Plexiform neurofibroma

Uncommon variant of neurofibroma.

Benign tumour of peripheral nerves (WHO grade I), arising from a proliferation of all neural elements.

Plexiform neurofibromas are essentially pathognomonic of neurofibromatosis type 1 (NF1). Unlike small sporadic localised neurofibromas and diffuse cutaneous neurofibromas.

Plexiform neurofibromas usually presents with symptoms due to large size and site [1].

Treatment

Plexiform neurofibroma treatment

Outcome

These tumours are at a significant risk of eventual malignant transformation.

Tonsgard et al. examined the incidence and radiologic characteristics of plexiform neurofibromas in neurofibromatosis-1 (NF-1) to define a cohort at greatest risk for malignant nerve-sheath tumors.

Plexiform neurofibromas are a frequent complication of NF-1. They can impair function, produce disfigurement, and be the site for the development of malignant nerve-sheath tumors. The incidence and natural history of plexiform neurofibromas is unknown.

CT imaging of the chest, abdomen, and pelvis was performed in 91 of 125 consecutive adults (age, > or = 16 years) with NF-1.

Twenty percent of patients had plexiform neurofibromas of the chest in the paraspinal, mediastinal, or supraclavicular area. Approximately 40% of patients had abnormal abdominal/pelvic scans. The paraspinal, sacral plexus, sciatic notch, and perirectal regions were the most common sites. Most plexiform neurofibromas were asymptomatic. Imaging also revealed a number of tumors, including malignant nerve-sheath tumors, adrenal tumors, carcinoids, and schwannomas.

The frequency of plexiform lesions and other tumors in NF-1 indicates that clinicians should monitor young adults carefully; however, imaging characteristics alone cannot reliably distinguish benign from malignant lesions [2].

Cervical cord compression from cervical root neurofibromas represents an important clinical problem in patients with neurofibromatosis type 1 (NF1), but is rarely reported. The aim of this study was to describe the clinical presentation and follow-up of children and adults with NF1 and cervical cord

compression. A retrospective review of clinical records and neuroimaging studies from two large tertiary care centres between 1996 and 2006 was performed. 13 patients with NF1 and cervical cord compression were identified. Age at presentation ranged from 9 to 61 years. The most common presentation was progressive quadriparesis. 11 of 13 patients underwent cervical decompression and subtotal resection of the associated neurofibroma. The majority of patients had recovery of neurological function and no further clinical progression. Progressive neurological deficit (typically quadriparesis), rather than neuroimaging appearances, should dictate the need for surgery [3].

1)
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