

A RARE CASE OF EXTRA-INTRAMEDULLARY DORSAL TANYCITIC EPENDYMOMA, RADICALLY REMOVED WITH INTRAOPERATIVE NEUROPHYSIOLOGICAL MONITORING

GIUSEPPE CICERO², ANTONELLA GIUGNO¹, ROSARIO MAUGERI¹, FRANCESCA GRAZIANO¹, LUIGI BASILE¹, DOMENICO G. IACOPINO¹

¹Department of Experimental Biomedicine and Clinical Neurosciences, School of Medicine, Neurosurgical Clinic, University of Palermo, Palermo, Italy - ²Department of Surgical, Oncological and Oral Sciences, School of Medicine, Neurosurgical Clinic, University of Palermo, Palermo, Italy

ABSTRACT

Introduction: *Tanycytic dorsal extra and intramedullary ependymoma is a rare form of tumor.*

From the histological point of view, these tumors show several aspects that make difficult the differential diagnosis from schwannomas and pilocytic astrocytomas. Tanycytic variant, often occurs in the thoracic tract of the spinal cord, and it is constituted by tanycytes, that are typical elongated and bipolar cells that give to the tumor fibrillary aspects. Tanycytic variant has been recently characterized as a variant of ependymoma, since the 2000 World Health Organization (WHO) system.

Case presentation: *A 57 years old woman presented with intractable back pain often radiating to the left leg. Neurological exam revealed mild weakness in left tight flexion. No sensory or sphincterical disturbances were present. A dorso-lumbar Magnetic Resonance Imaging (MRI) with contrast medium showed a well-demarcated T12 intradural extramedullary lesion, suggestive for schwannoma. The tumor was radically removed, with an excellent neurological outcome, and was then characterized as a grade II tanycytic ependymoma.*

Conclusion: *To differentiate the diagnosis between extramedullary ependymomas and schwannomas, meningiomas or astrocytomas is necessary a histopathological examination and a close follow up period is recommended since the tumor could evolve into higher grade. Neurophysiological monitoring is necessary for a satisfactory neurological outcome.*

Key words: *Intraoperative neurophysiological monitoring, Extramedullary ependymoma, Tanycytic ependymoma; Differential diagnosis.*

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Introduction

Spinal cord ependymomas are a subgroup of spinal gliomas, originating from cells lining of central canal of the spinal cord. They are slow growing tumors, typically benign intradural intramedullary lesions, and they account for about 40-60% of all primary spinal cord tumors⁽¹⁾. They are also the most common primary intramedullary spinal cord tumors in adult, sometimes localized also in the filum terminalis, being very rare the coexistence of an extramedullary component⁽²⁾. The histopathological classification individuates myxopapillar ependy-

mas and subependymomas that are grade I World Health Organization (WHO) ependymomas (grade II WHO), and grade III WHO anaplastic ependymomas⁽³⁾. Tanycytic variant, often occurs in the thoracic tract of the spinal cord, and it is constituted by tanycytes, that are typical elongated and bipolar cells that give to the tumor fibrillary aspects. Tanycytic variant has been recently characterized as a variant of ependymoma, since the 2000 WHO system⁽⁴⁾. From the histological point of view, these tumors show several aspects that make difficult the differential diagnosis from schwannomas and pilocytic astrocytomas⁽⁴⁾.

According to the available literature, there are only 18 reported cases of primary intradural extramedullary ependymomas (IDEM), which are exceptional outside the filum terminalis. Here we describe an extremely rare case of grade II tanyctic intradural extramedullary D12 ependymoma, successfully treated in a radical manner with the aid of intraoperative neurophysiological monitoring. In this case report, we have reported a rare case of tanyctic grade II dorsal extra and intramedullary ependymoma.

Tanyctic variant ependymoma was firstly described in late 70's by Friede and Pollak⁽⁵⁾, that reported 11 cases, 2 supratentorial, 3 infratentorial and 6 spinal^(3,5). However, it has been definitively characterized only in the 2000 WHO classification.

It takes origin from spindle shape bipolar cells, tanyctes, name coming from Greek meaning "to stretch", that usually are found lying along the ependymal wall of the third ventricle. These cells can resemble astrocytes but they differ from astrocytes for a characteristic "salt and pepper" aspect of the nucleus; furthermore, tanyctes do not form rosettes and perivascular pseudorosettes in a similar manner to schwannomas. Therefore, immunohistochemistry demonstrate positivity for Glial fibrillary acid protein (GFAP), S100 protein and Epithelial-Membrane Antigen (EMA) only in tanyctic ependymomas, whereas astrocytomas stain for GFAP and OLIG2 (a member of the group of basic helix-loop-helix transcription factors). Otherwise, schwannomas shows positivity for collagen type IV and Laminin unlike tanyctic ependymomas and astrocytomas.

Spinal tanyctic ependimomas often involve the cervico-thoracic region when exclusively intramedullary, whilst extra and intramedullary tumors are frequently found in the lower thoracic tract⁽⁴⁾. Sometimes they are associated with a syrinx, especially when located in the conus medullaris. From the analysis of previous literature, only 17 spinal tanyctic ependymomas are reported. Of all these, only two showed extramedullary localization, one of them totally removed. According to previous literature, after radical resection of spinal tanyctic ependymomas, no chemio or radiotherapy is required. Moreover, even if low grade tumors, it has been described anaplastic transformation in three previous reports^(6,7,8).

Case presentation

A 57-year-old woman was admitted to Department of Neurosurgery with a 2-year history of progressive paraparesis. For several years she had experienced pain radiating into both legs. For almost a year she had been unable to walk, even with support. The neurologic examination revealed a severe paraparesis, loss of sensory from T12 level, no bowel or bladder incontinence. No sensory or sphincter disturbances were present. The patient underwent a dorso-lumbar Magnetic Resonance Imaging (MRI) with contrast medium that showed a well-demarcated intradural extramedullary lesion, extending in the cranio-caudal direction for about 4 cm, with inhomogeneous intensity in both T1 and T2 sequences and evenly enhancing after contrast medium, extended also to the adjacent dura. The lesion caused bone erosion of the posterior part of D12 and enveloped the correspondent left nerve root with enlargement of the conjugation foramen, causing also dislocation of the conus medullaris on the right (Fig. 1).

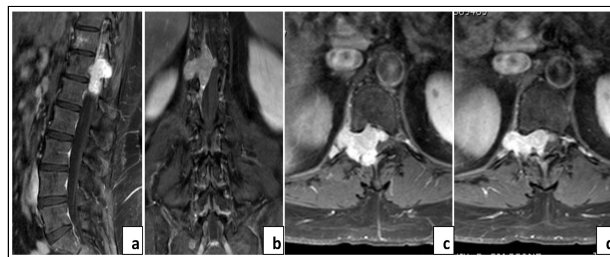


Figure 1: Pre-operative dorso-lumbar MR images. T1 weighted Gd-enhanced **a)** sagittal, **b)** coronal and **c), d)** axial reconstruction showing the massive enhancement of the intradural extramedullary dumbbell tumor causing dislocation of the conus medullaris.

For this morphological behavior, and in particular in relation to foramen invasion, the first hypothesis was of a D12 root schwannoma or, less probably, a meningioma. With the aid of intraoperative neurophysiological monitoring, a posterior dorso-lumbar approach was performed centered on D11-D12-L1 and L2 spinous processes. After D12 and L1 spinosectomy, a left D12 laminectomy allowed the visualization of a greyish lesion, quite bloody, with elastic consistence that partially infiltrated the adjacent bone and the ligamentum flavum.

The tumor appeared as a unique piece with the nerve root. It was followed along its course until completely exposed. Vascular supplies to the lesion were coagulated and interrupted.

The capsule was bluntly dissected and then opened. Internally the tumor showed a soft yellow aspect and was debulked partially in small fragments and partially with the ultrasonic surgical aspirator. The stimulation of the involved root did not reveal evoked response to motor potential, so the root could be sacrificed allowing the radical resection of the tumor. After internal debulking, the remaining part of the lesion was removed en-bloc from the foramen. The other vascular and nervous structures were respected and then the dura was repaired with autologous fibrin glue and watertight suture.

Finally, a D11-D12-L1-L2 arthrodesis with titanium pedicular screws was performed to ensure adequate stabilization of the vertebral tract operated. Postoperative dorsal MRI with contrast medium confirmed the treatment was radical, and a dorsal Xray and Computed tomography scan (CT scan) documented the right positioning of the screws. (Fig. 2).

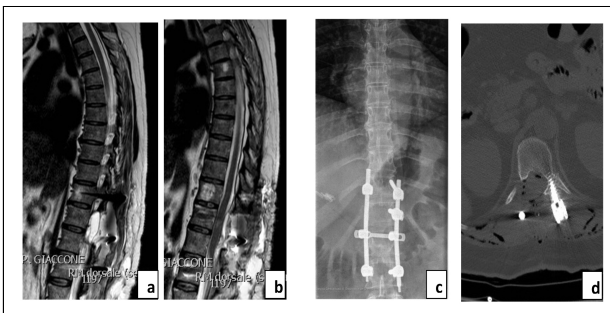


Figure 2: Post operative Neuroimaging. **a),b)** Dorso lumbar MR T2 weighted sagittal reconstruction showing CSF collection and complete removal of the lesion. **c)** Dorso-lumbar Xray showing the posterior transpedicle fixation. **d)** Ct bone window axial reconstruction showing ipsilateral arctrectomy and controlateral pedicle screw.

No postoperative neurological deficits were encountered and the patient referred a progressive recovery of the motor function of the left leg and remission of back pain.

Anatomic-pathologic evaluation showed a mesenchymal neoplastic proliferation, constituted by hypercellular spindle shape elements with hyperchromic and prominent nucleoli, alternated to hypocellular areas in a loose stromal matrix. Immunohistochemical examination documented immunopositivity for S100, GFAP, and Vimentin. Furthermore there was no staining with p53, CD34, Cd57, Calretinin. The Ki-67 index was 3-4%. Taking together these data, the tumor was characterized as a WHO grade II tanyctic ependymoma.

No adjuvant postoperative therapies were needed.

Long-term neurological outcome was excellent with complete recovery of left leg strength and no late recurrence of the tumor was encountered.

Discussion and conclusion

Spinal ependymomas are tumors usually located in extramedullary space and in the filum terminalis. Here we described an extremely rare case of intradural extramedullary D12 tanyctic ependymoma, radically and successfully operated with the aid of neurophysiological monitoring.

Intraoperative neurophysiological monitoring is widely used from 2000's to improve spinal surgery safety and reduce the incidence of neurological complications. To date, there are few studies concerning the real indications and possible benefits from this procedure in spinal surgery⁽⁹⁾. Usefulness is proven for scoliosis surgery, while less strong evidence is documented for tumoral epidural compressions⁽¹⁰⁾. According to a metanalysis of the literature from 1990 and 2009 by Fehlings et al.⁽¹¹⁾, multimodality intraoperative neuromonitoring is recommended in all surgery treatments "where the spinal cord or nerve roots are deemed to be at risk, including procedures involving deformity correction and procedures that require the placement of instrumentation".

In fact, it reliably detects intraoperative spinal or nerve injuries, also reducing the rate of postoperative neurological deficits⁽¹¹⁾. Otherwise, Zielinski et al.⁽¹²⁾ evaluated the effectiveness of neurophysiological monitoring in 74 patients underwent to spinal surgery (extradural, intradural extramedullary and intradural intramedullary), 38 of them operated with intraoperative monitoring. The number of neurological complications was significantly lower only in the intramedullary group, while similar data were encountered in the other groups comparing patients treated with and without neurophysiological monitoring.

Of all 18 previous reported extramedullary ependymomas, 16 were radically removed. Only one of them presented complete neurological recovery.

This reported, is the second case of totally removed extramedullary tanyctic ependymoma, the first described in literature with intraoperative neurophysiological monitoring. No post-operative neurological deficits were encountered, with com-

plete neurological recovery and after one year there was no recurrence. A multidisciplinary team, with specific expertise, is necessary.

This case report allows us to understand that the histopathological examination is necessary for a precise discrimination from schwannomas, meningiomas and astrocytomas and close follow up period is recommended since the tumor could evolve into higher grade⁽¹³⁾ moreover is recommended in a close follow -up period because the tumor could evolve into a higher grade. Neurophysiological monitoring is necessary for a satisfactory neurological outcome.

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Corresponding author

PROF GIUSEPPE CICERO, MD, PhD
 Department of Surgical, Oncological and Oral Sciences,
 University of Palermo, Palermo
 Medical Oncology
 Via del Vespro 129
 90127 Palermo
 (Italy)