Case Report
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Ganglioneuroma of Lumbar Nerve Root: A Case Report[1]
요추 신경근에 발생한 신경절신경종: 증례 보고[1]

Minhye Jeong, MD[1], Seunghun Lee, MD[1], Kyung Bin Joo, MD[1], Ki-Seok Jang, MD[2], Jiyoon Bae, MD[3]

Departments of [1]Radiology, [2]Pathology, Hanyang University Seoul Hospital, Seoul, Korea
[3]Department of Pathology, National Police Hospital, Seoul, Korea

Ganglioneuroma is a rare, benign, slow-growing, well-differentiated tumor consisting of ganglion cells and Schwann cells. Ganglioneuromas originate from neural crest cells and can affect any part of the sympathetic tissue from the skull base to the pelvis. However, ganglioneuroma occurring in the nerve root is extremely rare. We describe a 50-year-old man with ganglioneuroma involving the right 5th lumbar nerve root. The ganglioneuroma showed intermediate signal intensity on the T1-weighted image and high signal intensity on the T2-weighted image with homogeneous enhancement on the gadolinium-enhanced T1-weighted image.

Index terms
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Corresponding author: Seunghun Lee, MD
Department of Radiology, Hanyang University Seoul Hospital, 222 Wangsimni-ro, Seongdong-gu, Seoul 133-792, Korea.
Tel. 82-2-2290-9164 Fax. 82-2-2293-2111
E-mail: radsh@medimail.co.kr

INTRODUCTION

Ganglioneuroma is a rare, benign, slow-growing, well differentiated tumor originating from neural crest cells that form the sympathetic nervous system (1, 2). It is considered to be part of neuroblastic tumors (1, 3, 4). Depending on the degree of cellular and extracellular maturation, neuroblastic tumors are divided into neuroblastomas (most immature, undifferentiated, and with the most malignant potential), ganglioneuroblastomas (intermediate malignant potential), and ganglioneuromas (fully differentiated, most benign) (1, 3, 4). Ganglioneuromas can affect any part of the sympathetic tissue along the line of the embryonic neural crest, from the skull base to the pelvis (1). Ganglioneuroma occurring in the nerve root is very rare (1, 5). Thus, we present the radiologic and pathologic features of ganglioneuroma presenting as a right 5th lumbar nerve root mass. Radiological studies and literatures are also reviewed.

CASE REPORT

A 50-year-old man presented with a 3-month history of lower back pain and urinary frequency. He was diagnosed with hypertension and chronic kidney disease 15 years ago, and underwent renal transplantation in 1999 due to end stage renal disease associated with hypertension. After renal transplantation, CellCept® (Roche, Basel, Switzerland) was given as a immunosuppressive agent. He had taken CellCept® (Roche, Basel, Switzerland) 1000 mg per day for about 11 years.

On physical examination, paresthesia was detected along the dermatome of the right L5 nerve root. Motor functions of the bilateral hips, knees, and ankles were normal. Deep tendon reflexes were also intact. There were no pathologic reflexes.

A routine urine analysis revealed the following: white blood cell (WBC) 5-9/high power field (HPF), red blood cell 0-1/HPF, Epithelial cell 0-1/HPF, bacteria; nothing. Urine cytology revealed many neutrophils. Urine culture indicated Escherichia

coli. The patient's WBC count, blood urea nitrogen and creatinine had risen to 12.9 × 10^3/mm^3 (4.0-10.0 × 10^3), 30 mg/dL (7-20) and 1.6 mg/dL (0.4-1.4), respectively. Tumor markers (alpha-fetoprotein, carcinoembryonic antigen, CA19-9, CA125, and prostate-specific antigen) were normal. He was treated for his urinary tract infection using a third-generation cephalosporin. A mild improvement of urinary frequency was shown after medical treatment.

Plain radiograph of the hip demonstrated no abnormal findings. Magnetic resonance imaging (MRI) of the lumbar spine (Fig. 1A-D) revealed a 5.1 × 2.8 × 2.7 cm sized, dumbbell-shaped mass along the right 5th lumbar nerve root engulfing the right side of the iliac vessel. The mass showed intermediate signal intensity on the T1-weighted image and high signal intensity on the T2-weighted image with homogeneous enhancement on the gadolinium-enhanced T1-weighted image. On whole body positron emission tomography-computed tomography (PET-CT) (Fig. 1E), the mass disclosed mild metabolic activity (maximum standardized uptake value: 1.86) along the right 5th lumbar root.

Considering that patients under immunosuppressive treatment after renal transplantation have a higher risk for developing malignant neoplasms and since our patient had a dumbbell-shaped tumor along the lumbar nerve root showing intermediate signal intensity on the T1-weighted image and high signal intensity on the T2-weighted image with homogeneous contrast enhancement on MRI, the clinical impression was a lymphoma derived from the lumbar nerve root.

Medial facetectomy at the level of L5, S1 on the right and biopsy of the right L5 nerve root were performed.

On microscopic examination (Fig. 1F), mature ganglion cells were seen to be scattered among spindle cells, which were suggestive of ganglioneuroma. There were no immature or neurobastic components.

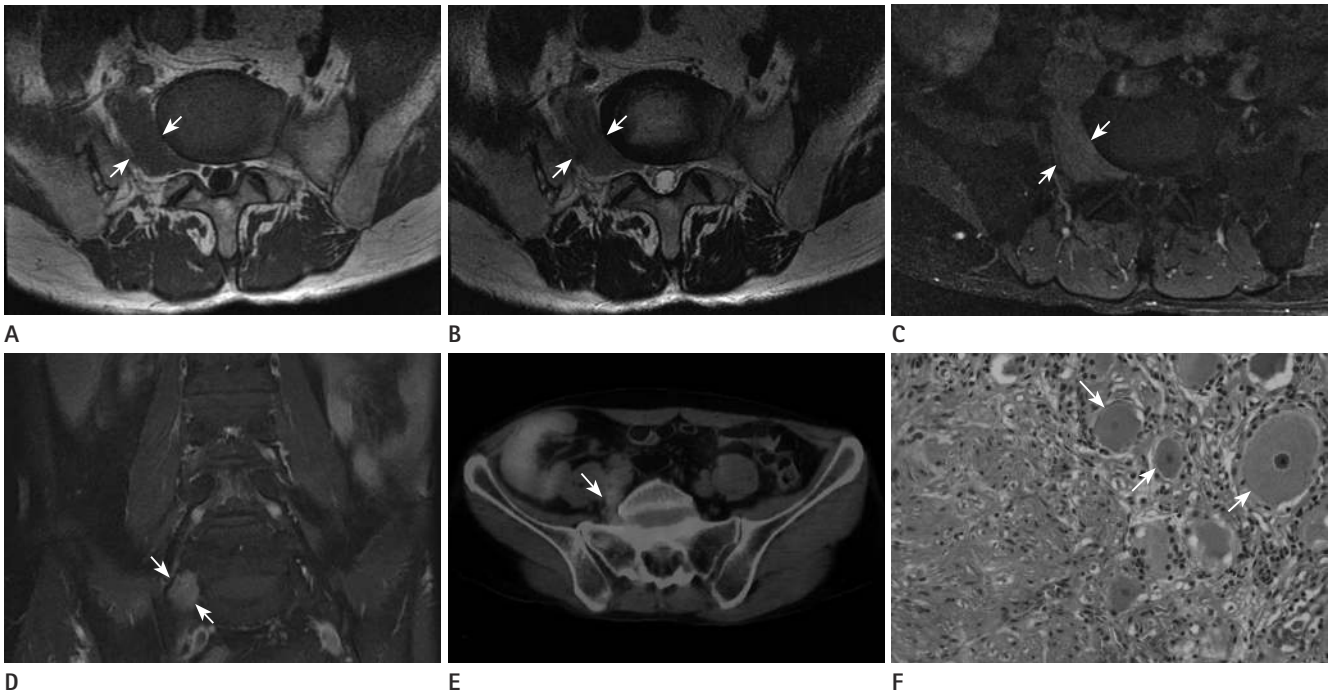

Fig. 1. A 50-year-old man with ganglioneuroma.
A, B. Axial T1 weighted **(A)** and T2 weighted images **(B)** show a lobulated mass (arrows) with intermediate signal intensity on the T1 weighted image and high signal intensity on the T2 weighted image along the right 5th lumbar nerve root.
C. Homogeneous enhancement was observed along the right 5th lumbar nerve root (arrows) on the axial contrast-enhanced T1-weighted image.
D. Coronal contrast-enhanced T1-weighted image shows a lobulated mass (arrows) arising from the right 5th lumbar nerve root with homogeneous enhancement.
E. Axial ^{18}F-FDG PET-CT image reveals a lobulated mass (arrow) with mildly increased FDG uptake (SUVmax: 1.86) in the right 5th lumbar nerve root.
F. Hematoxylin and Eosin photomicrograph (× 400) shows mature ganglion cells (arrows) scattered in spindle cell stroma.
Note.—^{18}F-FDG PET-CT = fluorine-18 fluorodeoxyglucose positron emission tomography-computed tomography, SUVmax = maximum standardized uptake value

DISCUSSION

Ganglioneuromas are a class of peripheral neuroblastic tumors defined as childhood embryonal tumors of migrating neuroectodermal cells originating from neural crest cells that form the sympathetic nervous system (1). Peripheral neuroblastic tumors include three subgroups depending on cellular and extracellular differentiation: neuroblastomas (most immature, undifferentiated, and with the most malignant potential), ganglioneuroblastomas (intermediate malignant potential), and ganglioneuromas (fully differentiated, most benign) (1, 3, 4). Ganglioneuromas consist entirely of ganglion cells and Schwann stroma (4).

Ganglioneuroma occurs most frequently in children and young adults under the age of 30 and rarely in those older than 60 years (3). It shows a slight female predominance (1.13 to 1.5 : 1) (3). It occurs any part of the sympathetic tissue exists from the skull base to the pelvis, but mainly from the paraspinal sympathetic chain ganglia (1). More specifically, the most common sites include the posterior mediastinum, adrenal gland, and in the lumbar and pelvic retroperitoneal space (1, 3). Unusual cases have been reported in the spermatic cord, heart, intestine, and bone (3, 4). Most cases involve the paraspinal region with intraspinal extension extradurally through the neural foramen, leading to dumbbell shaped tumors (6). This also leads to extradural compression of the spinal cord (6). Shephard and Sutton (5) reported that dumbbell-shaped gangioneuromas constitute 3.5% of all ganglioneuromas. Spinal ganglioneuroma occurs most frequently in the cervical spine, followed by thoracic spine and lumbar spine (6). Ganglioneuroma occurring in the nerve root is very rare (1, 5, 7). A few cases of ganglioneuroma occurring in the cervical nerve root have been reported (5, 7).

Findings such as the widening of the neural foramen and thickening of the nerve root help to narrow down the differential diagnosis to schwannoma, neurofibroma, meningioma, or ganglioneuroma (2). Ganglioneuroma is distinguished from other tumors, such as lymphomas, chordomas, soft-tissue sarcomas, Ewing's sarcoma, osteosarcomas, chondrosarcomas, and bone metastases, because it doesn't have the ability to involve, invade and erode bones (2). In the differential diagnosis of extradural nerve root tumors, metastatic diseases, lymphomas, and Ewing's sarcoma should be considered (8-10).

Discrete punctuate calcifications can be seen in about 42-60% of ganglioneuromas (3, 4). MRI typically shows low signal intensity on T1-weighted images and heterogeneous high signal intensity on T2-weighted images, with gradual increasing enhancement on dynamic images (3).

Tumors with intermediate to high signal intensity on T2-weighted MR images are caused by abundant cellular and fibrous components and only a small amount of myxoid stroma (4). In the case of lymphoma, MRI appearance is isointense on T1-weighted images and iso- to hyperintense signal intense on T2-weighted images (8). When a lymphoma is located in the epidural space, it can extend into the paraspinal space through the intervertebral foramen, leading to a dumbbell shaped tumor (8). Therefore, it would be difficult to differentiate ganglioneuroma from lymphoma. As our patient had a history of renal transplantation and a dumbbell-shaped mass showing intermediate signal intensity on the T1-weighted image and high signal intensity on the T2-weighted image, the most possible diagnosis was lymphoma.

Those with markedly high signal intensity on T2-weighted images are due to a large amount of myxoid stroma and relatively few cellular and fibrous components (4). A large amount of myxoid matrix and numerous fibrous capsules result in late contrast enhancement (3). Preoperative diagnosis of ganglioneuroma is usually difficult, thus histopathologic evaluation of the biopsy specimen is necessary for making an accurate diagnosis (3).

Histologically, ganglioneuroma consists of ganglion cells and mature Schwann cells arranged in interlacing fascicles mixed with mature ganglion cells (3).

The treatment of choice is surgical resection (3). Our patient underwent biopsy of the tumor only due to the inoperable location of the tumor. Follow up abdomen CT and PET-CT scans revealed no evidence of progression of the tumor.

Adjuvant systemic chemotherapy and local radiotherapy have limited roles due to their benign biological nature (3). The long term prognosis is excellent regardless of tumor location as long as total tumor excision is performed (3). However, local recurrence has been reported after surgical resection, so regular radiologic follow up with neurologic examination and radiologic evaluation is necessary even after complete excision (3, 4).

In conclusion, the present extremely rare case of ganglioneuroma of the lumbar nerve root illustrates occurrence. This case well illustrates that ganglioneuroma should be included in the

differential diagnoses for patients with dumbbell shaped lumbar nerve root tumors.

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요추 신경근에 발생한 신경절신경종: 증례 보고[1]

정민혜[1] · 이승훈[1] · 주경빈[1] · 장기석[2] · 배지윤[3]

신경절신경종은 천천히 자라고 잘 분화된 드문 양성 종양이며, 신경절세포와 슈반세포로 이루어져 있다. 신경절신경종은 신경능선세포에서 기원하며 교감신경이 위치하는 두개저에서부터 골반에 이르기까지 어디에서든 발생할 수 있다. 그러나 신경근에서 발생하는 신경절신경종은 매우 드물다. 저자들은 50세 남자 환자에서 우측 5번째 신경근을 침범한 신경절신경종의 증례를 보고한다. 신경절신경종은 MRI T1 강조영상에서 중등도 신호강도, T2 강조영상에서 고등도 신호강도를 보였으며, 가돌리늄 조영증강 T1 강조영상에서 균질한 조영증강을 보였다.

한양대학교병원 [1]영상의학과, [2]병리과, [3]국립경찰병원 병리과