(International Print/Online Journal)

SJIF IMPACT FACTOR: 5.565
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume 6, Issue 2 , Page No: 696-698
March-April 2023

IJMSCR

Spinal Ganglioneuroma: A Rare Case Report

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Type of Publication: Case Report
Conflicts of Interest: Nil

Abstract

Ganglioneuromas are rare peripheral neuroblastic tumours derived from neural crest cells. They are well-differentiated benign neoplasms. We report an unusual case of ganglioneuroma found located in the lumbar spine extending into the extradural compartment invading the psoas muscle in 52-year-old man who presnted with lower back pain of 5 months duration. Radiology showed a heterogeneously hyperintense lesion L1-L2 right foraminal level, causing widening of the neural foramina and extending laterally into the right psoas muscle, splaying its fibers. Radiological features raised the suspicions of peripheral nerve sheath tumour, which later turned out to be a ganglioneuroma on histopathological examination.

Keywords: Ganglioneuroma, Peripheral neuroblastic tumour, Lumbar Spine

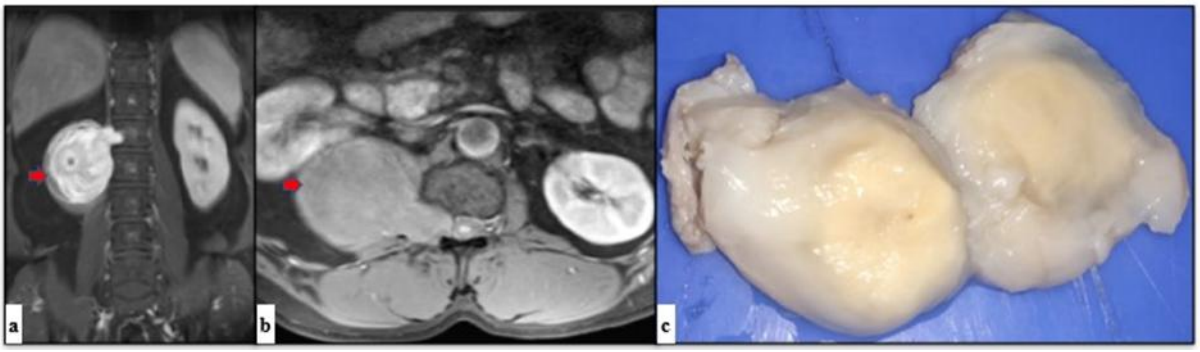
Introduction

Peripheral neuroblastic tumors originating from the neural crest cells, encompass a broad group of lesions including neuroblastoma, ganglioneuroblastoma and ganglioneuroma, characterized by their grade of neuroblastic differentiation and degree of schwannian stromal development. Ganglioneuroma, is a schwannian stroma dominant tumor, most commonly involving the adrenal glands and abdominal, pelvic or thoracic ganglia. [1] According to the International Neuroblastoma Pathology Classification (INPC), it has a favorable outcome irrespective of the patient's age at diagnosis. [1,2]

Case Report

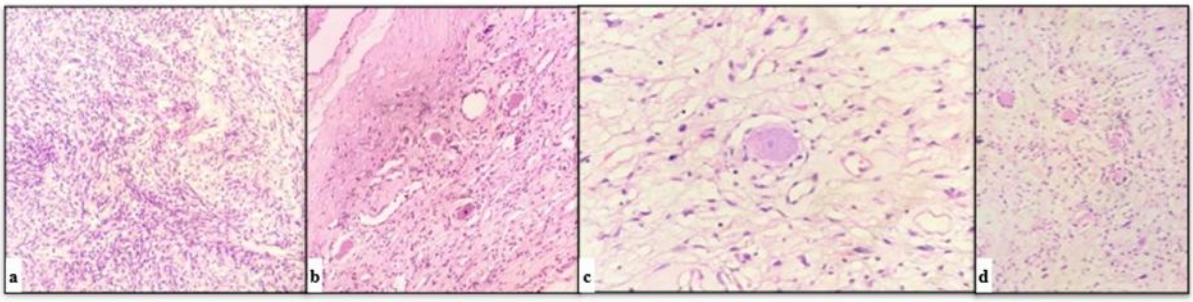
A 52-year-old male, came with complaints of lower back pain for 5months, which was insidious in onset, gradual in progression, aggravated by walking and relieved by rest. For the last 2months, the patient developed radiating pain to bilateral lower limbs. He was known case of systemic hypertension on treatment for the past 2 years. There was no history of trauma, fever, loss of weight or appetite. Local examination of lumbar spine showed a positive FABER test, complete range of motion against maximal resistance and intact sensations in the bilateral lower limbs. MRI Lumbar spine with contrast showed a dumbbell shaped extramedullary heterogeneously hyperintense lesion measuring 5.6x5.9.6.9cm epicentered at the level of L1-L2 right foraminal level, causing widening of the neural foramina and extending laterally into the right psoas muscle, splaying its fibers. (Fig.1a,b) These features were indicative of a nerve sheath tumor, probably schwannoma or neurofibroma. After obtaining due consent, patient underwent a posterior D12-L2 stabilization, L1 laminectomy, tumor and right L1 nerve root excision under general anesthesia. We received a globular encapsulated mass measuring 8x7x2cm, with a smooth external surface and a solid, pale yellow, translucent cut surface. (Fig.1c).

Fig.1 a), b) MRI Lumbar spine with contrast showing a dumbbell shaped extramedullary lesion at L1-L2 right foraminal level c) Gross picture of the excision specimen



Microscopic examination of the haematoxylin and eosin-stained formalin fixed paraffin embedded tissue sections revealed a well circumscribed capsulated neoplasm composed of cells with scant to moderate eosinophilic cytoplasm and uniform elongated, spindled, wavy nuclei arranged in fascicles separated by areas of myxoid and hyalinised stroma and interspersed lymphocytes, few mast cells and occasional macrophages. (Fig.2a) Focal areas in the periphery of the lesion show scattered mature ganglion cells with abundant eosinophilic cytoplasm and uniform round nuclei surrounded by neurofibroma like stroma. (Fig.2b,c,d) Adjoining areas show unremarkable nerve fibres. (Fig.2b) There is no evidence of necrosis or mitotic activity. Based on the above histopathological findings a diagnosis of ganglioneuroma was made.

Fig.2 a) Schwannian stroma b), c), d) Scattered ganglion cells in a neurofibroma like stroma



Discussion

Ganglioneuromas are rare, slow growing benign mature neuroblastic tumors. They are mostly diagnosed in children, rarely affecting older age groups. They account for 0.1-0.5% of the CNS tumours. [3] Approximately 10% ganglioneuromas affect the spinal canal. Paraspinal tumors present as dumbbell shaped masses extending extradurally into the ipsilateral spinal canal through the neural foramina. [4,5] Spinal ganglioneuromas commonly present with back pain, weakness, and paresthesia due to mass effect. [3,4] Few cases of ganglioneuroma have been reported to present with symptoms related to aberrant hormone secretion from the tumour. [6,7] Rarely, ganglioneuromas can be associated with neurofifibromatosis type-1 (NF-1) and multiple endocrine neoplasia (MEN) 2B. These patients should hence be tested for these disorders, especially if there is significant family history. [8] Total surgical resection remains the best treatment option for symptomatic solitary ganglioneuroma with a favorable prognosis in the long-term. [9,10] Apart from typical histopathological features of ganglioneuroma, IHC can also help to differentiate it from other neurogenic tumors like schwannoma and neurofibroma. Nerve fibers and Schwann cells in ganglioneuroma, like the other two tumors are S100+. [1] NSE and NF proteins are positive in mature ganglion cells whereas immature ganglion cells are synaptophysin+ and NF negative. [11]

Conclusion

Ganglioneuromas located in the paraspinal region with extradural intraspinal extension through the ipsilateral neural foramina are a rare occurrence. Their diagnosis requires comprehensive

Volume 6, Issue 2; March-April 2023; Page No 696-698
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histopathological analysis along with clinico-radiological correlation. Thus, ganglioneuroma should be included in the differential diagnosis of a para-spinal extradural mass.

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