Bilateral spinal neurofibroma

see also Cervical bilateral spinal neurofibroma.

Neurofibromatosis type 1 associated with wide, symmetrically distributed spinal neurofibromas are not common. Furthermore, occurrence is still rarer in members of the same family in familial spinal NF1 (FSNF1) [1].

Retroperitoneal plexiform neurofibromas have a characteristic appearance on CT scans. They are typically bilateral, symmetric, low-attenuation masses in a parapsoas or presacral location. Asymmetry in size and attenuation of a larger mass suggests the possibility of a malignant tumor of the nerve sheath. Recognition of the CT features of a retroperitoneal plexiform neurofibroma can obviate the expense, pain, and risk of an unnecessary biopsy. Conversely, detection of findings suggestive of malignant tumor can lead to appropriate recommendation of biopsy [2].

Pascual-Castroviejo et al. present 8 patients (5 M and 3 F) with sporadic or non-familial spinal neurofibromatosis 1 (non-FSNF1) associated with bilateral spinal neurofibromas involving all of the paraspinal nerves.

This is the first series of such association described in the literature. Their ages ranged from 6 months to 20 years (average 9.8 years) at the time of radiological diagnosis. This presentation appears to be earlier than in familial spinal neurofibromas in NF1 (FSNF1). Predisposition to malignancy probably is greater in the non-FSNF1 type. MRI studies were performed routinely in all patients with NF1 and these were complemented with MRI enhanced with gadolinium and repeated at different ages in cases with paraspinal tumors. Coronal views provided the best evidence for the presence of neurofibromas in every spinal nerve. The size of the tumors and the clinical complications increased with advancing age in most patients. Giant plexiform tumors were often seen in the cervico-thoracic region. Malignant peripheral nerve sheath tumors (MPNST) were found in one patient with a sciatic tumor and another patient died suddenly at home without necropsy or pathological study. Voluminous paraspinal neurofibromas can be at risk for malignancy. More frequent neuroimaging studies may be necessary for an earlier detection. Early surgical treatment to anticipate the occurrence of MPNST during surveillance could be an option. Bilateral spinal neurofibromas are found in both patients who inherited the NF1 and in those due to de novo mutations [3].

1)
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2)
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3)
Pascual-Castroviejo I, Pascual-Pascual SI, Viaño J, Velazquez-Fragua R, López-Gutierrez JC. Bilateral spinal neurofibromas in patients with neurofibromatosis 1. Brain Dev. 2012 Aug;34(7):563-9. doi: 10.1016/j.braindev.2011.09.011. Epub 2011 Oct 14. PubMed PMID: 21999966.