**Introduction**

Small neoplastic lesions of the vertebral column and the spinal canal are often misdiagnosed as intervertebral disc disease owing to the commoner occurrence of the latter [1]. Rarely, a neoplastic lesion may coincide with a herniated intervertebral disc usually without a topographic relationship in between [8]. Concomittant appearance of a disk herniation and a neoplastic lesion compromising the same nerve root represent a unique situation [1]. We herein report the unique case of a pseudotumor of the L3 dorsal root ganglion (DRG) adjacent to a far-lateral L3-4 disc herniation.

**Case history**

A 61-year-old physician presented with excruciating pain in her left leg. The mild leg pain that has been around the left knee and around the posterior hip for some weeks suddenly became unbearable one afternoon. The patient came to emergency room 8 h after the onset of the pain. Upon further questioning, the patient confessed that she had not emptied her bladder since the pain started. Past medical history was positive for a distant episode of trigeminal neuralgia, cluster headache, and an episode of significant back and right leg pain 7 years ago and a recent pulmonary embolus. Past surgical history included total hysterectomy and bilateral

**Abstract**

Dorsal root ganglia are oval enlargements on the dorsal nerve roots and contain the cell bodies of sensory neurons. Asymmetry of dorsal root ganglia may occur naturally, yet natural occurrence of gigantic dorsal root ganglion (DRG) is rare. The patient was 61-year-old woman who presented with atypical symptoms like neuropathic pain and urinary distention. Neuroimaging has shown left L3-4 far-lateral disc herniation and a gigantic L3 DRG. At surgery, the dural sheath of the ganglion had to be opened and a firm, yellow-colored abnormal tissue was exposed. The abnormal tissue considered to be a tumor of neural origin was gross totally excised and the patient’s symptoms ceased immediately after surgery. Histopathological examination of the specimen revealed nothing more than normal DRG morphology. At 4 months postoperatively, the patient is well with mild L3 hyperesthesia and hyperalgesia. Dural sheath opening in neurosurgery is not a routine practice. The sheath may need to be opened when surgeon suspects of a tumor, a free disc fragment and any inflammation within the ganglion. Operative morphology of a severely edematous but non-tumoral (pseudotumor) ganglion has not previously been documented.

**Keywords** Dorsal root ganglion · Ganglionectomy · Magnetic resonance imaging · Microsurgery
salpingo-oophorectomy after a serious pelvic inflammatory disease and abscess formation within the uterine tube.

In the emergency room, the patient was in extreme pain. Any movement or touch would exacerbate the pain. Right leg-raising test was positive at 60 and left leg-raising test was positive at 20°. Power was normal in both legs, including knee extension. Patellar and the ankle reflexes were normoactive bilaterally. Abdominal examination for a distended bladder was fruitless because of softened subcutaneous fat. Lumbar MR showed disc degeneration and mild bulging at L3-4, L4-5 and L5-S1 levels yet at L3-4 level there was foraminal and a round extraforaminal disc herniation (Fig. 1a, b). Lumbar MR scan also verified a fully distended bladder (Fig. 1c). Considering vaque radiological findings in contrast to serious pain and urinary retention, the MR examination was extended to cover the whole cervico-thoracic and thoracic levels. There was no cord compression.

The next morning, urinary retention re-occurred and an indwelling urinary catheter had to be placed. An ultrasound of the abdomen and pelvis was not contributory except identification of few benign adhesions secondary to previous pelvic surgery and a distended gall bladder. Meanwhile the left knee pain and the left sciatica got only a bit better but the patient described additional occasional electric shock like pain radiating medially from the left groin to the left knee. Lumbar computerized tomography (CT) to rule out a small bone chip originating from end-plate attached to a free disc fragment was done next. Axial CT scans showed the presence of a left-sided round hyperdense extraforaminal lesion at L3-4 level that was reported to be consistent with far-lateral disc herniation (Fig. 2a). Coronal reformatted CT images better demonstrated the lateral extent of the disc herniation (Fig. 2b). After an unsatisfactory 2-day narcotic treatment with codein 60 mg q6h, gabapentin 300 mg twice a day was started. On the third day of admission, a new pain emerged around the left inguinal area. By this time, the left patellar reflex diminished and left knee extension got 20% weaker. IV steroid (80 mg methyl prednisolon as the loading dose and 40 mg twice a day as the maintenance dose) was then started. Steroid alleviated a significant portion of the pain. Tapering steroid dose immediately caused return of intense pain, so surgery was considered as the last resort.

The patient was informed to consent for the removal of far-lateral disc herniation at L3-4 level. After medial facetectomy and ligamentous removal, the left L4 nerve and the L3-4 disc space were exposed. The bulging disc was compressing the L4 root from its shoulder. After standard discectomy, the L4 root became mobile. Next, lateral disc space was emptied by angling the rongeur laterally. Yet, disc compression alone was far from explaining the dramatic clinical picture. After total removal of left L3 inferior facet and L4 superior facet and undercutting the L3 pedicle, the left L3 root was identified and traced laterally. L3 nerve was more or less normal sized yet L3 DRG was extremely different than normal. The DRG was 3–4 times bigger than a normal DRG. The L3 root was totally immobile (Fig. 3a). The DRG felt extremely gritty. The dural sheath of nerve root and the DRG was opened. A yellow-colored firm tissue came into view (Fig. 3b, c). There was good cleavage from the dural sheath and the underlying septum. The healthy ventral rootlets were pushed anteriorly and inferiorly by the mass (Fig. 3d). Overall, the mass lesion within the root sleeve was totally extirpated. Although no cerebrospinal fluid (CSF) was seen in the operative field, the dural sheath was sutured with interrupted prolene sutures and the suture was reinforced with tissue fibrin glue. On postoperative day 1, the urinary catheter was removed and the patient emptied her bladder spontaneously. The original pain was totally gone. The left knee function returned to normal. On third postoperative day, she complained of mild hyperesthesia and hyperalgesia over the left knee which did not affect her daily life. At 4 months postoperatively, she is leading a normal life with gabapentin 300 mg twice a day.

The histopathological specimen was extensively studied and did not reveal anything other than normal DRG tissue with mature ganglion cells, axons and Schwannian stroma supporting the axons (Fig. 4).

Discussion

Dorsal root ganglia are oval enlargements on the dorsal nerve roots and contain the cell bodies of sensory neurons. Although both the DRG and the ventral motor root are surrounded by a common dural sheath, a thin fibrous septum separates both. The DRG are surrounded by fat within the intervertebral foramen. According to the baseline anatomic information that Hasegawa et al. [5] obtained from male volunteers using MRI, the mean width, length and height is 5.7×7.1×7.3 mm³ for L3 DRG, 6.2×8.4×8.2 mm³ for L4 DRG, 5.9×9.4×8.3 mm³ for L5 DRG and 6.2×11.2 mm² for S1 DRG, respectively (data on the height of S1 DRG was not given in this study). The lower was the spinal level, the bigger was the ganglion size. In that study, the DRGs were located in lumbar intervertebral foramen in 92% of L1, 98% of L2, 100% of L3 and L4 and 95% of L5 levels. The S1 DRG was intraspinal in 79%. However analyzing patients presenting with low-back pain or sciatica, Hamanishi and Tanaka [4] found that 100% of L2 DRG, 48% of L3 DRG, 27% of L4 DRG and 12%
of L5 DRG were located extraforaminally while 52% of L3 DRG, 72% of L4 DRG and 75% of L5 DRG were located intraforaminally.

Preoperative radiological diagnosis of spinal nerve tumors producing radiculopathy or masquerading as spinal and/or root pain has been facilitated by combined
use of CT and MR. Intradural extramedullary tumors like schwannoma or extradural tumors like neurofibroma frequently affect the DRG and appear as dumbbell-shaped in 15% of cases [10, 14]. Diagnosis of tumors that had already enlarged the neural foramen is usually straightforward. However, a smaller pathology intrinsic to the root or the DRG will first be expected to cause an asymmetry as also depicted by MR scans in our patient. Yet asymmetry of DRGs may also occur naturally [4]. Hamanishi and Tanaka [4] found asymmetrical locations and sizes in 11% of the reviewed DRGs. Asymmetry at L3-4 level is rarer. Out of 104 patients that they reviewed, only one had DRG asymmetry at L3-4 level. Two cases who had asymmetrically gigantic DRGs similar to our case underwent surgery at different

Fig. 3 Operative microphotographs show that: a the left L3 root and the DRG are extremely swollen, b after opening the dural nerve sheath, the upper end of a yellow colored tumor is being dissected, c the lower end of the tumor is being dissected with better cleavage, d after the removal of the tumor normal appearing ventral motor rootlets come into view, e root size return to normal after suture closure of the dural sheath (black arrows)

Fig. 4 Photomicrographs a with hematoxylin and eosin and b with neurofilament (NF) protein immuno-stain demonstrate axons (left) and ganglion cells dispersed in a cellular stroma consisting of bundles of elongated spindle cells (right) (×200 original magnification). This represents normal DRG morphology
spinal levels [4]. In the first case, the lowered pedicle appeared to push down and kink the intraspinal portion of the L5 DRG. In the second case, the mass mimicking the gigantic DRG was histologically confirmed to be very old sequesterated nucleus pulposus at L5-S1 level. However, anteriorly compressed DRG could not be detected by MR scan at all [4].

As for MR signal intensity, schwannomas and neurofibromas will often appear iso- to hypointense on T1- and hyperintense on T2-weighted MR images [14]. The left L3 DRG in our case appeared isointense on both T1- and T2-weighted MR images. Intermediate signal on T2-weighted MR image usually suggests the presence of a tumor with numerous cellular and fibrous components and very little myxoid stroma. Unfortunately, paramagnetic contrast was not given in our case. One can argue that the preoperative radiological diagnosis of a tumor could have been possible if intravenous gadolinium was given. Although gadolinium enhanced MR imaging allows for better detection of intranuclear edema with diffuse enhancement along the involved nerve roots, this characteristic probably would not have helped us in diagnosing a DRG tumor since DRG always enhance with contrast medium because of a less developed blood–nerve barrier anyway. Recently, a French group studying enhancement pattern on 180 healthy DRG also concluded that 100% demonstrated significant and homogeneous enhancement [3]. Therefore, even if we had used intravenous paramagnetic contrast preoperatively, it would probably be impossible to differentiate between an intraradicular tumor and a healthy DRG.

It was only at surgery that the left L3 DRG was found to be extremely swollen and had to be opened. A nerve root and a DRG may become swollen because of both biochemical irritation of nucleus pulposus and the mechanical compression of the disc material [2, 6]. The mechanical compression of the DRG can increase the endoneural fluid pressure within the DRG almost threefolds [11]. Yet since the dural sheath is not an elastic tissue, a DRG can hardly ever reach the gigantic size that we have observed. Moreover, the far-lateral disc protrusion at L3-4 level observed at surgery (in contrast to the extent of the herniation as seen on CT and MR images) could not have exerted the force necessary to cause a significant endoneural fluid pressure rise within the left L3 DRG. Therefore, the adjacent far-lateral disc herniation probably just coexisted with the gigantic left L3 DRG of unknown origin and served a better purpose by attracting our surgical attention to this part of the spine. Not only the size of the involved DRG, but also its firm feeling suggested the presence of something unusual inside it. Although intraradicular disc herniations occur more commonly at L5-S1 and L4-5 levels [12], the surgeon’s impression before the microsurgical opening of the dural sheath was more inclined to a rare intraradicular disc herniation rather than a tumor.

Dural opening in similar cases totally depend on surgeon’s experience and/or attitude or on the medico-legal restrictions of the preoperative consenting procedure. By opening the dural sheath of a nerve root or a DRG the surgeon not only will prolong the operative procedure but also will take the morbidity risk associated with CSF leak and difficulty with wound healing. Moreover, extra expense of tissue fibrin glue and/or external lumbar drainage may also trigger an hesitation before dural opening in the currently managed health care systems. If a gross pathology like a tumor or a disc fragment is not readily encountered between the rootlets, chances are histopathology will be the only diagnostic tool. In addition to a real mass lesion like tumor or disc material, inflammatory reactions within a DRG—although very rare—may also cause a tumor-like lesion. A team from Tübingen, Germany recently has reported a very similar case presenting with low back pain and progressive hypesthesia of the thigh [10]. MR imaging with contrast enhancement revealed an intraspinal–extradural mass at L1-2 level compressing the right L2 root. The surgical part of their case is almost identical to ours. The surgical team emptied the disc and the nerve was still compressed and appeared to be enlarged. In the end, they had to open the dural sheath and resect the indurated and swollen part of the ganglion. Histopathological diagnosis was ganglionitis characterized by loss of ganglion cells, increased number of nodules of Nagcotte and diffuse infiltration of T cells.

Ganglionectomy as performed for cancer pain or non-cancer pain (for failed back surgery syndrome or chronic lumbar radiculopathy) was originally described by Osgood et al. [9] and by definition is an extradural operation [13]. Wilkinson and Chan [15] in their recent publication advocated the use of microscope for the procedure. After exposure of the ganglion and surrounding plexus and compression of the venous plexus with cottonoids, the longitudinal incision path along the dural sheath is coagulated using low-current bipolar. Once the dural sheath is incised dorsally using a no. 11 knife, ganglion tissue will come into view. Young [16] described the DRG as a round, yellow, irregular structure. Wilkinson and Chan [15] described the ganglion as having an appearance similar to fat, with tiny “sparkles” within. The latter description is for ganglia from patients with failed back surgery syndrome or chronic lumbar radiculopathy. In rare circumstances like we had, differentiation between an healthy and a tumoral ganglionic tissue may be extremely difficult even with the operating microscope. At this stage, the surgeon has to decide between a biopsy or total removal of the abnormal looking ganglionic tissue. Biopsy may be safer but then there is the possibility of sampling error. If total removal of the DRG is aimed (sensory ganglionectomy), then the afferents and efferents of the ganglion are elevated first.
and then the ganglion is separated from the underlying septum. To lessen the likelihood of neuroma formation, the proximal end of the ganglion is either hemoclipped or cut with CO2 laser [7]. If leakage of CSF is not encountered (which usually is the case) a watertight closure of the perineurium is not necessary. Some even advocated to leave an autogenous fat graft in the surgically created defect [15]. As for complication of the procedure, deafferentiation pain has been exaggerated because it is usually transient and never reaches to the point of anesthesia dolorosa [13, 15]. Taha et al. [13] states that there is no uniformly beneficial treatment for deaffarentiation pain after ganglionectomy but our patient definitely benefited from brief use of gabapentin.

In conclusion, we reported the unique case of a pseudotumor of L3 DRG. The patient presented with atypical symptoms like neuropathic pain and urinary distention. Preoperative MR scans showed a gigantic left L3 DRG yet this asymmetry was masked by the presence of an adjacent ipsilateral far-lateral disc herniation. Attention to foraminal and extraforaminal part of the spine is of utmost importance in the evaluation of lumbar MR scans. Surgery was both diagnostic and therapeutic in this case. Before opening the dural sheath of a nerve root or DRG, a surgeon should realize that if a tumor or a disc fragment does not come out at once, then it may only be through histopathology that a proper diagnosis can be reached.

References

1. Albert FK, Oldenkott P, Bieker G, Danz B (1988) Lumbar intervertebral disk herniation with a concomitant nerve root neurinoma at the same site. Case report and review of the literature. Neurochirurgia 31:222–225
2. Aota Y, Onari K, An H, Yoshikawa K (2001) Dorsal root ganglia morphologic features in patients with herniation of the nucleus pulposus: assessment using magnetic resonance myelography and clinical correlation. Spine 26:2125–2132
3. Demondion X, Leroy X, Lappegue F, Drizenko A, Francke JP, Cotten A (2001) Lumbar spinal ganglia enhancement after gadolinium chelate administration: a radio-histologic correlation. Surg Radiol Anat 23:415–419
4. Hamamishi C, Tanaka S (1993) Dorsal root ganglia in the lumbosacral region observed from the axial views of MRI. Spine 18:1753–1756
5. Hasegawa T, Mikawa Y, Watanabe R, An HS (1996) Morphometric analysis of the lumbosacral nerve roots and dorsal root ganglia by magnetic resonance imaging. Spine 21:1005–1009
6. Kobayashi S, Yoshizawa H, Yamada S (2004) Pathology of lumbar nerve root compression. Part 2: morphological and immunohistochemical changes of dorsal root ganglion. J Orthop Res 22:180–188
7. Kuzbari R, Liegl C, Neurmayer C, Moser H, Burggasser G, Holle J, Gruber H, Hoppak W (1996) Effect of the CO2 milliwatt laser on neurona formation in rats. Lasers Surg Med 18:81–85
8. Oro JJ, Geise AW (1983) Dumbell ganglioneuroma of the lumbar spine associated with a herniated intervertebral disc: case report. Neurosurgery 13:711–714
9. Osgood CP, Dujovny M, Faille R, Abassy M (1976) Microsurgical lumbosacral ganglionectomy. Technical note. J Neurosurg 45:113–115
10. Roser F, Ritz R, Morgalla M, Tatagiba M, Bornemann A (2005) Spinal nerve root ganglionitis as a cause of disc herniation. J Neurosurg Spine 2:472–475
11. Rydevik BL, Myers RR, Powel HC (1989) Pressure increase in the dorsal root ganglion following mechanical compression: closed compartment syndrome in nerve roots. Spine 14:574–576
12. Süzer T, Tahta K, Erdal C (1997) Intraradicular lumbar disc herniation: case report and review of the literature. Neurosurgery 41:956–959
13. Taha JM, Abdel Aziz KM, Andaluz N (2004) Dorsal rhizotomy and dorsal root ganglionectomy. In: Winn R (ed) Youmans neurological surgery, vol 3, 5th edn. Saunders, Philadelphia, pp 3033–3043
14. Tekkökk IH, Akpinar G, Güngen Y (2004) Extradural lumbosacral cavernous hemangioma. Eur Spine J 13:469–473
15. Wilkinson HA, Chan AS (2001) Sensory ganglionectomy: theory, technical aspects and clinical experience. J Neurosurg 95:61–66
16. Young RF (1990) Dorsal rhizotomy and dorsal root ganglionectomy. In: Youmans JR (ed) Neurological surgery, vol 6, 3rd edn. Saunders, Philadelphia, pp 4026–4035