ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

www.surgicalneurologyint.com

Surgical Neurology International

Editor-in-Chief: Nancy E. Epstein, MD, Clinical Professor of Neurological Surgery, School of Medicine, State U. of NY at Stony Brook.

SNI: Pediatric Neurosurgery

Editor
Frank Van Calenbergh, MD
University Hospitals; Leuven, Belgium

SNI
Open Access

Case Report

Ectopic dorsal root ganglion in cauda equina mimicking schwannoma in a child

Nobuya Murakami[1], Ai Kurogi[1], Satoshi O. Suzuki[2], Naoko Akitake[3], Takafumi Shimogawa[4], Nobutaka Mukae[5], Koji Yoshimoto[4], Takato Morioka[6]

[1]Department of Neurosurgery, Fukuoka Children's Hospital, Fukuoka, [2]Department of Psychiatry, Shourai Hospital, Karatsu, [3]Department of Urology, Fukuoka Children's Hospital, [4]Department of Neurosurgery, Graduate School of Medical Sciences, Kyushu University, Fukuoka, [5]Department of Neurosurgery, Iizuka Hospital, Iizuka, [6]Department of Neurosurgery, Hachisuga Hospital, Munakata, Japan.

E-mail: *Nobuya Murakami - murakami.n@fcho.jp; Ai Kurogi - aironron8282@gmail.com; Satoshi O. Suzuki - sosuzuki@shouraikai.jp; Naoko Akitake - akitake.n@fcho.jp; Takafumi Shimogawa - shimogawa28@gmail.com; Nobutaka Mukae - mukae@ns.med.kyushu-u.ac.jp; Koji Yoshimoto - yoshimoto.koji.315@m.kyushu-u.ac.jp; Takato Morioka - takatons1227@gmail.com

***Corresponding author:**
Nobuya Murakami,
Department of Neurosurgery,
Fukuoka Children's Hospital,
Fukuoka, Japan.

murakami.n@fcho.jp

Received : 05 December 2022
Accepted : 13 January 2023
Published : 27 January 2023

DOI
10.25259/SNI_1089_2022

Quick Response Code:

ABSTRACT

Background: A heterotopic dorsal root ganglion (DRG) is sometimes observed in the vicinity of dysplastic neural structures during surgery for open spinal dysraphism; however, it is rarely associated with closed spinal dysraphism. Distinguish from neoplasms by preoperative imaging study is difficult. Although the embryopathogenesis of a heterotopic DRG has been speculated to be migration disorder of neural crest cells from primary neural tube, its details remain unelucidated.

Case Description: We report a pediatric case with an ectopic DRG in cauda equina associated with a fatty terminal filum and bifid sacrum. The DRG mimicked a schwannoma in the cauda equina on preoperative magnetic resonance imaging. Laminotomy at L3 revealed that the tumor was entangled in the nerve roots, and small parts of the tumor were resected for biopsy. Histopathologically, the tumor consisted of ganglion cells and peripheral nerve fibers. Ki-67 immunopositive cells were observed at the periphery of the ganglion cells. These findings indicate the tumor comprised DRG tissue.

Conclusion: We report detailed neuroradiological, intraoperative and histological findings and discuss the embryopathogenesis of the ectopic DRG. One should be aware of the possibility of ectopic or heterotopic DRGs when cauda equina tumors are observed in pediatric patients with neurulation disorders.

Keywords: Neural crest cell, Neural tube, Neurulation disorder, Fatty terminal filum, Satellite glial cells

INTRODUCTION

A heterotopic dorsal root ganglion (DRG) is sometimes observed in the vicinity of dysplastic neural structures during repair surgery for myelomeningocele, as has been reported in Lendon and Emery's autopsy study in which 63 of 95 spinal cords from cases with open spinal dysraphism were associated with heterotopic ganglion tissues.[11] With regard to closed spinal dysraphism, two cases with split cord malformation have been reportedly associated with heterotopic paramedian DRGs; this was revealed by postoperative histopathological investigation in an infant case[4] and by autopsy in an adult case.[20] Ectopic or heterotopic

ganglion cells, that are not fully formed DRG, are sometimes microscopically revealed by postoperative histopathological investigation in a normal terminal filum and in cases with neurulation disorders such as spinal lipoma, limited dorsal myeloschisis, and retained medullary cord.[5,10,13-15,17,18,21] Conner *et al*. recently described an ectopic DRG in cauda equina in an adult case without spinal cord malformation, which had been preoperatively recognized as a cauda equina tumor.[1]

We treated a pediatric case with an ectopic DRG in cauda equina who had a fatty terminal filum and bifid sacrum. On preoperative magnetic resonance imaging (MRI), the DRG mimicked a schwannoma in the cauda equina. We report detailed neuroradiological, intraoperative, and histological findings and discuss the embryopathogenesis of the lesion.

CASE REPORT

A 4-year- and 6-month-old boy was referred to us due to intermittent dribbling of urine and involuntary loss of a small amount of stool, both of which had been present since birth. No family history was noted. Cognitive development was considered to be appropriate for his age. Neurologically, his anal reflex was weak, but he had no motor dysfunction or sensory disturbance in the lower extremities. He had a tiny dimple at the lumbosacral region. There were no cutaneous stigmata indicative of Café-au-lait spot. His general condition was good, except for short stature which was being followed by a pediatrician.

MRI demonstrated a small tumor in the cauda equina that had isosignal intensity to the spinal cord on three-dimensional T1-weighted imaging (3D-T1WI) and 3D heavily T2WI (3D-hT2WI) and was slightly enhanced with gadolinium [Figures 1a-e]. A spinal lipoma of filar type with the conus medullaris at L2 level was also demonstrated. Detailed imaging study using 3D-hT2WI[12] revealed that the tumor was located in the left dorsal nerve roots of L3 and L4, presumed from the entry point of the root into the dural sac [Figure 1f]. Local bulging was observed at the root sleeves of L3 and L4, indicating the presence of normally-positioned DRG. A 3D reconstructions of computed tomography image revealed spina bifida below S2 and abnormal fusion of the lamina of L5-S2 [Figure 1g]. No intracranial tumor was observed on brain MRI. Urological examination revealed bladder dysfunction consisting of low compliance and decreased urine retention.

Although we considered the possibility of a neoplasm, as the patient's bladder dysfunction had existed since birth and had not progressed, a follow-up MRI was performed 7 months later; this revealed that the cauda equina tumor remained unchanged in size. The diagnosis was uncertain and we proposed diagnostic surgery for the cauda equina tumor, as well as prophylactic untethering of the spinal cord for the fatty terminal filum. Because his family was concerned about postoperative neurological deterioration due to possible damage to the cauda equina, only untethering of the cord was performed at 5 years and 2 months of age.

The fatty filum was severed through L5-S1 interspace. The cauda equina observed in the operative field appeared normal. Histologically, the fatty filum consisted of fibroadipose tissue with a tiny glial fibrillary acidic protein immunopositive tissues. Postoperatively, no *de novo* neurological abnormalities were observed. His family subsequently provided consent for the patient to undergo surgery for the cauda equina tumor.

Partial removal of the cauda equina tumor was performed at 5 years and 6 months of age. Laminoplastic laminotomy at L3 revealed a spherical tumor with a long axis of 10 mm at the left dorsolateral portion in the dural sac [Figure 2a-1]. The tumor was surrounded by nerve roots; a few normal-appearing nerve roots ran over the surface of the tumor and extended caudally, while some thinner nerve roots ended at the tumor surface [Figure 2a-2]. Direct stimulation of the tumor surface and the adhering nerve roots showed no evoked compound muscle action potentials on intraoperative neurophysiological monitoring (IONM) of the legs and anus. The roots were not determined to be non-functional because they could be dorsal roots on the basis of their anatomical location. Because the nerve roots intimately adhered to the tumor, we considered it impossible to dissect the tumor without damaging the nerve roots, and therefore, small parts of the tumor were resected for biopsy. Postoperatively, no *de novo* neurological abnormalities were observed. MRI performed after the second surgery revealed that the untethering of the cord was achieved and the cauda equina tumor remained unchanged [Figure 2b].

Histopathologically, the resected section consisted of ganglion cells and peripheral nerve fibers [Figures 2c-e]. At the periphery of the ganglion cells, a few Ki-67 immunopositive satellite glial cells or macrophages were observed. No neoplastic component was present. These findings indicated that the tumor comprised DRG tissue.

DISCUSSION

A DRG is derived from neural crest cells located in the dorsal region of the primary neural tube. Some neural crest cells migrate through somite and stop midway lateral to the neural tube, giving rise to the DRG.[7,22] Dorsal nerve roots are formed by nerve processes from neuroblasts in the

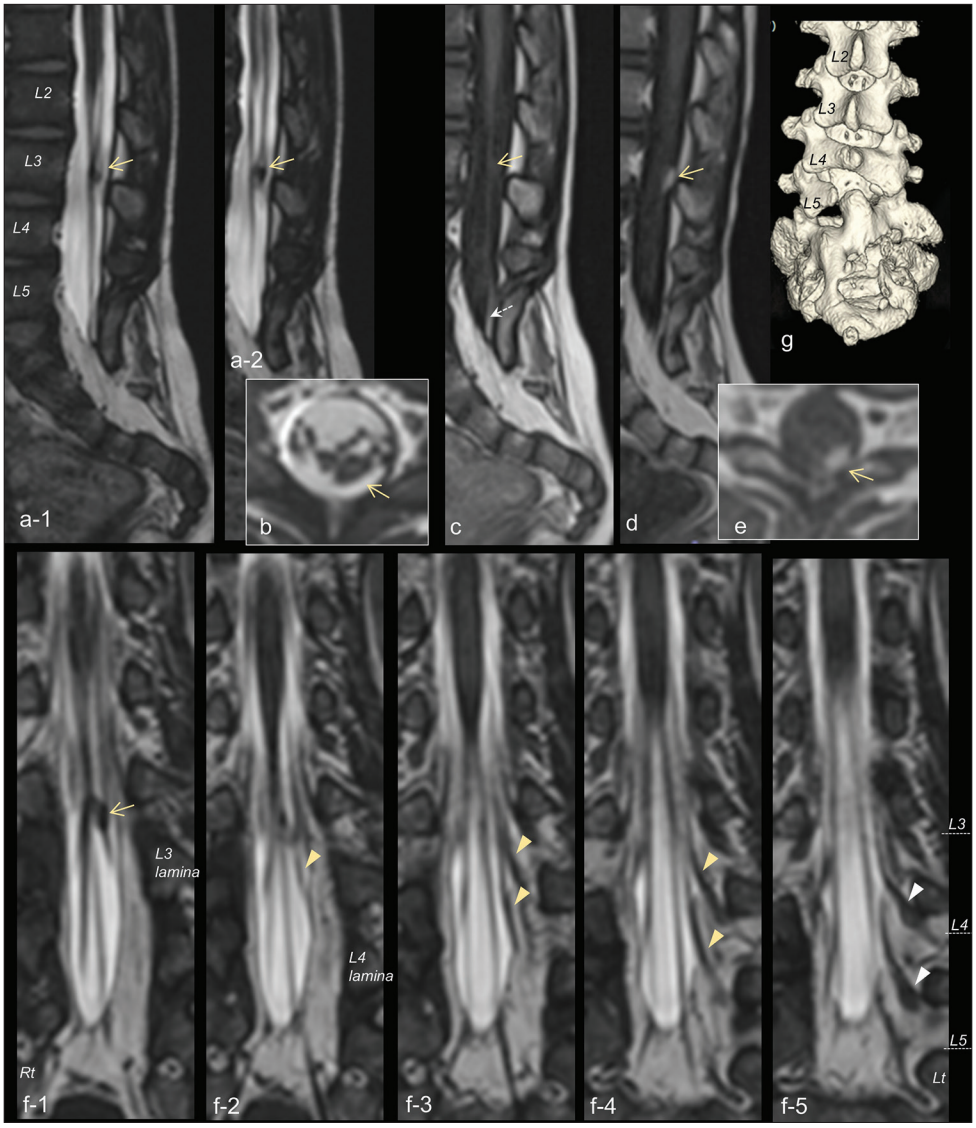

Figure 1: Sagittal (a-1 and 2) and axial (b) views of three-dimensional heavily T2-weighted imaging (3D-hT2WI) demonstrating a tumor (yellow arrow) in the cauda equina. (c) 3D T1-weighted images (3D-T1WI) showing the isointense lesion (yellow arrow) of the cauda equina and a high intensity lesion (dotted arrow) of the terminal filum. Sagittal (d) and axial (e) views of 3D-T1WI demonstrating the tumor enhanced with gadolinium (yellow arrow). (f-1-5) Serial coronal views of 3D-hT2WI delineating that the tumor (yellow arrow) is located in the dorsal nerve roots (yellow arrowheads) that enter the dural sac at the left L3-L4 and L4-L5 intervertebral space. Normally-positioned dorsal root ganglions (white arrowheads) are observed at the root sleeves as local bulging. The level of each vertebral body is indicated by a dashed line. (g) 3D reconstructions of computed tomography image revealing spina bifida below S2 and abnormal fusion of the sacral lamina of L5-S2.

ganglion. The differentiation of neural crest cells into DRG is modulated by signals from the neural tube, including Wnt and bone morphogenetic protein.[6,9] Delayed and incomplete closure of the neural tube is postulated to cause a delay in migration of the neural crest cells and subsequent ectopia to explain the association of heterotopic DRG with open spinal dysraphism[11] and diastematomyelia.[20] The present case is associated with a fatty terminal filum and sacral bony malformation, which are thought to be related to secondary neurulation failure but not primary neural tube closure.[16,23] To the best of our knowledge, the association of fully formed heterotopic or ectopic DRG with secondary neurulation disorders has not been reported; hence, its impact on the embryopathogenesis is uncertain. From the neuroimaging

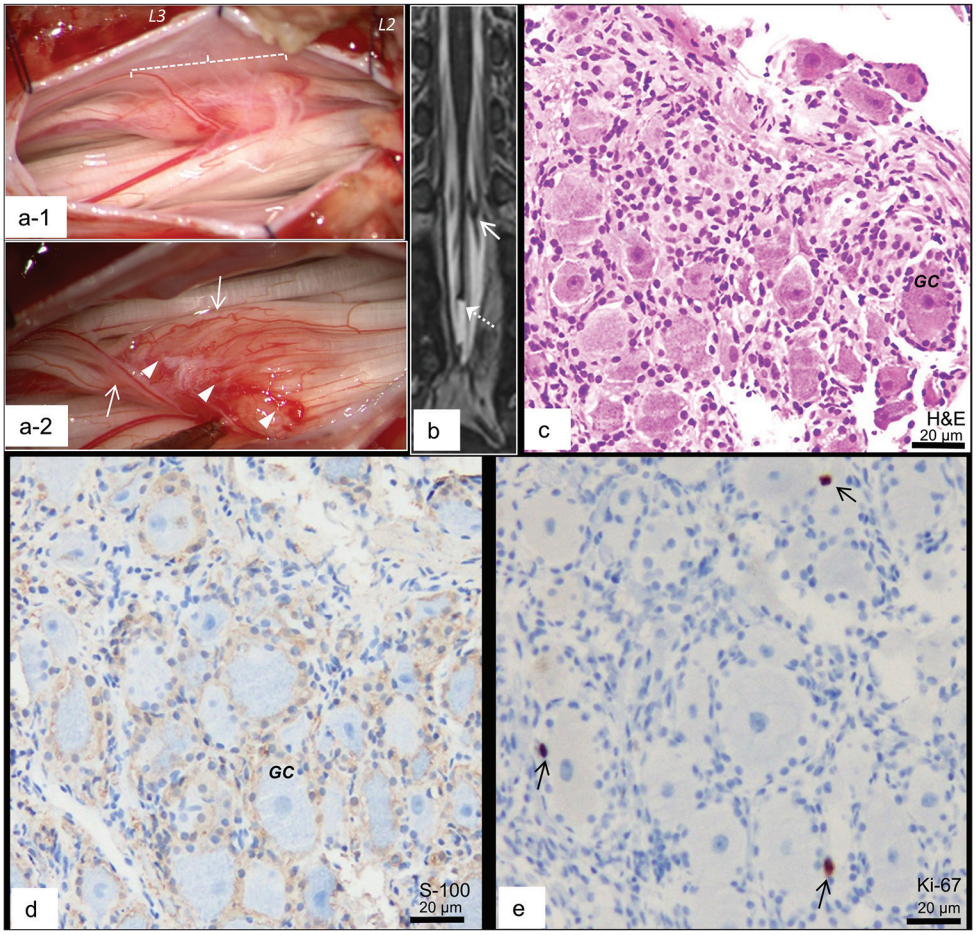

Figure 2: (a-1 and 2) Microscopic view of the surgery for the cauda equina tumor. Laminotomy at L3 revealed a tumor (a-1; blanket length of 10 mm). (a-2) A few nerve roots running over the lesion surface and extending caudally (arrows); some thinner nerve roots ending at the tumor surface (arrow heads). (b) Coronal view of 3D-hT2WI after the second surgery demonstrated that the cauda equina tumor (arrow) remained unchanged and the untethering of the cord was achieved (dotted arrow). (c-e) Histopathological findings of the resected parts of the tumor stained with hematoxylin and eosin (H & E; c), and immunostained with S-100 (d) and Ki-67 (e). (c) The resected section consisted of ganglion cells (GC) and peripheral nerve fibers. The periphery of the ganglion cells is immunopositive for S-100 (d), a few of which are immunopositive for Ki-67 (arrows; e).

finding of the present case that the dorsal nerve roots entangled with the ectopic DRG possibly connected to the normally positioned DRGs in the root sleeves, we speculated that some of the ganglion cells might have successfully migrated to the future formation site of the root sleeves and developed to the DRGs, while some cells might have been left behind at the intradural region and develop into the ectopic DRG.

A human DRG normally consists of cell bodies of primary sensory neurons surrounded by satellite glial cells, macrophages, and bundles of peripheral nerve fibers.[3] Histologically, the ectopic DRG in the present case contains all these features. Ki-67 immunopositive cells at the periphery of ganglion cells, which were probably satellite glial cells or macrophages surrounding the glial cells, might have been a population of self-renewing cells.[8] Although these histopathological features indicate that the ectopic DRG in the present case might be active, whether it is functional remains uncertain, as it could not be confirmed using IONM. From a clinical perspective, the contribution of the ectopic DRG to the patient's symptom was likely small, because its location at the L3-4dorsal roots failed to explain his neurological bladder, and it had no mass effect.

Tumors of the cauda equina in pediatric cases include schwannoma, neurofiboma, myxopapillary ependymoma,

dermoid/epidermoid cyst, and paraganglioma.[2] Ectopic DRGs in both the present and previously reported cases[1] were enhanced with gadolinium on MRI, probably because of rich vascular supply[3] and weak blood-nerve barrier,[19] which may make it difficult to rule out neoplasms preoperatively.

CONCLUSION

The present case demonstrate that heterotopic or ectopic DRGs could occur in cauda equina in pediatric patients with neurulation disorders and mimic neoplasms on imaging study. Careful observation of neurological symptoms combined with repeated MRI examination, and diagnostic surgery, when required, is necessary.

Ethics statement

The authors confirm that written informed consent was obtained from the family of the infant described in this report. The authors declare that this work complies with the guidelines for human studies and the research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

The Research Foundation of Fukuoka Children' Hospital.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Conner AK, Fung KM, Peterson JE, Glenn CA, Martin MD. Ectopic ganglion in cauda equina: Case report. J Neurosurg Spine 2016;24:937-40.
2. Engelhard HH, Villano JL, Porter KR, Stewart AK, Barua M, Barker FG, *et al*. Clinical presentation, histology, and treatment in 430 patients with primary tumors of the spinal cord, spinal meninges, or cauda equina. J Neurosurg Spine 2010;13:67-77.
3. Haberberger RV, Barry C, Dominguez N, Matusica D. Human dorsal root Ganglia. Front Cell Neurosci 2019;13:271.
4. Hamasaki T, Makino K, Morioka M, Hasegawa S, Kurino M, Kuratsu JI. Histological study of paramedian dorsal root ganglia in an infant with split cord malformation. Case report. J Neurosurg 2006;104(6 Suppl):415-8.
5. Hiraoka A, Morioka T, Murakami N, Suzuki SO, Mizoguchi M. Limited dorsal myeloschisis with no extradural stalk linking to a flat skin lesion: A case report. Childs Nerv Syst 2018;34:2497-501.
6. Kalcheim C, Le Douarin NM. Requirement of a neural tube signal for the differentiation of neural crest cells into dorsal root ganglia. Dev Biol 1986;116:451-66.
7. Kasemeier-Kulesa JC, Kulesa PM, Lefcort F, Zochodne D. Imaging neural crest cell dynamics during formation of dorsal root ganglia and sympathetic ganglia. Development 2005;132:235-45.
8. Krishnan A, Bhavanam S, Zochodne D. An intimate role for adult dorsal root ganglia resident cycling cells in the generation of local macrophages and satellite glial cells. J Neuropathol Exp Neurol 2018;77:929-41.
9. Krispin S, Nitzan E, Kalcheim C. The dorsal neural tube: A dynamic setting for cell fate decisions. Dev Neurobiol 2010;70:796-812.
10. Kural C, Guresci S, Simsek GG, Arslan E, Tehli O, Solmaz I, *et al*. Histological structure of filum terminale in human fetuses. J Neurosurg Pediatr 2014;13:362-7.
11. Lendon RG, Emery JL. Heterotopic dorsal-root ganglion cells around the spinal cord in children with spina bifida aperta. Dev Med Child Neurol Suppl 1976;37:16-21.
12. Morioka T, Hashiguchi K, Yoshida F, Nagata S, Miyagi Y, Mihara F, *et al*. Dynamic morphological changes in lumbosacral lipoma during the first months of life revealed by constructive interference in steady-state (CISS) MR imaging. Childs Nerv Syst 2007;23:415-20.
13. Morioka T, Suzuki SO, Murakami N, Shimogawa T, Mukae N, Inoha S, *et al*. Neurosurgical pathology of limited dorsal myeloschisis. Childs Nerv Syst 2018;34:293-303.
14. Morioka T, Suzuki SO, Murakami N, Mukae N, Shimogawa T, Haruyama H, *et al*. Surgical histopathology of limited dorsal myeloschisis with flat skin lesion. Childs Nerv Syst 2019;35:119-28.
15. Morioka T, Murakami N, Kurogi A, Mukae N, Shimogawa T, Shono T, *et al*. Embryopathological relationship between retained medullary cord and caudal spinal lipoma. Interdiscip Neurosurg 2022;29:101534.
16. Morota N, Ihara S, Ogiwara H. New classification of spinal lipomas based on embryonic stage. J Neurosurg Pediatr 2017;19:428-39.
17. Mukae N, Morioka T, Suzuki SO, Murakami N, Shimogawa T, Kanata A, *et al*. Two cases of large filar cyst associated with terminal lipoma: Relationship with retained medullary cord. World Neurosurg 2020;142:294-8.
18. Murakami N, Morioka T, Shimogawa T, Mukae N, Inoha S, Sasaguri T, *et al*. Ependyma-lined canal with surrounding neuroglial tissues in lumboscaral lipomatous malformations: Relationship with retained medullary cord. Pediat Neurosurg 2018;53:387-94.
19. Reinhold AK, Rittner HL. Characteristics of the nerve barrier and the blood dorsal root ganglion barrier in health and disease. Exp Neurol 2020;327:113244.
20. Ross GW, Swanson SA, Perentes E, Urich H. Ectopic midline spinal ganglion in diastematomyelia: A study of its connections. J Neurol Neurosurg Psychiatry 1988;51:1231-4.
21. Shirozu N, Morioka T, Inoha S, Imamoto N, Sasaguri T. Enlargement of sacral subcutaneous meningocele associated

with retained medullary cord. Childs Nerv Syst 2018;34:1785-90.
22. Tosney KW. The early migration of neural crest cells in the trunk region of the avian embryo: An electron microscopic study. Dev Biol 1978;62:317-33.
23. Yang J, Lee JY, Kim KH, Wang KC. Disorders of secondary neurulation: Mainly focused on pathoembryogenesis. J Korean Neurosurg Soc 2021;64:386-405.

How to cite this article: Murakami N, Kurogi A, Suzuki SO, Akitake N, Shimogawa T, Mukae N, *et al*. Ectopic dorsal root ganglion in cauda equina mimicking schwannoma in a child. Surg Neurol Int. 2023;14:33.