

## Case Report

# Multicentric extramedullary myxopapillary ependymomas: Two case reports and literature review

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## Abstract

**Background:** Ependymoma has been described typically as an intramedullary tumor derived from ependymal cells. Intradural extramedullary presentation is rarely described and almost always as a unique lesion. Myxopapillary ependymoma is a histological variant that distinguishes from the ordinary type of ependymoma because of its generally better prognosis. We present two cases of multicentric extramedullary myxopapillary ependymomas.

**Case Description:** Case 1 was a 30-year-old man with progressive paresthesia and paresis in the lower limbs, urinary sphincter disturbances, gait instability, ataxia, and chronic low back pain with multiple intradural extramedullary lesions at C2-C3, D2-D4-D5, and D12-L1. Case 2 was a 32-year-old man, presented with low back pain and mild paresthesia in the right lower limb. Magnetic resonance imaging (MRI) showed multiple intradural extramedullary lesions with homogeneous enhancement after gadolinium injection at C7, D2, D4, D5, D8, D10, D11, L1, L3, L5, S1, and S2. Complete tumor resection of the approached tumors was archived in both cases. Histological studies confirm myxopapillary ependymomas. Patient's neurologic outcome was good and no residual tumor was present at MRI control at 10 years in case 1 and 12 months in case 2.

**Conclusions:** We report the first two cases of multicentric extramedullary myxopapillary ependymomas, this etiology must be taken into account in the differential diagnosis of intradural extramedullary tumors.

**Key Words:** Extramedullary ependymoma, multicentric ependymoma, myxopapillary ependymoma

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## INTRODUCTION

Ependymoma has been described typically as an intramedullary tumor derived from ependymal cells. Their location is most commonly proximal to the normal

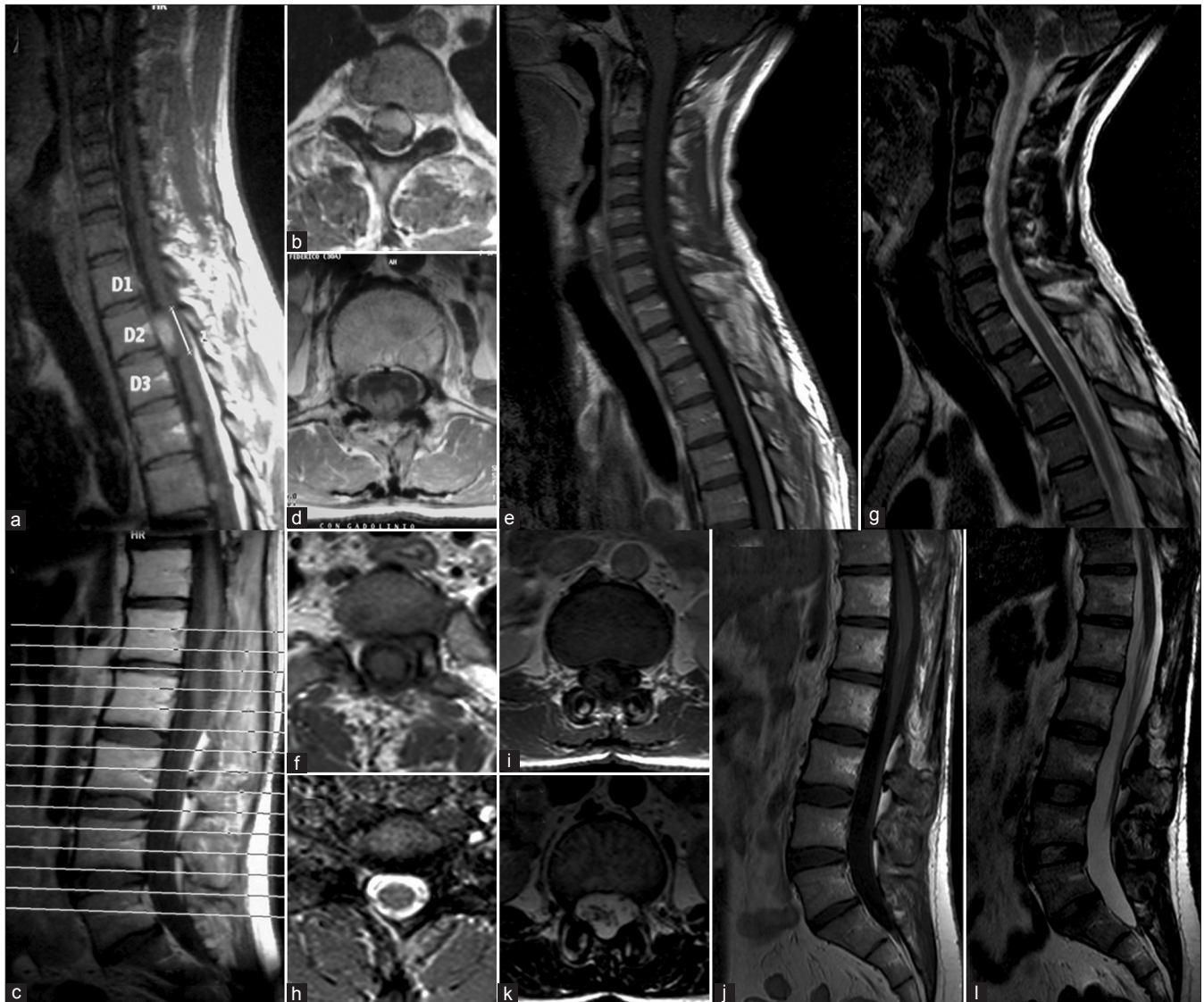
site of these cells. Intradural extramedullary presentation is rarely described except for those located at the terminal filum or conus medullaris. Myxopapillary ependymoma is a histological variant distinguishes from the ordinary type of ependymoma because of its generally better

prognosis<sup>[2]</sup> [Table 1]. The authors present two cases of this unusual location and histological variant, and discuss the incidence, histopathogenesis, and treatment of this rare clinical presentation.

## CASE I

A 30-year-old man was referred to our neurosurgical department presenting progressive paresthesia and paraparesis, urinary sphincter disturbances, gait instability, ataxia, and chronic low back pain. Neurological examination showed bilateral hypoesthesia at D2 and perineal region was not associated with radicular pain and hyperreflexia in lower-limbs. Spinal magnetic resonance imaging (MRI) revealed multiple intradural extramedullary lesions, isointense on T1, hyperintense

on T2, and fluid attenuated inversion recovery (FLAIR) sequences with intense contrast enhancement, at C2-C3, D2-D4-D5, and D12-L1 [Figure 1 shows no recurrence in T2. I, J, K and L, demonstrate no tumor at D12-L2 level in 10-year postoperative control"]. Cranial MRI did not reveal any abnormality. The D12-L1 tumor was located posteriorly in the spinal canal displacing anteriorly the conus medullaris and was removed through a D12-L2 laminectomy in first place. The D2 antero-medullary mass compress and displace posteriorly the spinal cord. After D1-D3 laminectomy and midline durotomy the left dental ligament was cut and retracted to improve surgical field. The tumor has soft consistence and a fleshy-gray appearance and was completely encapsulated by a thin fibrous capsule with a well delimited cleavage plane. A microvascular attachment to the spinal cord



**Figure 1:** (a and b) Demonstrate D2 antero-medullary enhanced tumor on T1 sequence. (c and d) Show D12-L1 lesion posteriorly located in the spinal canal displacing anteriorly the conus medullaris. (e and f) 10-year postoperative control MRI showing no recurrence on T1. (g and h) Shows no recurrence in T2. (i, j, k and l) demonstrate no tumor at D12-L2 level in 10-year postoperative control

was identified, coagulated, and divided. The tumor was debulked and completely resected using microsurgical techniques. Histological results inform a myxopapillary ependymoma. Patient's neurologic condition improved after surgery and he was discharged with a mild lower limb paresis and D2 left radicular pain, which resolved with rehabilitation 6 months after treatment. Adjuvant radiotherapy was performed and he had no tumor progression in 10 years follow up.

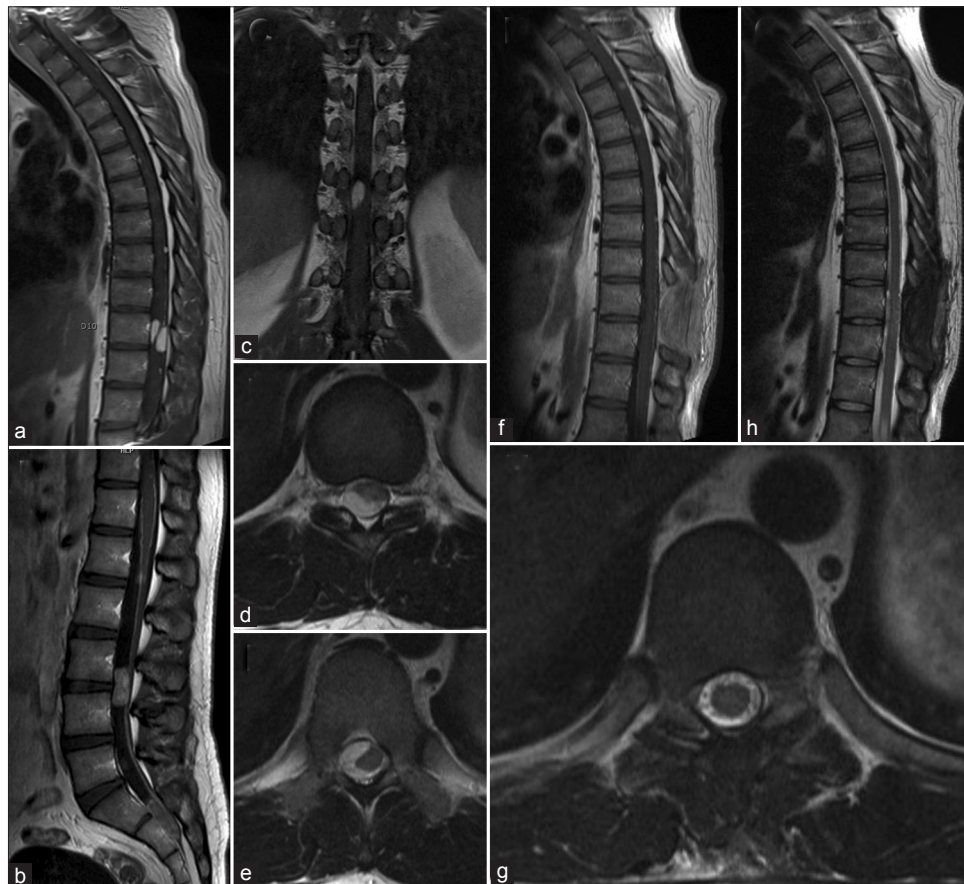
## CASE 2

A 32-year-old man, presented with a 6-month history of low back pain and mild paresthesia in the right lower limb without dorsalgia or radicular pain. Neurological examination showed bilateral hypoesthesia at the T-8 level and hyperreflexia in lower limbs (right greater than left). MRI showed multiple intradural extramedullary lesions with homogeneous enhancement after gadolinium injection at C7, D2, D4, D5, D8, D10, D11, L1, L3, L5, S1, and S2 [Figure 2]. D10 tumor was "C" shaped occupying 60% of the spinal canal without foramen invasion or bone destruction. The tumor compresses anteriorly, right-lateral and posteriorly the spinal cord.

Cranial MRI was normal. The tumor was approached through a D9-D10 laminectomy. Surgical findings include partially encapsulation, soft discretely hemorrhagic consistence with a well delimited cleavage plane, and no medullary connection. The D10 right root was involved by the tumor but not infiltrated. After debulking complete tumor removal was archived under microscope magnification. Histological tumor diagnosis was also myxopapillary ependymoma [Figure 3]. Referred symptoms remain unchanged and control MRI at 12 months after surgery showed no recurrence or spinal cord damage. No adjuvant therapies were performed.

## DISCUSSION

Extramedullary ependymomas appears to be more frequent in the 5th decade of life<sup>[3]</sup> and females and only 20 cases of this unusual location have being described in literature.<sup>[3-10]</sup> The myxopapillary histological variant has a slight male predominance and a peak age in young adults at 3<sup>rd</sup> decade.<sup>[3]</sup> Myxopapillary ependymomas often arise from filum terminale and are encountered in the conus medullaris area, this location "technically speaking" is also extramedullary, and the subpial



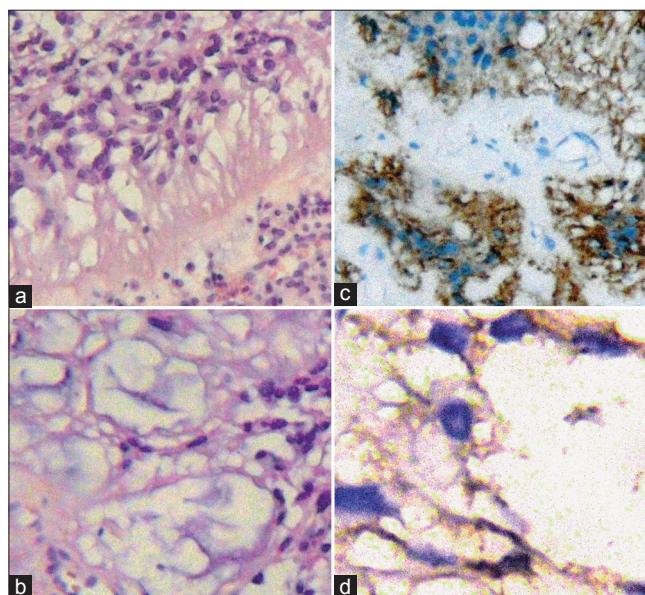
**Figure 2:** (a and b) Shows sagittal multiple spine enhanced tumours. (c) Demonstrate coronal view of D10 tumor on T1-weighted sequence. (d and e) Presents axial view of "C" shaped D10 lesion. (f) Present 12-month postoperative T1-weighted contrast sequence. (g and h) Demonstrate 12-month postoperative control on T2-weighted images



medullary location is the result of its cerebrospinal fluid (CSF) seeding.<sup>[10]</sup> So far only one case was reported in a focal extramedullary location<sup>[5]</sup> besides the conus and filum terminale. It has been postulated that intradural extramedullary ependymomas develop from ectopic ependymal cells that migrates from the neural tube to the extramedullary space during its closure,<sup>[12]</sup> this neuroaxis invagination is evidenced by the tumor encapsulation of the pia mater and arachnoid membrane in almost all cases.<sup>[3]</sup> Some authors describe CSF dissemination as a result of aggressive tumor behavior<sup>[10]</sup> or anaplastic transformation of initial benign lesions.<sup>[17]</sup> Although most of these extramedullary tumors were described in a single level, Iunes<sup>[11]</sup> described the first report of a multifocal intradural extramedullary grade II ependymoma. This spread could be the result of drops metastasis from these primary lower spinal lesions. We present two cases of this extremely rare multicentric dissemination of a grade I, myxopapillary ependymomas.

Histologically, ependymal rosettes and perivascular pseudorosettes that characteristically mark ependymomas are generally absent or are not well formed in the myxopapillary subtype, although mucoid stroma, which is a distinctive feature of myxopapillary ependymoma, is not observed in classic ependymomas or subependymomas. Parietal hyalinization of vessels is frequently seen in myxopapillary subtypes. Mitotic figures, calcifications, necrosis, and cyst formations are not usually present [Table 1]. Myxopapillary ependymoma is histopathologically benign, grade I in the World Health Organization (WHO) classification, however, malignant transformation can occur, leading to recurrence or

intradural dissemination.<sup>[10,14-20]</sup> Outcome is strongly influenced by the extent of surgical resection, which is the best predictor of good prognosis. Sonneland *et al.*<sup>[21]</sup> reported a mean survival of 19 years in patients who underwent gross total resection as compared with 14 years for patients who underwent subtotal resection. In the case of multilevel compromise, asymptomatic lesions of this histopathological variant can be observed at appropriate time intervals due to lack or slow growth during follow-up as shown in Case 1. Otherwise,



**Figure 3:** (a) Neoplastic proliferation of ependymal cells, eosinophilic cytoplasm and hyperchromatic nuclei with mild anisokaryosis and papillary structures. (b) Abundant myxoid matrix. (c and d) Immunohistochemistry were performed with antigen retrieval methods informing positive stain to GFAP and S-100 protein

**Table 1: Demographic, histological and prognosis characteristic of ependymomas**

	Ependymoma	Ependymoma myxopapillary	Subependymoma	Anaplastic Ependymoma
Peak age	Children	Young adults	Adults	Children > Adults
Gender	Males = Females	Males ≥ Females	Males > Females	
Ependymal rosettes	+	-	-	+
Perivascular pseudorosettes	+	-	-	+
Mucoid Stroma	-	+	-	-
Papillary architecture	-	+	-	-
Cellularity	+++	+	++	++++
Mitoses	±	± (rare)	±	+
Calcification	±	-	±	+
Necrosis	±	-	±	±
Nuclear pleomorphism	±	±	±	±
Vascular proliferation	-	-	-	±
S-100	+	+	+	+
GFAP	+	+	+	+
Prognosis	Dependent on extent of resection	Good	Good	Worse

rare cases of aggressive behavior have been reported involving dissemination within and outside the central nervous system.<sup>[22,23]</sup> A close follow-up is recommended because of the potential for anaplastic transformation, recurrence, or metastasis.<sup>[13]</sup> The utilization of adjunctive radiotherapy has shown a significant improvement in overall survival even posterior to gross total resection.<sup>[1,21]</sup> In our 1 case we use it, presenting no tumor recurrence in 10 years.

## CONCLUSIONS

We report the first two cases of multicentric extramedullary myxopapillary ependymomas. This etiology must be taken into account in the differential diagnosis of intradural extramedullary tumors, a complete imaging study of the entire neuroaxis should be performed always in order to exclude the presence of tumor dissemination. In multifocal presentation, asymptomatic lesions of this histopathological variant may be observed at appropriate time intervals due to lack or slow growth.

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