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# Bilateral plexiform neurofibromas of the brachial and lumbosacral plexuses

Neurofibroma plexiforme bilateral dos plexos braquial e lombossacral

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Neurofibromatosis type 1 (NF-1) is one the most common autosomal dominant conditions affecting the nervous system<sup>1</sup>. Plexiform neurofibroma (PN) is a benign tumor that basically defines the diagnosis of NF-1. These entities consist of multiple, twisted masses that grow along the axis of a large nerve, infiltrating and separating normal nerve fascicles. Malignant transformation is the main associated complication. Surgery is frequently reserved until PNs have progressed to the point of causing functional compromise, esthetic deformity or pain<sup>2</sup>. This paper describes a case of plexiform neurofibromas of the brachial and lumbosacral plexuses, with presentation of the clinical and radiological findings.

## CASE REPORT

An 18-year-old female patient was referred to a neurosurgery reference service with a history of mass lesion in the left cervical region, progressing to the right cervical region two months after the onset of symptoms. Physical examination revealed a nodule, painless upon palpation, in both cervical regions (Fig 1). “Café au lait” spots were seen on the patient’s back and cutaneous and subcutaneous neurofibromas were seen over the extremities. The patient reported family history of NF-1, but she had not undergone any clinical follow-up. Neurological examination was normal.



Fig 1. Color photograph showing left-sided neck mass.

Magnetic resonance imaging (MRI) of the brachial plexus was solicited and revealed the T2 weighted sequence, multiple, hyperintense, lobular, solid, expansive tumors in the distribution of the brachial plexus bilaterally extending to the anterior mediastinum and invading the spinal canal (Fig 2). Lumbosacral plexus MRI showed impairment of both plexuses and suggested the presence of a tumor toward the lumbosacral neural foramina (Fig 3) and its extension to the pelvic region (Fig 4).

An incisional biopsy was performed on the left cervical tumor and the histological findings were compatible with neurofibroma.

During the four-year follow-up involving imaging exams, the patient had been asymptomatic and free of neurological deficit. Thus, the decision was to maintain conservative management.

## DISCUSSION

PN is a common manifestation of NF-1. This entity may be classified as a benign peripheral nerve sheath tumor that involves multiple nerve fascicles or branches of major nerves<sup>3</sup>.

MRI is the method of choice for the diagnosis and follow-up of neurofibromas. In T1 weighted image, a PN is isointense in relation to muscle tissue. In T2 weighted sequence, a PN is seen as a hyperintense, homogeneous image with “target sign”, owing to the central fibrous tissue surrounded by myxoid tissue<sup>4</sup>.

Malignant transformation is the leading complication of PN. The lifetime risk of this transformation is estimated at 15 to 20% in patients with NF-1<sup>5</sup>. Clinical signs of malignant degeneration include the emergence of consistent pain, neurological deficit and an increase in the tumor mass. In the present case, the patient did not present clinical signs of malignant transformation throughout the entire follow-up.

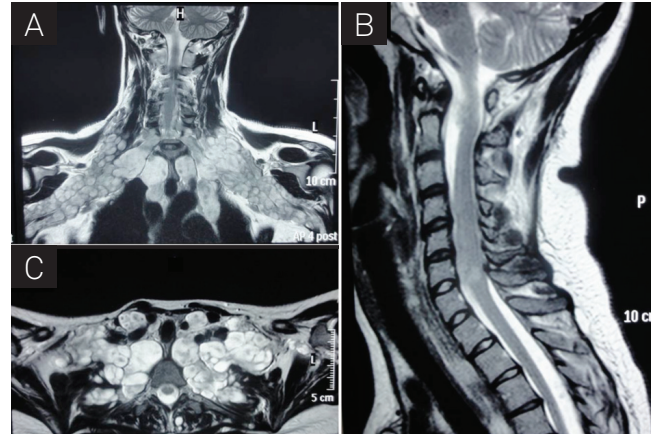


Fig 2. (A) Coronal T2 weighted magnetic resonance imaging showing the brachial plexuses bilaterally irregular, enlarged and hyperintense, resembling a “bag of worms”. (B) Sagittal T2 sequence shows the extension of PNs into the spinal canal with indentation of the spinal cord at the C3–C6 level. (C) Axial T2 weighted magnetic resonance imaging showing many hyperintense nodules in the cervical paravertebral space.

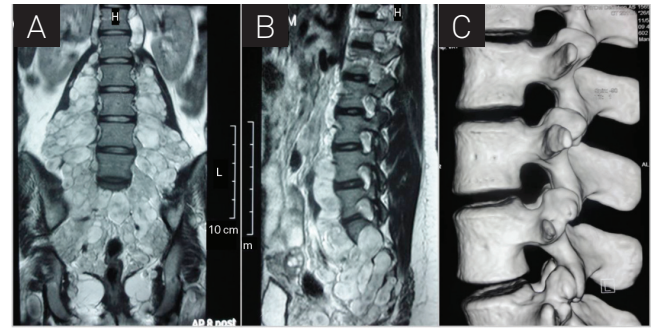


Fig 3. (A) Coronal T2 weighted MRI showing extensive PNs extending along the course of multiple nerve roots. (B) Sagittal T2 weighted MRI of the lumbar spine showing a large mass with multiple areas of low signal intensity centrally and high signal intensity peripherally. This finding is referred to as the target sign. (C) 3D CT scan reconstruction showing widening of the neural foramina at multiple levels of the lumbar spine.

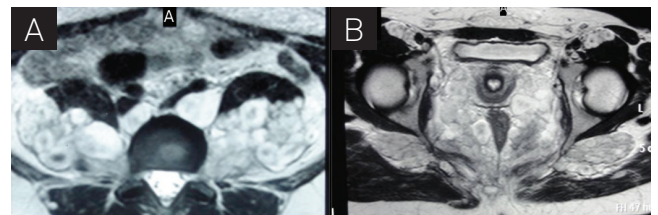


Fig 4. (A) Axial T2 weighted MRI of the lumbar spine showing a large mass in the lumbar paravertebral space. (B) Axial T2 image showing deep, nodular neurofibroma of sciatic nerve in pelvic and gluteal region.

The treatment of PNs in patients with NF-1 consists of regular follow-up examinations involving serial physical evaluations and neuroimaging studies<sup>2</sup>. This was the conduct used in the present case.

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