CASE REPORT

Ganglioneuroma arising from the L5 nerve root: A rare case report

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ABSTRACT

Ganglioneuroma is a rare, benign, slow-growing, well-differentiated tumor consisting 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 44-year-old man with ganglioneuroma involving the right fifth lumbar nerve root.

Key words: Ganglioneuroma, lumbar nerve root, spine

Introduction

Ganglioneuromas originate from neural crest cells and can affect any part of the sympathetic tissue from the skull base to the pelvis. Those arising from spinal nerve root are extremely rare.[1]

Case Report

A 44-year-old male presented with low backache that aggravated on cough and strain. He also complained of numbness on both side saddle and toe and left calf muscle pain. On examination, root pain was along left L5 distribution with numbness in L5 dermatone. magnetic resonance imaging was suggestive of nerve root tumor [Figure 1]. Laminectomy in L5-S1, and excision if nerve root tumor after keeping perineural sheath intact. Tumor was excised end to end nerve sheath closed. On microscopic examination, there were clusters as well as scattered variably sized mature ganglion cells embedded in a stroma comprising of Schwann like cells with an elongated wavy serpentine nucleus [Figure 2]. The tumor was diagnosed as ganglioneuroma of L5 nerve root. Postoperatively, there was improvement with complete relief of pain and numbness around the saddle area and little toe in 7 days.

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Discussion

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. Ganglioneuromas are a class of peripheral neuroblastic tumors that 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).

Central nervous system ganglioneuromas are rare and most often occur in children and young adults with most prevalent

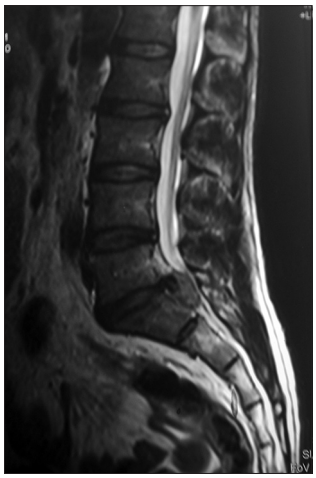

Figure1: Magnetic resonance imaging revealing the tumor arising from L5 nerve root

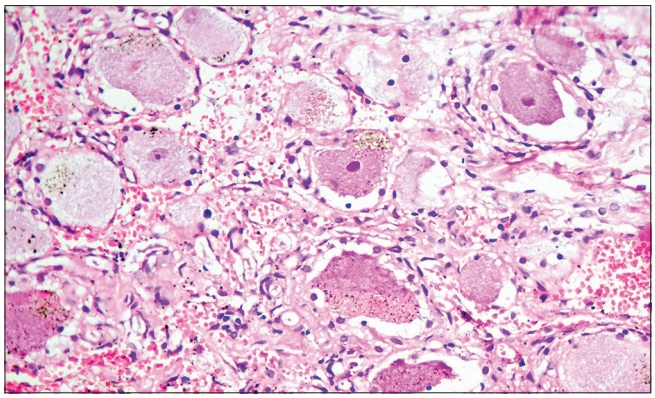

Figure 2: Photomicrograph revealing mature ganglion cells in a Schwann cell rich stroma, (H and E, ×40)

locations being the third ventricle and hypothalamus.[1] Most cases involve the paraspinal region with intraspinal extension extradurally through the neural foramen, leading to dumbbell-shaped tumors.[2] This also leads to extradural compression of the spinal cord.[2] Spinal ganglioneuroma occurs most frequently in the cervical spine, followed by thoracic spine and lumbar spine.[2] Ganglioneuroma occurring in the nerve root is very rare.[3-6] A few cases of ganglioneuroma occurring in the cervical nerve root have been reported.[4,5]

Ganglioneuroma occurs most frequently in children and young adults under the age of 30 and rarely in those older than 60 years.[7] It shows a slight female predominance.[7]

Clinical sign and symptoms of spinal ganglioneuromas vary depending on its location in the spine. Sometimes, they are detected incidentally on radiographic studies done for some other reasons as they are asymptomatic. There are usually signs of spinal cord compression such as motor and sensory deficits and/or bowel and bladder disturbances. Pain may be variable, and local pain is generally dull, but it could be burning and lancinating in extremities. Scoliosis has been reported in patients with dumbbell ganglioneuromas.[2,4]

Characteristic histological findings help to distinguish these tumors from the schwannoma, neurofibroma, or meningioma. In the differential diagnosis of extradural nerve root tumors, metastatic diseases, lymphomas, and Ewing's sarcoma also should be considered.[8-10]

The treatment of choice is surgical resection.[7] Adjuvant systemic chemotherapy and local radiotherapy have limited roles due to their benign biological nature.[7] The long-term prognosis is excellent regardless of tumor location as long as total tumor excision is performed.[7] 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.[7]

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