



## Case Report

# Spinal intradural extramedullary granular cell tumor

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## ABSTRACT

**Background:** Granular cell tumors (GCTs) are uncommon peripheral nerve sheath tumors of Schwann cell origin that may occur throughout the body. However, they rarely occur in the spinal canal.

**Case Description:** A 49-year-old male presented with burning sensation in the left knee. The MRI of the lumbar spine showed an L3-L4 intradural extramedullary tumor. Complete surgical resection was successfully performed and the L3 root burning improved. Histopathologically, the lesion proved to be a benign GCT.

**Conclusion:** Spinal GCTs are rare benign tumors that may be found in an intradural extramedullary location in the spine. The preferred treatment is complete surgical resection as subtotal/partial resection may result in recurrence warranting radiation therapy.

**Keywords:** Granular cell tumor, Intradural extramedullary, Intramedullary, Nerve sheath tumor, Spinal canal

## INTRODUCTION

Granular cell tumors (GCTs) are uncommon soft-tissue tumors that were initially described as granular cell myoblastoma by Abrinkosoff in 1926.<sup>[15]</sup> They most commonly occur in the deep dermis and subcutaneous tissue of the head-and-neck region, trunk, and proximal extremities.<sup>[18]</sup> They may also arise in visceral sites such as the gastrointestinal and respiratory tracts.<sup>[18]</sup> GCTs of the spinal canal are extremely rare with only few cases reported in the literature. We report a case of a 49-year-old man who was found to have an intradural extramedullary GCT at the L3-L4 level.

## CASE DESCRIPTION

A 49-year-old male presented with burning sensation and pain in the left knee for 2 years' duration. The pain was severe and refractory to pain medications. The lumbar MRI showed a well-circumscribed soft-tissue lesion in the left L3-L4 neural foramen along the course of the left L3 nerve root. It was isointense to hypointense on T1W, hypointense on T2W images [Figure 1] and enhanced with contrast.

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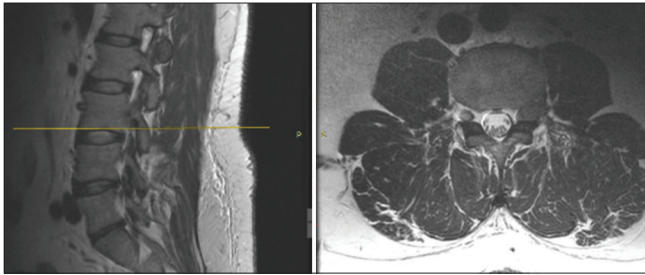
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## Surgery

Under fluoroscopic guidance, a bilateral L3-L4 laminectomy was performed. The pedicles of L3 and L4 were exposed along with the transverse processes. The traversing L3 nerve root was identified and the tumor was completely dissected and resected off the root. Postoperatively, the patient sustained improvement in the burning sensation and exhibited no new motor deficit.

## Histology

The histological examination was consistent with a benign GCT. It revealed polygonal tumor cells with abundant, eosinophilic, and granular cytoplasm arranged in sheets [Figure 2a]. There was no mitotic activity, necrosis, marked



**Figure 1:** Lumbo-sacral spine MRI T2 sequence showing soft tissue lesion at the level of L3-L4 (yellow bar).

pleomorphism, or tumor spindling. Immunohistochemically, the tumor cells were diffusely positive for S-100, SOX-10, and inhibin [Figures 2b-d] and showed patchy positivity for CD68. They were negative for CD34 and EMA. In addition, the Ki-67 proliferation index was approximately 5%.

## DISCUSSION

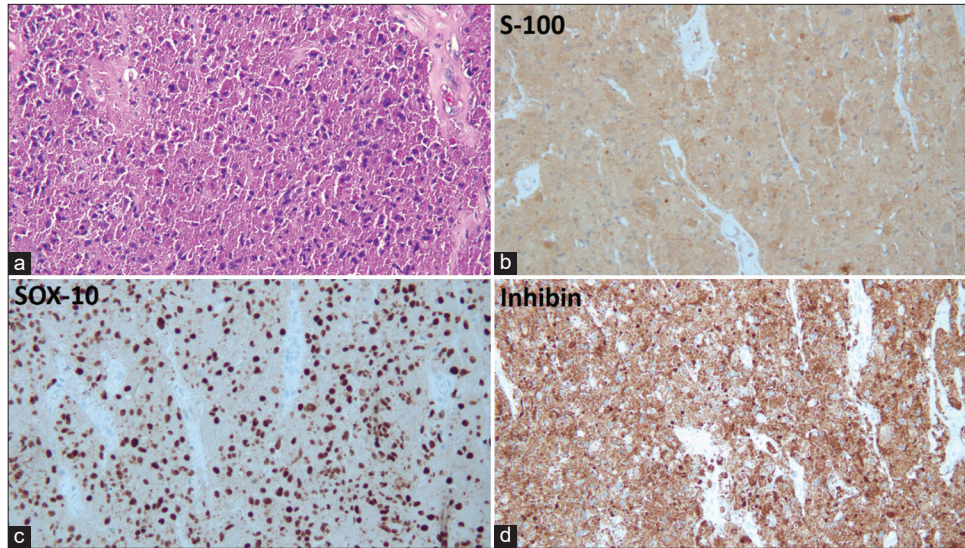
GCTs are peripheral nerve sheath tumors that show neuroectodermal differentiation.<sup>[18]</sup> They are believed to be of Schwann cell origin based on immunohistochemical and ultrastructural findings.<sup>[17]</sup> They may occur throughout the body, with the tongue being the most common site accounting for 25% of cases.<sup>[18]</sup> Despite their Schwann cell derivation, GCTs of the spinal canal are very rare. Review of the English literature revealed 20 cases of spinal intradural GCTs [Table 1].<sup>[1-17,19]</sup> The age of the patients ranged from 9 to 49 years, with the exception of one case that was reported as an incidental autopsy finding at the age of 73. The mean age at presentation is 23 years. There is a female predominance where 16 out of 21 patients (76%) are female. GCTs can be seen throughout the spinal canal: six of the reported cases were in the cervical region, four cases in the thoracic region, three cases in the thoracolumbar region, seven cases in the lumbar region, and one case in the sacral region.

The majority of the reported cases of spinal GCTs have an intradural extramedullary location. The differential

**Table 1:** Summary of the reported cases of spinal intradural granular cell tumors.

Case report	Age	Sex	Spinal level	Extent of resection	Recurrence	Additional treatment
Markesbery <i>et al.</i> , 1973 <sup>[8]</sup>	73	F	C1. EM	Autopsy finding	Autopsy finding	Autopsy finding
Stromblad <i>et al.</i> , 1987 <sup>[14]</sup>	10	F	T12-L1. EM	C	NER	None
Critchley <i>et al.</i> , 1997 <sup>[5]</sup>	17	F	T9-T10. IM	INC	Yes, at 12 months	Radiation
Burton <i>et al.</i> , 1997 <sup>[4]</sup>	22	F	C1. IM	INC	Yes, at 7 months	Radiation
Takayama <i>et al.</i> , 2004 <sup>[15]</sup>	49	M	L2. EM	C	NER	None
Qu <i>et al.</i> , 2009 <sup>[10]</sup>	16	F	T11-T12. EM	NS	NS	None
Weinstein <i>et al.</i> , 2010 <sup>[17]</sup>	20	F	L1-L2. EM	C	NER	None
Lee <i>et al.</i> , 2013 <sup>[6]</sup>	22	F	T1-T2. EM	C	NER	None
Lee <i>et al.</i> , 2013 <sup>[6]</sup>	21	F	C5-C6. EM	C	NER	None
Vaghasiya <i>et al.</i> , 2014 <sup>[16]</sup>	13	F	L1-L3. EM	INC	NER	None
Brown <i>et al.</i> , 2015 <sup>[3]</sup>	13	F	C4-C5. IM	INC	NS	None
Li <i>et al.</i> , 2016 <sup>[7]</sup>	9	M	T11-T12. EM	ST	Yes, at 12 months	None (asymptomatic)
Li <i>et al.</i> , 2016 <sup>[7]</sup>	12	F	L1. EM	ST	NER	None
Shen <i>et al.</i> , 2016 <sup>[11]</sup>	35	F	C4-C5. EM	C	NER	None
Al-Karawi <i>et al.</i> , 2016 <sup>[11]</sup>	11	M	L2-L3. EM	NS	NER	None
Bhati <i>et al.</i> , 2017 <sup>[2]</sup>	13	F	T12-L1. EM	Likely INC	Yes, at 7 months	Incomplete surgical excision and radiation
Mundi <i>et al.</i> , 2017 <sup>[9]</sup>	21	F	S1. EM	INC	NER	None
Soni and Bansal 2018 <sup>[13]</sup>	11	F	T12-L1. EM	INC	Yes	Incomplete surgical excision and radiation
Xue <i>et al.</i> , 2019 <sup>[19]</sup>	26	F	L4. EM	NS	NER	None
Soldozy <i>et al.</i> , 2020 <sup>[12]</sup>	15	M	C7. EM	C	NER	None
Current case	49	M	L3-L4. EM	C	NER	None

F: Female, M: Male, EM: Extramedullary, IM: Intramedullary, C: Complete, INC: Incomplete, NS: Not stated, ST: Subtotal, NER: No evidence of recurrence



**Figure 2:** Spinal granular cell tumor. (a) The tumor is composed of sheets of polygonal tumor cells with abundant eosinophilic granular cytoplasm (Hematoxylin and Eosin, original magnification  $\times 20$ ). (b-d) The tumor cells are diffusely positive for S-100, SOX-10, and inhibin (original magnification  $\times 20$ ).

diagnosis for tumors in this location includes neurofibroma, schwannoma, meningioma, paraganglioma, epidermoid cyst, intradural dissemination, and, in the cauda equina region, myxopapillary ependymoma. Those tumors can be difficult to distinguish from GCTs on MRI.<sup>[15]</sup> Hence, histopathologic diagnosis is needed for a definitive diagnosis.

Three spinal GCTs were described as being intramedullary.<sup>[3-5]</sup> The significance of the presence of an intramedullary component is that the tumor may not be amenable to complete surgical resection because of the associated increased risk of morbidity. In the three reported cases of intramedullary GCTs, complete surgical resection was not possible and the tumors locally recurred within a year in two of the cases.<sup>[4,5]</sup> In the third case, follow-up information about the patient was not provided.<sup>[3]</sup>

The vast majority of GCTs behave in a benign fashion; however, they may recur if incompletely excised.<sup>[18]</sup> None of the reported cases of spinal GCTs was a malignant GCT and none metastasized. GCTs are treated by complete surgical excision, when feasible. Local recurrence was seen in cases that were incompletely excised.<sup>[2,4,5,7,13]</sup> Of the five cases that locally recurred, two cases were subsequently treated with surgical excision and radiation therapy, two cases were treated with radiation therapy, and one case was asymptomatic so only follow-up was performed.<sup>[2,4,5,7,13]</sup>

## CONCLUSION

GCTs are peripheral nerve sheath tumors that rarely occur in the spinal canal where they are usually intradural/extramedullary in location. The optimal treatment is gross

total surgical excision, as subtotal/partial removal may result in recurrence and the need for adjuvant radiation therapy.

## Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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## Conflicts of interest

There are no conflicts of interest.

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