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*Original Article*

# Tanycytic ependymoma in a 76-year-old Puerto Rican male

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**Abstract:** Ependymoma is a slowly growing tumor in children and young adults originating from the wall of the ventricles or from the spinal canal that is composed of neoplastic ependymal cells. Tanycytic ependymoma is a rare variant of ependymoma usually arising in the intra medullary spine. The World Health Organization classifies the tanycytic ependymoma as a grade II tumor. The diagnosis of tanycytic ependymoma is challenging since the morphology of the lesions resemble those found in schwannoma and astrocytomas. In the present study, we show a case of a 76 years old male with a progressive paraparesis for 8 years, due to a spinal tumor. Radiological and histological studies were used to classify the tumor as tanycytic ependymoma. Therefore, it is important to be aware of tanycytic ependymoma and its immunohistochemistry profile in older patients, especially within the Caribbean Hispanic population. To our knowledge this is the oldest patient known to have this rare tumor and the first case reported in Puerto Rico.

**Keywords:** Tanycytic ependymoma, Caribbean Hispanic, tumor, spinal cord, ethnicity

## Introduction

Tanycytic ependymoma is a rare variant of ependymoma that usually arises in the cavity of the spinal cord, but also in the lateral, third, or fourth ventricle. This tumor was initially described by Friede RL & Pollak A. in 1978 [1]. It was included in the 2000 WHO Classification of Tumors of the Central Nervous System as a grade II tumor [2]. Tanycytic ependymoma cells are derived from tanycytes which are special and unique ependymal cells [3]. The term comes from the Greek word tanus which means elongated. "Tanycytic" refers to its spindle elongated cell morphology [4].

This tumor closely resembles schwannoma and pilocytic astrocytoma, which makes the diagnosis more challenging and difficult [5-9]. Tanycytic ependymoma is formed of spindle fibrillary cells with oval and elongated nuclei that are similar to schwannoma, but with some peculiarities For example, schwannoma tumors have Antoni A & B cellular pattern and tend to be more cellular than tanycytic ependymoma on the other hand, Pilocytic astrocytoma has cells with a long thin hair-like process unlike tanycytic ependymoma in which the cells are more oval and wide. Those important characteristics can pass unnoticed with H&E stain, inducing a diagnostic error [3, 10].

Tanycytic ependymoma normally shows a strong immunoreactivity for glial fibrillary acid protein (GFAP) and is irregularly positive for S-100. In contrast, schwannomas are negative for GFAP and strongly and uniformly positive for S-100. Pilocytic astrocytoma, however, in addition to the piloid architecture, presents Rosenthal fibers and eosinophilic granular bodies [3, 10]. Some types of meningiomas can also resemble tanycytic ependymoma but they will present psammoma bodies and they are epithelial membrane antigen (EMA) positive and GFAP negative [10].

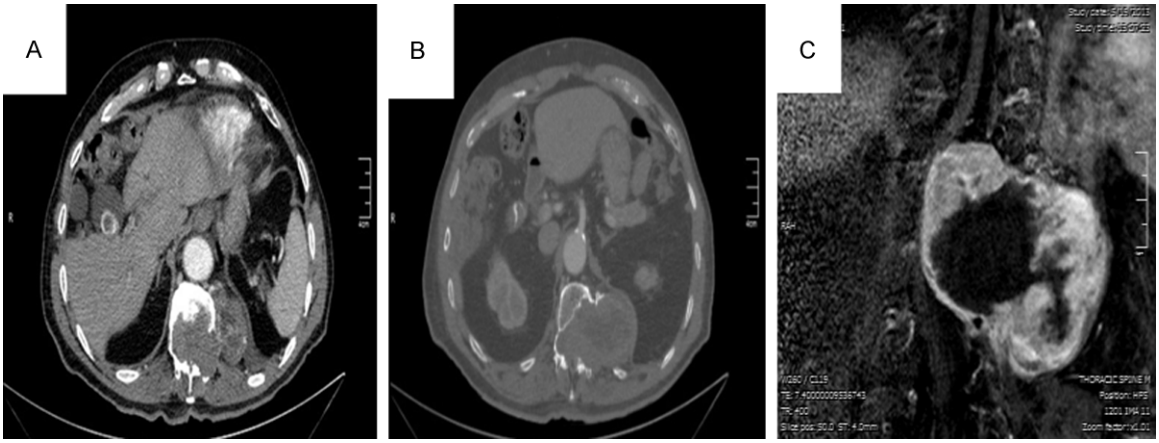


**Figure 1.** CT scan of the abdomen with intravenous contrast. A, B: Axial images in soft tissue and bone windows showed a soft tissue mass centered at the T12. C: Sagital CT scan showed that the soft tissue mass extended from T11 to L1.

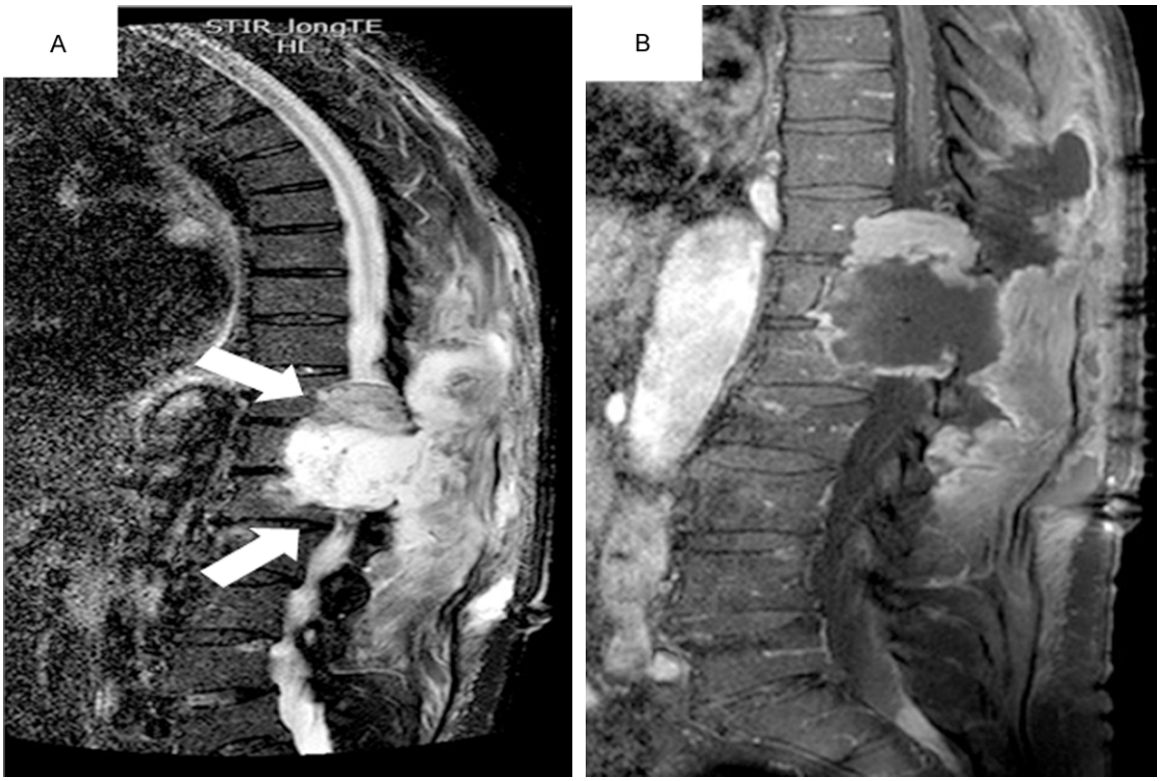


**Figure 2.** Post-operative MRI of the thoracic spine. A, B: Post-operative MRI of the thoracic spine showed large necrotic mass.

We present a rare case of a Puerto Rican 76 year-old male with progressive movement impairment in his lower extremities that became wheelchair bound. Radiological and histological analyses confirmed the diagnosis of tanycytic ependymoma. This case demonstrates that tanycytic ependymoma is a disease that could be detected in older subjects and among Caribbean Hispanics, indicating that this rare disease should be considered regardless of age and ethnic background.

## Case report

### *History and examination*

A 76 year-old Puerto Rican man, developed a progressive weakness in the lower extremities

and became wheelchair bound. He has a previous history of hypertension, myocardial infarction, coronary artery disease and hypothyroidism. The symptoms worsened and five years later, he developed severe constipation and a neurogenic bladder which required a colostomy and cytostomy, respectively. The patient was re-examined eight years after the onset of symptoms. At this time, he developed bilateral upper extremity edema, bilateral foot drop, absent patellar and Achilles reflexes and impaired ambulation. A CT scan showed a paravertebral tumor. The patient underwent a T11-L1 left laminectomy and removal of the tumor.

*CT of the abdomen with intravenous contrast*

Axial images in soft tissue and bone windows show a large soft tissue mass within the spinal canal centered at the T12 level (**Figure 1A** and **1B**). The mass measures approximately 7.2 cm × 6.6 cm × 9.6 cm (craniocaudal × anteroposterior × transverse diameters). The sagittal image shows the craniocaudal extent of the mass, within a dilated vertebral canal, spanning from T11 to L1 (**Figure 1C**). The mass remodels, erodes and severely scallops the anterior and posterior elements of the T11, T12 and L1 vertebral bodies. The core of the mass is hypodense relative to the periphery, suggesting a necrotic core. The lesion markedly expands the left T12-L1 neural foramen, extending extra-spinally along the paraspinal musculature.

*Post-operative MRI of the thoracic spine*

Coronal post-gadolinium T1-weighted sequence with fat suppression shows the large, centrally necrotic mass (**Figure 2A**). The intraspinal component markedly displaces and compresses the distal thoracic cord towards the right side. The subarachnoid space immediately above and below the lesion is expanded, suggesting an extra-medullary origin (**Figure 2A**, white arrows). The large extra-spinal component is through a markedly enlarged T12-L1 neural foramen (**Figure 1A**). The sagittal post-gadolinium T1-weighted image with fat suppression shows extensive post-operative changes in the subcutaneous and paraspinal tissues of the lower back (**Figure 2B**). The intra-spinal craniocaudal extent of the lesion is well-depicted on this image, spanning from T11 to L1. Prominent enhancing serpiginous structures along the posterior surface of the thoracic cord represent dilated spinal veins, suggesting spinal venous hypertension.

*Surgical procedure*

A laminectomy at T9-L3 was performed to expose the tumor. The lesion was identified and removed. A specimen consisting of multiple irregular fragments of pink-tan friable tissue measuring 8 × 7 × 3 cm in aggregate was sent for pathological analysis.

*Pathological findings*

Microscopically, the tumor was composed of spindle cells with a palisading pattern that resemble Verocay bodies of schwannoma and clear cells in some areas (**Figure 3A**). Also, it was detected prominent hyaline blood vessels, macrophages and few giant cells (**Figure 3B**). The tumor cells where strongly and uniformly positive for glial fribillary acidic protein (GFAP) and only scattered tumor cells were positive for S-100 (**Figure 3C** and **3D**, respectively). These findings support the diagnosis of tanycytic ependymoma.

## Discussion

Based in the review of 32 cases reported in the literature [11-43], excluding our case, tanycytic ependymoma does not have a sex or age predisposition, but is more commonly found in males, with a 55% percent of the cases being at the range of 31 to 60 years old [references] (**Figure 4A** and **4B**). These tumors are usually found in the spinal cord (19 cases of 33), with most cases allocated at the cervical area, with only nine localized in the Cerebrum and four unknown locations (**Figure 4C**). Most cases of tanycytic ependymoma were reported in Japan, followed by USA. In this case report, the patient is a 76 years old male, with a tumor in the spinal cord specifically in thoracic area T11-L1. After an intense search in the literature, we report the first case of a 76 old man of Hispanic descent with this type of tumor.

The most common symptoms of tanycytic ependymoma is the gradual deterioration of the motor functions and/or sensory disturbances, like heaviness, numbness, and weakness of the low back, lower extremities including the waist (paraparesis) accompanied with gait dis-

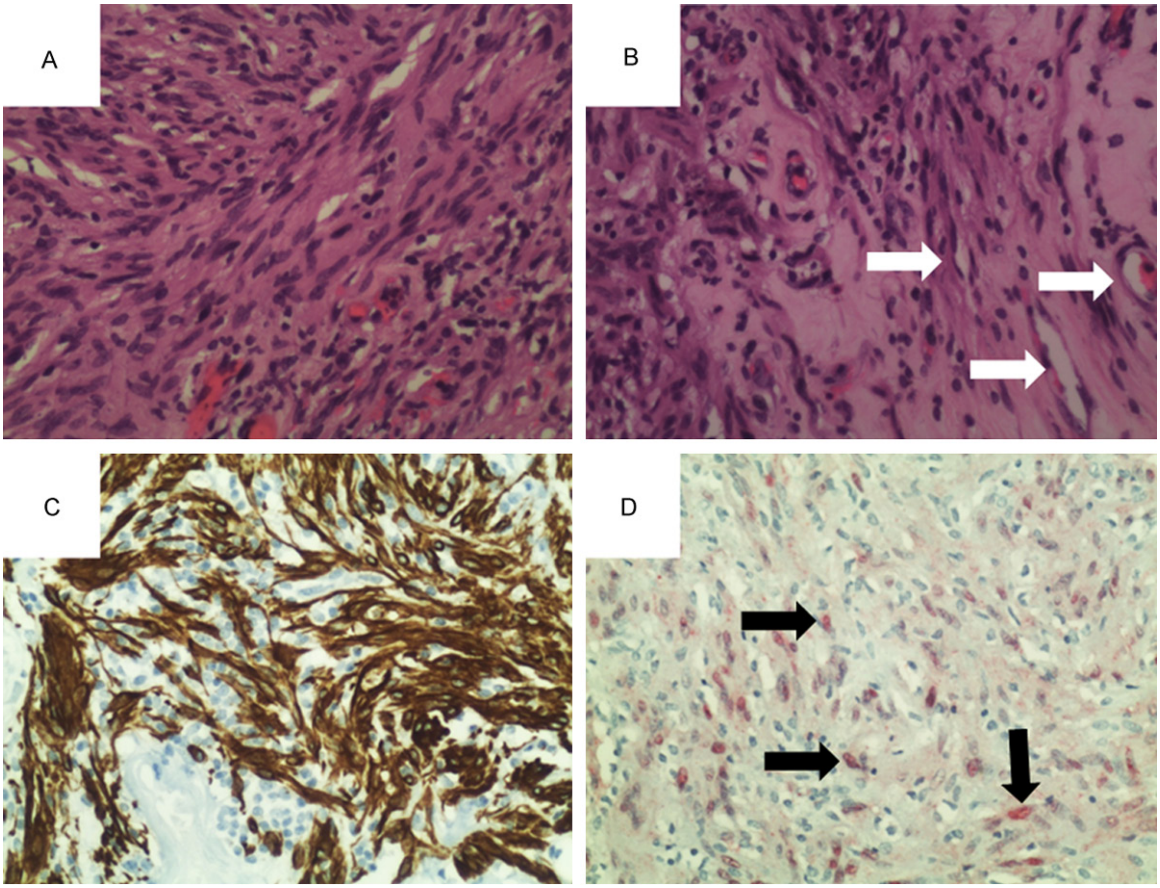


**Figure 3.** Pathological fingings. Histological analysis. A: Spindle tumor cells with a palisading pattern resembling Verocay bodies of schwannoma (H&E staining). B: Thick walled blood vessels are seen within the tumor (H&E staining). C: Tumor cells are strongly and uniformly positive for GFAP. D: Only scattered tumor cells are positives for S100. All images are at 200× magnification.

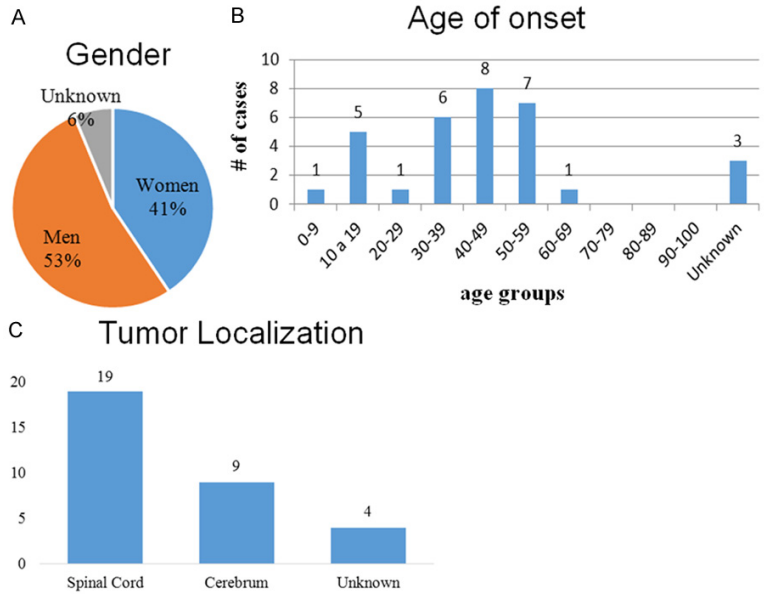


**Figure 4.** Description of reported cases. A-C: Description of gender, age of onset and tumor localization of tanycytic ependymoma cases reported in the literature.

turbance and urinary incontinence [7, 8, 11-19]. These symptoms are related to the most frequent locations of this type of tumor, the spinal cord. In the cases were the tumor is located in the cerebrum, the headache is the most common symptom [9, 20]. There is also three patients reported presenting convulsion seizures [21-23]. Tanycytic ependymoma have been reported associated to type 2 neurofibormatosis (4 cases) [8, 24, 25], Multiple endocrine neoplasia [11] and subependymoma [27]. In the case presented here, the patient developed progressive weakness of the lower extremities, causing him to

become wheelchair bound, developed severe constipation and neurogenic bladder.

In conclusion, tanycytic ependymoma is a rare neoplasia of CNS, most frequently seen in the spinal cord, with an age of onset in the 30-60 age range. This tumor can be confused with other lesions, in particular schwannoma. It is important to be aware of this variant of ependymoma and to know its morphology and immunohistochemical profile, especially in patients older than 70 years old, and of Caribbean Hispanic descent.

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