the perivascular and neural space. They can also be found in the spinal cord and represent the common progenitor cells of both ependymal cells and astrocytes (8).

Tanycytic ependymoma is a form of ependymoma that was initially described by Friede and Pollak in 1978, who represented it as neoplasm of low-to-moderate cellularity characterized by a flow of elongated cells with moderate nuclear pleomorhism and usually absent mitotic figures (9). In these lesions, the classic ependymal rosettes and perivascular pseudorosettes are replaced by more fibrillar cells (10). Neoplastic cells usually do not exhibit anaplastic cytological features, and although it has been assigned for grade II lesions in the current WHO classification (7), it is generally a slow-growing and noninvasive tumor (11).

Just like in the case presented here, the clinical presentation correlates with the anatomic location of the neoplasm. Contrast-enhanced MR imaging remains the

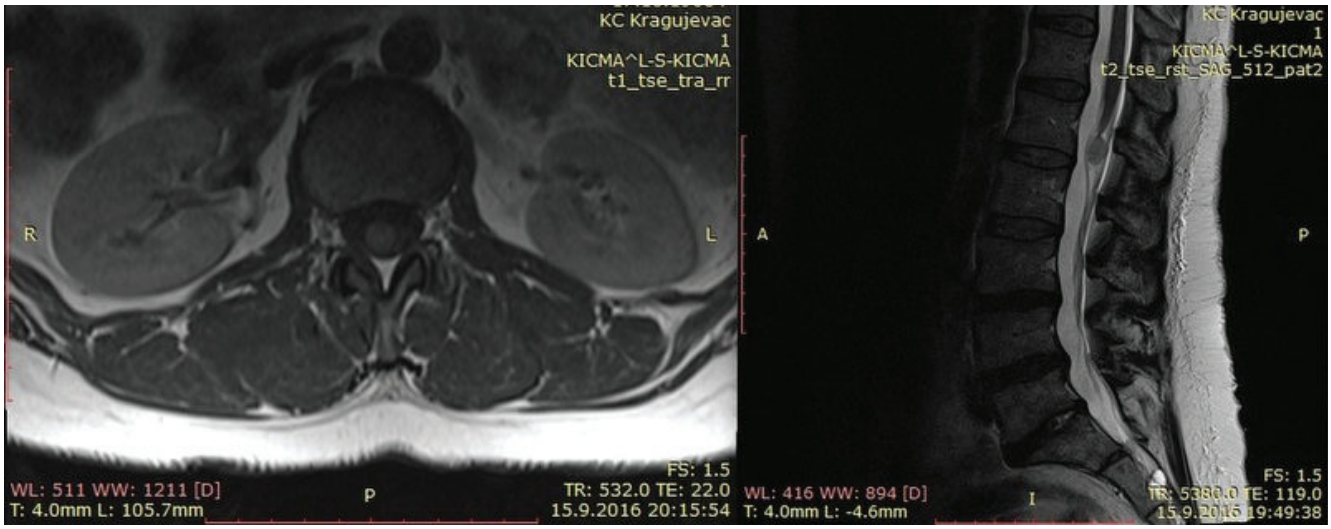


**Figure 1.** Lesion showed isointensity on the MRI T1-weighted image (1a, transversal plane) and slightly higher signal intensity than the spinal cord on the T2-weighted image (1b, sagittal plane) with minimal enhancement after gadolinium administration.

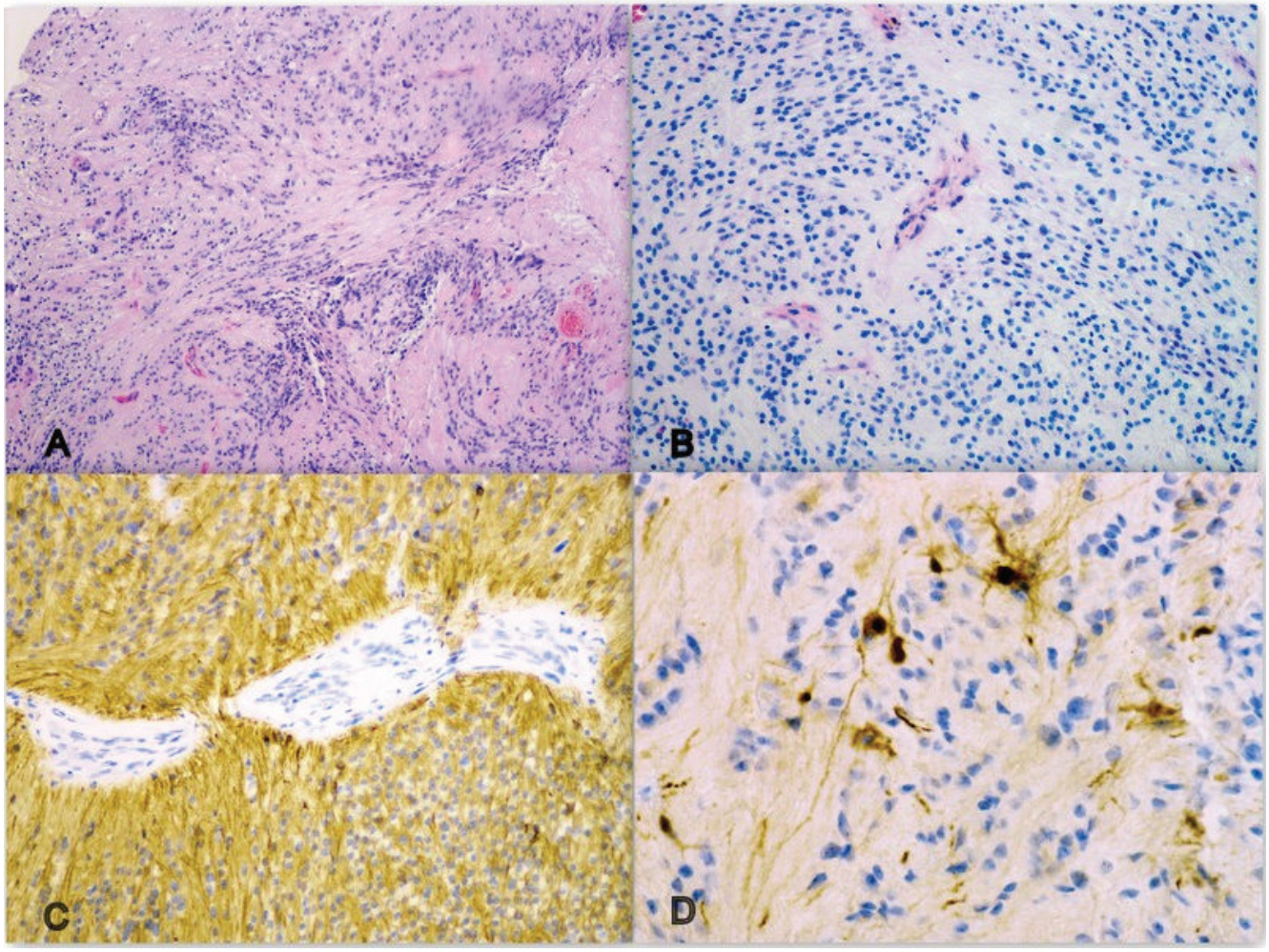


**Figure 2.**

A. HE x100 1 - The moderate cellular tumor tissue of glial origin and solid and fascicular structure.
B. HE x200 1 - The tumor cells contain round to oval nuclei with the grainy chromatin and form rare structures such as perivascular pseudorosettes.
C. GFAP x200 3 –The tumor cells show diffuse immunoexpression of glial fibrillary acidic protein (GFAP).
D. S100 x400 3 – The tumor cells show individual expression of S100 protein.

radiological investigation of choice. Due to the similar radiological picture as in myxopapillary ependymomas and cystic schwannomas, the final decision in resolving the diagnostic suspense rests with the pathologist.

Intraoperatively, the tumors have a clear cleavage in regard to neural structures but require a microneurosurgical technique for their removal. Tumors usually have minimal vascularity with cystic component, which contain dark-colored fluid. Among cases reported so far (4, 5), no increase in neurological deficits has been noted. Additionally, no tumor recurrence has been detected in the patients, which indicate favorable outcomes, without adjuvant therapy.

The low incidence of these tumors and atypical histological image, which is distinct from the typical features of commonly encountered ependymomas, can present a challenge in terms of making an accurate diagnosis. However, the presence of spindle cells, eosinofil cytoplasm, oval isomorphic nuclei and the absence of Rosenthal fibers indicate the ependymoma rather than other similar tumors (pilocytic astrocytoma, schwannoma, fibroblastic meningioma) (11). Careful immunohistochemical and ultrastructural analyses are necessary to establish the diagnosis of tanycytic ependymoma. Considering the fact that among other ependymomas in the cauda equine region the most frequent type is myxopapillary ependymoma, the diagnosis of the afore mentioned ependymoma can be excluded in this particular case due to the absence of papillary architecture and myxoid degeneration.

## CONCLUSION

Tanycytic ependymoma at the region of filum terminale is extremely uncommon and only three cases have been described in the literature. The treatment of tanycytic ependymomas should be conducted in the same way as ordinary ependymomas, since there is no current evidence suggesting that these morphologically distinct tumors differ in terms of biological behavior. A careful histological inspection with utilization of immunohistochemical stains and ultrastructural microscopy may be necessary to distinguish tanycytic ependymoma from other neoplasms such as schwannoma and pilocytic ependymoma. Awareness of this transitional form of ependymoma among neurosurgeons and pathologists may avoid incorrect surgical approaches and postoperative course.

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