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Lumbar neurofibroma: aetiology, clinical presentation, surgical indications, and surgical technique. A focused view about surgical experience in Viseu, Portugal

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Keywords
neurofibroma, lumbar, root, plexus

ABSTRACT

Lumbar neurofibroma is a benign neoplasm that originates from peripheral nerves, specifically from Schwann cells. While it can occur at various anatomical sites, its manifestation in the lumbar region has specific clinical and neurosurgical aspects, which are of great relevance for accurate diagnosis and effective treatment. This article addresses the causes, clinical presentation, indications for surgical treatment, and the surgical technique applied to lumbar neurofibromas, based on key studies and articles published in the last ten years.

ETIOLOGY AND CAUSES

Neurofibromas can be classified as either sporadic or hereditary, with the latter often associated with Neurofibromatosis Type 1 (NF1), an autosomal dominant condition that results in multiple neurofibromas across various parts of the body. NF1 is caused by mutations in the NF1 gene located on chromosome 17, which codes for neurofibromin, a tumor suppressor protein. Patients with NF1 are predisposed to developing neurofibromas in various areas, including the spinal column, particularly in the lumbar region [1,2].

On the other hand, sporadic neurofibromas have no identifiable genetic cause and can occur at any age, with higher prevalence in young adults [3]. Pathogenesis involves abnormal proliferation of Schwann cells, fibroblasts, and other peripheral nervous system cells.

CLINICAL PRESENTATION

The clinical presentation of lumbar neurofibromas can vary significantly depending on the size, location, and compression of adjacent structures, such as nerve roots and the spinal cord. Common symptoms include low back pain, radiculopathy, sensory loss, or

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muscle weakness, with patients often affected by compression of spinal nerves exiting the lumbar spine [4].

In more severe cases, spinal cord compression can lead to more significant neurological symptoms, such as paralysis, urinary and fecal dysfunction, and loss of reflexes. Pain is a predominant symptom, which may be continuous or intermittent, often exacerbated by movement of the spine [5].

Additionally, patients with NF1 may present characteristics associated with the syndrome, such as café-au-lait spots and Lisch nodules, which can assist in the differential diagnosis [6]. Magnetic resonance imaging (MRI) is the imaging modality of choice for evaluating lumbar neurofibromas, enabling the identification of lesion extent, compression of surrounding structures, and the type of tissue involved.

Surgical indication

The main indication for surgery in cases of lumbar neurofibroma is the presence of progressive neurological symptoms or failure of conservative treatment. Surgery is indicated when there is evidence of nerve root or spinal cord compression causing debilitating pain, neurological deficit, or functional impairment [1].

Furthermore, surgical removal is recommended in symptomatic neurofibromas that interfere with the patient's daily activities or when malignancy is suspected, a rare but possible condition that can occur in long-standing neurofibromas. The evaluation of malignancy is based on clinical and histological characteristics, with malignant transformation to neurofibrosarcoma being a severe complication that requires urgent intervention [7].

Surgical technique

The surgical approach to lumbar neurofibromas must be carefully planned to minimize the risk of neurological injury and maximize tumor removal. The standard procedure involves lumbar laminectomy or extended laminectomy to ensure access to the neurofibroma, which is often located in extradurally or paraspinal regions. The choice of laminectomy type depends on the tumor's location and size [8].

The initial step in surgery involves careful exposure of the lumbar spinal structures and removal of the vertebral lamina, followed by identification and dissection of the neurofibroma. In cases of intradural neurofibromas, a more invasive approach may be necessary, with manipulation of the dura mater and nerve roots. The neurofibroma should be excised as completely as possible without compromising the integrity of the nerve roots, with attention to prevent damage to the spinal cord.

In some cases, intraoperative neurophysiological monitoring (IONM) may be helpful to protect neurological structures during excision, especially in tumors located in difficult-to-access areas [2]. After tumor resection, the lumbar spine may be stabilized, if necessary, with instrumentation, depending on the extent of bone removal. The goal is to ensure functional recovery without additional neurological deficits.

In our series in two cases mini retroperitoneal approach was uses for tumor removal and in one case the tumor was approached through extended laminectomy.

Postoperative considerations

Postoperative management of patients undergoing lumbar neurofibroma resection includes pain control, neurological monitoring, and early physiotherapy to promote mobility recovery. Full recovery may vary depending on the severity of preoperative symptoms and the extent of surgery. In NF1 cases, continuous monitoring is important to detect the development of new neurofibromas over time [4].

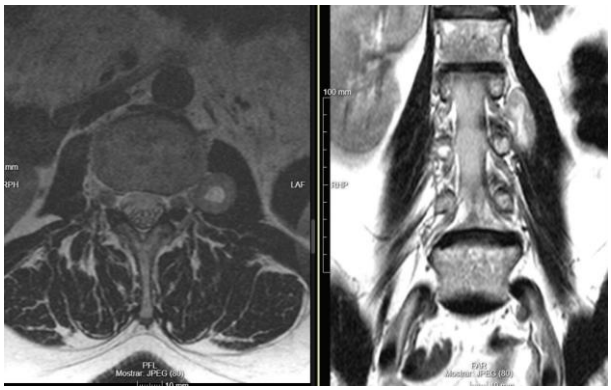


Figure 1. MRI extra canalar, intrapssoas muscle tumor, arising from left side L2 root.

Case 1.

59 years old lady with history of renal TB treated, later left side nephrectomy due to lithiasis, presenting left side radicular pain L3 treated

conservatively with no effect. Lumbar MRI showed extracanalar, intrapssoas muscle tumor, arising from left side L2 root (Fig. 1). She was operated through left side mini retroiperitoneal approach with total removal of the tumor with resolution of complaints. Two years after surgery she is doing well, MRI revealed no tumor recurrence (Fig. 2).

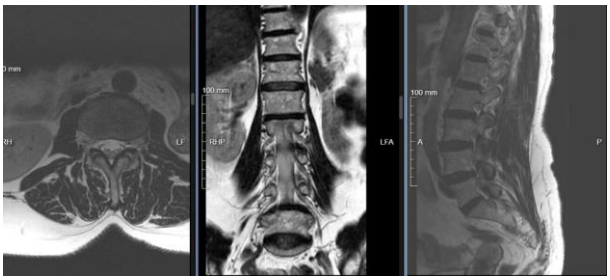
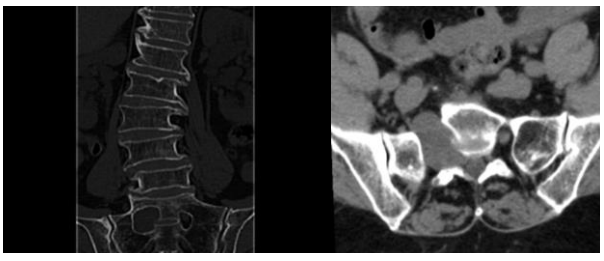


Figure 2. Postoperative MRI 2 years after surgery showing no recurrence of tumor.

Case 2.

73 years old lady with a long lasting hystory of right side ciatic pain, worthened by association of progressive neurologic claudication. CT scan showd severe L5-S1 stenosis and sacral errosion with enlagement of right side S1 foramina by a tumor arizing from extracanalar para of S1 root with evolution to presacral, retroperitoneal space (Fig. 3). She was submitted to psterior decompression of low lumar spine with fixation with transpedicular screws and S1 tumor total removal. 4 years after surgery she is doing well, complaining of numbness of righr leg, independent, using sporadically pain killers. CT scan 4 years after surgery shows no recurrence and the tumor bad is filled with bone (Fig. 4).



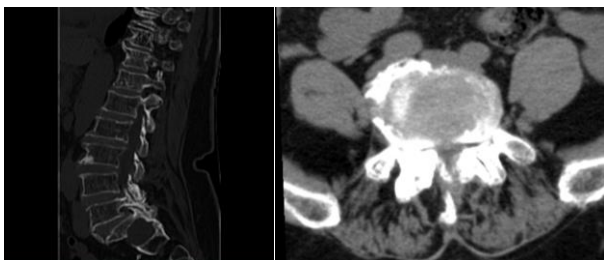


Figure 3. CT scan revealing enlargement of right-side S1 foramina by huge tumor, associated by L5-S1 severe central canal stenosis.

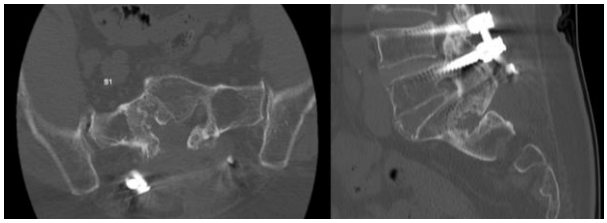


Figure 4. SC scan 4 years after surgery, partial bone filling of the tumor bed.

Case 3.

45 years old lady with complants of right side L2 pain with few effect with pain killers. MRI found left-side intrapssoas tumor adjacent to L3 root (Fig. 5). She was operated through left-side mini retroperitoneal approach with total removal of tumor and after two year MRI revealed no recurrence (Fig. 6).

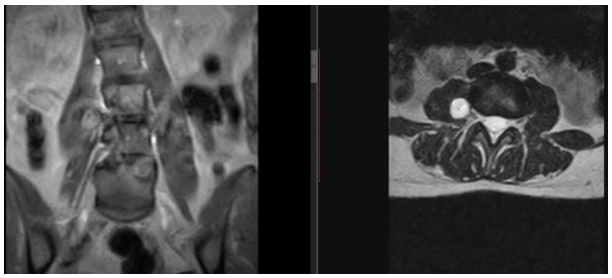


Figure 5. MRI right-side extra canalar, intrapssos tumor razing from L3 root.

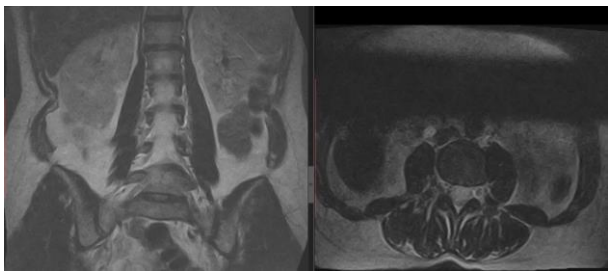


Figure 6. Postoperative MRI, 2 years after surgery, showing no recurrence of tumor.

Conclusion

Lumbar neurofibromas represent a significant clinical and neurosurgical challenge, requiring early diagnosis and appropriate management to prevent neurological complications. Surgical intervention is crucial in symptomatic cases, with tumor resection

being the most effective strategy to alleviate symptoms and improve the quality of life of patients. Modern intraoperative monitoring techniques and spinal stabilization options contribute to a high surgical success rate.

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