

## Case Report

### Giant Cell Ependymoma of The Filum Terminale: A Case Report

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#### ABSTRACT

Ependymomas accounts for about 2%–6% of CNS and 60%–70% of spinal cord tumors. Several histological patterns of these neoplasms are well known, but little attention has been devoted to a variant composed of giant cells. In spite of apparently “worrisome” histology, giant cell ependymoma seems to be a neoplasm with a relatively good prognosis. This report presents a case of giant cell ependymoma of the filum terminale in a 24-year-old woman and highlights the morphological diagnostic criteria for this rare tumor.

**Key words:** Ependymoma, Filum Terminale

#### Introduction

Ependymomas account for about 2–6% of CNS tumors and 60–70% of spinal cord tumors (1). They occur predominantly in children and young adults and are characterized histologically by the presence of perivascular pseudorosettes, in which tumor cells are arranged around vessels with an intervening zone consisting of thin ependymal processes directed toward the wall of the vessel, creating a characteristic pattern of halos around vessels(2). The WHO classification of ependymal neoplasms encompasses four groups: ependymoma and variants such as cellular, papillary, clear cell and tancytic ependymoma (WHO grade

II), anaplastic ependymoma (WHO grade III), myxopapillary ependymoma (WHO grade I), and subependymoma (WHO grade I)(2).

A rare variant of ependymoma is “giant cell ependymoma” (“GCE”) which has pleomorphic giant cells histologically and a good clinical outcome (3). To date only 12 cases of GCE have been documented in the literature (1, 3-6).

In the present paper, we describe a case of GCE arising from filum terminale in a 24-year-old subject.

#### Case report

A 24-year-old woman presented with a 6- month history of decreased sensation of lower extremities,

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accompanied by pain. Physical examination revealed bilateral hyporeflexia, mild paresthesia and paresis. Lumbosacral roentgenogram revealed no abnormal finding.

A sagittal, T2-weighted, magnetic resonance imaging (MRI) scan showed an oblong and isotense intramedullary lesion of filum terminale extending from L3 to L5 (Fig.1).

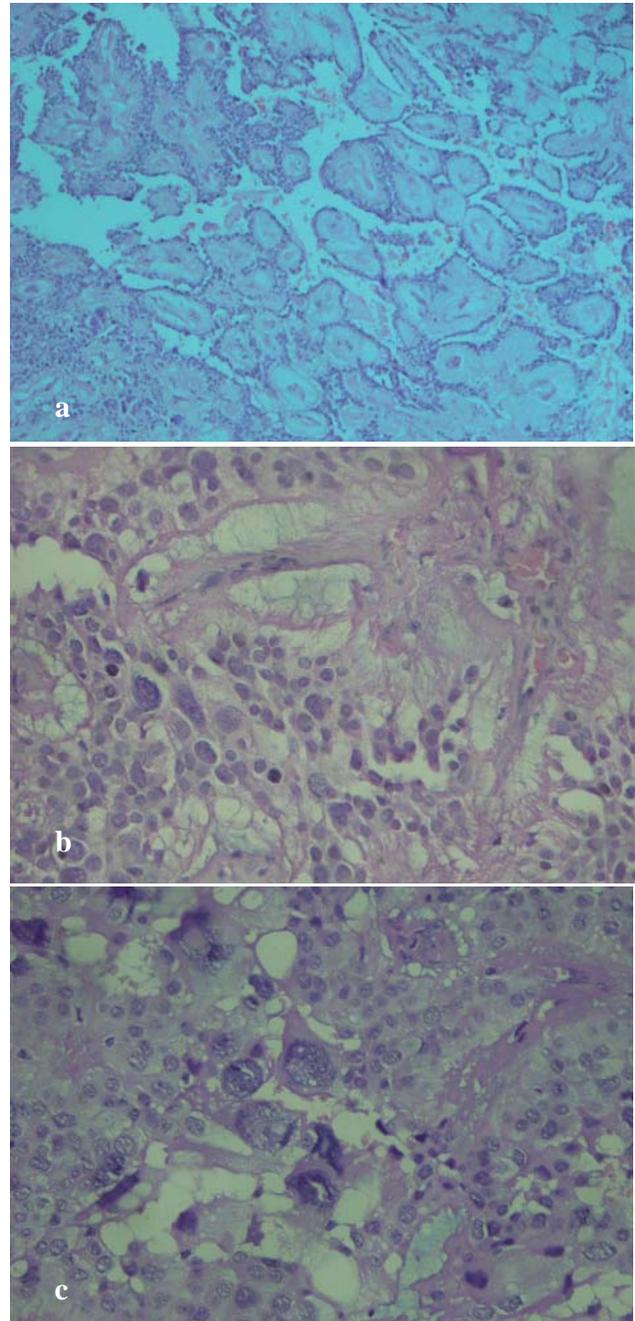


**Fig. 1:** A sagittal, T2-weighted, magnetic resonance imaging (MRI) demonstrating an oblong and isotense intramedullary lesion of filum terminale extending from L3 to L5.

The patient underwent laminectomy and a well-circumscribed mass attached to dorsal roots of spinal cord at the level of L3 to L5 was totally removed. Surgical specimen consisted of a well-defined sausage-shaped mass measured 5.5×1.5×1 cm with solid and creamy cut surface.

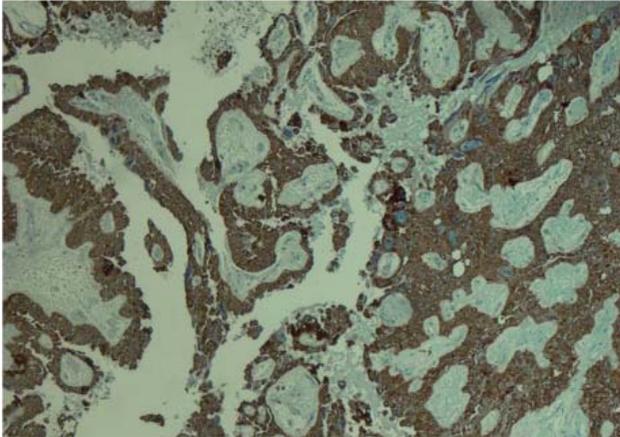
Histologically, tumor composed of papillary structures each surrounded by well-defined cuboidal or columnar cells. Dismorphism was apparent among the cellular population of neoplastic cells. A group of cells had rounded, ependymal type nuclei

with distinct margins and a delicate chromatin meshwork, and the others were pleomorphic giant cells with bizarre hyperchromatic nuclei, and abundant eosinophilic cytoplasm. The cores of the papillae consisted of central blood vessel surrounded by mucinous matrix (Fig. 2). Mitoses, necrosis, or vascular proliferation was not observed.



**Fig. 2a:** Microscopic view of tumor: papillary structures consisted of central blood vessel surrounded by mucinous matrix (H&E×40), **2b& 2c:** pleomorphic giant cells with bizarre hyperchromatic nuclei, and abundant eosinophilic cytoplasm (H&E×400)

Immunohistochemically both cellular populations strongly expressed GFAP (Fig.3) and EMA. The Ki-67 labeling index was about 1% and p53 was negative.



**Fig. 3:** Strong immunopositivity for GFAP

Considering the morphological aspect and immunohistochemical features, diagnosis of GCE of the filum terminale was made. In postoperative follow up no neurologic defect was detected.

## Discussion

Ependymomas are neuroepithelial neoplasms occurring mainly in childhood and in young adults. The majority of these neoplasms arise in association with the ependymal neuroepithelium of the ventricles and spinal central canal. Several histologic patterns of these neoplasms are well known, but little attention has been devoted to a variant composed of giant cells (3, 5).

Zec *et al.* first described two cases of GCE of the filum terminale in 1996(7). To date GCE are reported in 12 cases in the literature, majority of whom were seen in young people (3). Six tumors were localized in the vertebral canal (three arising from filum terminale(4, 7), one from cervical(8), and one from thoracic spinal cord(5), one was extended from cervical spinal cord to the fourth ventricle(8) and five intracranially(3). Our report is the fourth case of the filum terminale.

Histologically, GCE is moderately cellular tumour with a biphasic pattern. One pattern is characterized by a monomorphous proliferation of neoplastic cells with perivascular pseudorosettes

and the second is characterized by non-cohesive giant cells showing cellular pleomorphism, and considerable degenerative nuclear atypia. These cells are round or oval with eccentrically single or multiple hyperchromatic nuclei, evident nucleoli and plump eosinophilic cytoplasm (3). These nuclear changes are considered essentially degenerative in nature and probably do not presage a poor outcome (1, 7).

Mitoses and necrosis are rare and, as in the classical ependymomas, the MIB-1 or Ki-67 labeling index is low (1). In one reported case there was focal intratumoral calcification(1), but in our report, this was not the case.

The most important differential diagnosis of GCE is anaplastic ependymoma. The current WHO classification system divides ependymomas into grades II and III (the latter designated anaplastic ependymomas), but the criteria for anaplasia is a subject of debate. It has been proposed that a high mitotic rate (at least 5 mitoses/10HPF), microvascular proliferation, and the presence of areas of hypercellular, less differentiated tumor correlate best with poor outcome. Interestingly, necrosis (with the exception of pseudopalisading necrosis) is relatively common finding in ependymomas and does not have the same significance as seen in astrocytomas. Given the importance of accurate grading, it is recommended to assess ependymomas carefully for mitotic figures, including the use of Ki-67 immunohistochemical stain, especially in borderline cases (2).

Other differential diagnoses are clear cell ependymoma, anaplastic oligodendroglioma, rhabdoid/papillary meningioma, pleomorphic xanthoastrocytoma, subependymal giant cell astrocytoma and giant cell glioblastoma(3,9). Considering some features like the presence of bizarre and monstrous cells in ependymomal background with perivascular rosettes and absence of mitotic activity, necrosis and microvascular proliferation (glomeruloid) accompanied by immunohistochemical profiles can guide to correct diagnosis (3).

One must be particularly cautious in the cases in which limited biopsy samples have been obtained. There is also special limit for intraoperative diagnosis of this tumor as in one case intraoperative

analysis of frozen section tissue fragments suggested a malignant tumor (1, 5). Considering the surgical anatomical features of the mass and relatively ease of its dissection off the spinal cord can prevent of this error.

As the number of reported cases is limited, it is difficult to predict the biological behavior of this kind of ependymoma (3).

There are reported cases of GCE with anaplastic features, manifested by cellular pleomorphism, high cellularity, intensive mitoses with or without microvascular proliferation and necrosis (5, 9). These cases have been recurred within a year after surgery (3). Some reports suggested the role of p53 mutation or overexpression in anaplastic transformation in GCE, which correlated with poor prognosis (10). However, in other cases with gigantocellular morphology and marked pleomorphism but without features of anaplasia (such as our case), the course of disease was benign (3).

### Acknowledgements

The authors declare that they have no conflicts of interest.

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