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Artículo de revisión

Giant plexiform neurofibroma: an intraspinal dumbbell tumour with a huge retroperitoneal portion

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NEUROFIBROMA PLEXIFORME GIGANTE . UN TUMOR EN RELOJ DE ARENA CON UNA PORCIÓN GRANDE RETROPERITONEAL

RESUMEN

Una mujer de 32 años de edad con historia de un año de dolor lumbar y dos semanas de agudización con dolor irradiado a la extremidad pélvica derecha fue admitida en nuestro servicio. Los hallazgos radiológicos revelaron la presencia de una masa de apariencia tumoral retroperitoneal y parapélvica originándose de una raíz nerviosa lumbar. Se realizó tratamiento quirúrgico combinado, transabdominal y mediante laminectomía estándar, el tumor se resecó en su totalidad. En el posoperatorio la paciente mostró mejoría de los síntomas sin mostrar déficit alguno. El reporte histopatológico confirmó un neurofibroma plexiforme. El seguimiento posoperatorio a 24 meses muestra estudios radiológicos sin tumor residual y sin recidiva tumoral. El caso y su manejo son discutidos en el presente artículo.

Palabras clave: neurofibroma plexiforme, tumor retroperitoneal, raíz nerviosa lumbar, dolor lumbar.

ABSTRACT

A 32-year-odl-woman with a one year history of lower back pain and two weeks of worsening of pain in the right tight was admitted. Radiological findings revealed an extremely rare retroperitoneal and parapelvic tumour mass originating from a lumbal nerve root. A combined surgical treatment was performed transabdominally and through standard laminectomy and the tumor was totally resected. Following surgery the patient showed symptomatic improvement and she did not show any deficit. The histopathological report confirmed a plexiform neurofibroma. Follow-up investigations 24 months after surgery did not reveal any residual tumour. The case and its management are discussed.

Key words: plexiform neurofibroma, retroperitoneal tumour, lumbar nerve root, lumbar pain.

Primary retroperitoneal tumours are rare and present an astonishing variety of histological features. These tumours are associated with few characteristic clinical signs such as increase in abdominal size, irradiating pain in the lumbar region or in the back, and thus are usually diagnosed after they have reached an appreciable size[1,2].

Nerve sheath tumours of a large size such as plexiform neurofibromas or neurofibrosarcomas are often a peripheral form of neurofibromatosis[3]. Unlike solitary neurofibromas, which can be completely resected with minimal morbidity[4], plexiform neurofibromas are characteristically multiple and may involve any or all of the following sites: cranial and spinal nerve roots and ganglia, major nerves of the neck, trunk and limbs, including the sympathetic system, subcutaneous branches of major nerves and visceral sympathetic plexuses[5].

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We present a case of a plexiform neurofibroma with considerable retroperitoneal component as well as intraspinal extension and discuss the specific diagnostic and surgical procedures.

CASE REPORT

A 32-year-old woman with a one-year history of dull lower back pain came to hospital admission. She complained about increasing pain in the right side spreading into the groin and thigh. Her clinical and family history was inconspicuous for neurofibromatosis (von Recklinghausen's disease). Clinical examination revealed only hypaesthesia of the right thigh according to the dermatome L2 and L3. No cutaneous nodules or features of neurofibromatosis were detected. Abdominal palpation disclosed a huge solid mass in the right side and in the renal bed.

Radiological evaluation

Intravenous urography confirmed a calyceal dilatation and unilateral hydronephrosis due to a marked tumorous ureteric compression (figure 1). Computed tomography (CT) showed a hypodense, well demarcated space occupying process shifting the right kidney laterally (figure 2a, b). MRI revealed besides the retroperitoneal and parapelvic mass (10 x 9 x 11 cm) a solid intraspinal tumour (1.5 x 1.5 x 1.2 cm) with low signal intensity on T1-weighted images (not shown) and irregular enhancement of the tumour (figure 3a-e). Heterogeneous areas of increased and decreased signal intensity were obtained on T2-weighted images.

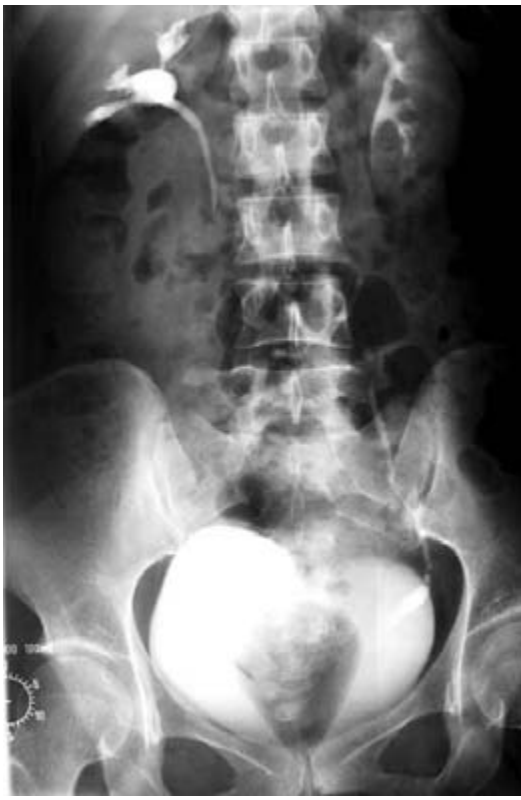

Figure 1. Excretory urography revealing a calyceal dilatation and unilateral hydronephrosis due to a marked tumorous compression.

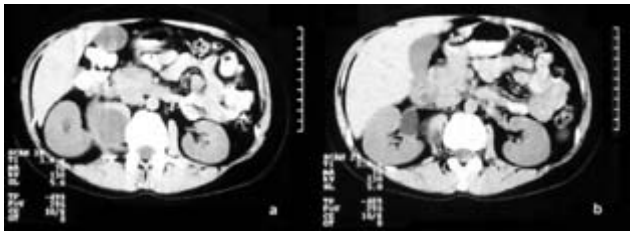

Figure 2. Unenhanced axial CT - scans **(a, b)** demonstrating a round, well-de-lineated retroperitoneal mass shifting the right kidney cranially and lat-erally.

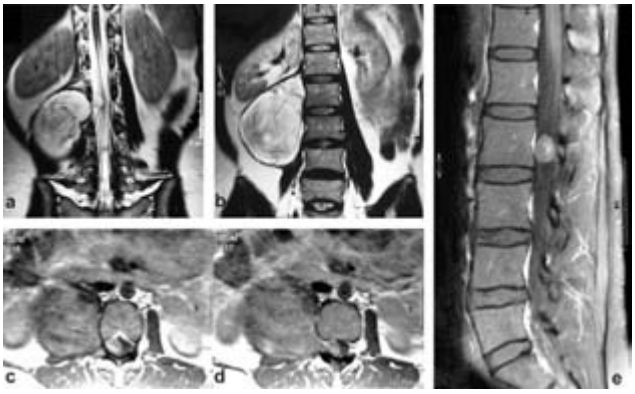

Figure 3. Coronal preoperative **(a, b)** T2-weighted MR-Images **(TR 2700, TE 150)** revealing the huge retroperitoneal and the intraspinal tumour. Axial, Gd-DTPA-enhanced T1-weighted MR-images **(TR 944, TE 12)** showing an extensive paraspinal and intraspinal plexiform neuro-fibroma **(c, d)**. Sagittal **(e)** Gd-DTPA enhanced T1-weighted MR-images **(TR 585, TE 12)** exhibit an intraspinal tumour at the level of L2.

OPERATION

Multidisciplinary input was essential to perform surgery. In supine position the extraspinal tumour was ressected transabdominally up to the region of the neural foramen. Because it was impossible to identify involved nerve fascicles and network of autonomic nerve filaments (renal plexus), the entire tumour was transsected at the foraminal level, marked by metal clips and totally removed.

Then in prone position, a standard laminectomy at L1 and L2, was performed to expose the intradural

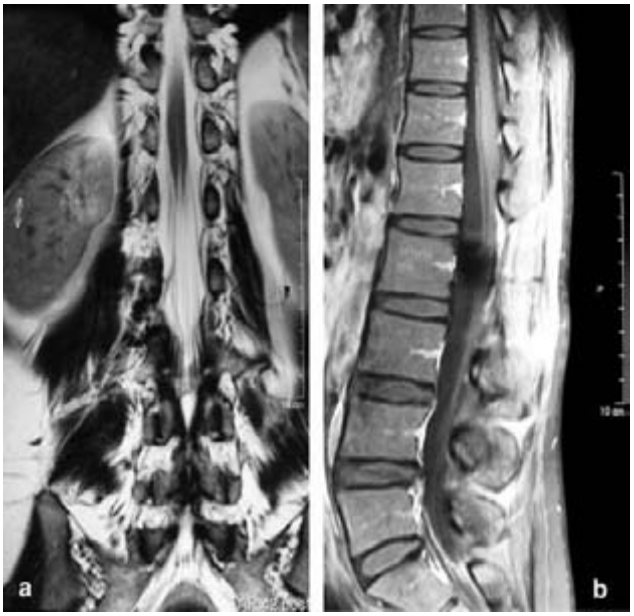

Figure 4. Postoperative coronal **(a)** T2- weighted image **(TR 2700, TE 150)** and sagittal Gd-DTPA enhanced **(b)** T1-weighted MR-images **(TR 540; TE 12)** confirmed a complete tumour resection 24 months after surgery.

tumoural mass. In contrast to the retroperitoneal tumour, which was firm, the intradural portion was soft allowing the use of the ultrasonic aspirator. The nerve roots emanating from the conus medullaris were delineated, the tumour was debulked to permit visualisation of the nerve roots, uninvolved rootlets were preserved and the resection was continued laterally toward the nerve root sleeve up to the neural foramen.

Postoperative neurological examination showed no paresis, hypaesthesia or impaired urinary function. Follow-up examination (MRI) 24 months later revealed complete tumour resection (fig. 4 a-b).

HISTOPATHOLOGICAL FINDINGS

The retroperitoneal tumour mass was greyish coloured with a shiny surface incorporating nerve rootlets with a cylindrical enlargement. The extra and intraspinal tumour was composed of neoplastic Schwann cells, perineurial-like cells and fibroblasts in a matrix of collagen fibres and mucosubstances. Extensive collagen formation took the form of characteristic bundles. Mitotic figures were absent; labelling indices for the proliferation marker (Ki 67) are below 1%. Immunohistochemical analysis for neurofilament revealed axonal structures indicating an infiltration of nerve fascicles histopathological diagnosis confirmed a plexiform neurofibroma.

DISCUSSION

Neurofibromas are common tumours of peripheral nerves[3,5] and can be found sporadically as solitary tumours unrelated to any apparent syndrome or as part of Recklinghausen's disease (neurofibromatosis type 1)[6]. Both types of neurofibromas show an identical histological pattern consisting of Schwann-cells, fibroblasts and perineural like cells. The affected nerve rootlets display an irregular enlargement, from which the term «plexiform» was originally derived. It is important to stress that «plexiform» does not imply involvement of a nerve plexus but implies a network-like growth of neurofibroma involving multiple fascicles of a nerve or multiple branches of a large nerve[7]. When the tumour becomes larger, degenerative involution occur, haemorrhage and cystic degeneration due to vascular thrombosis and subsequent necrosis.

An intraspinally localised plexiform neurofibroma is a rare entity[1] as described in our patient. The clinical history is non-specific with irradiating lumbar pain as leading symptom [2].

The radiological aspect depends on size, large tumours are characterized by degenerative changes (fig. 2 and fig. 3a, b) and pseudocystic areas[8]. MR-images of neurofibromas show different signal intensity characteristics, including hypointense or intermediate intensity on T1-weighted images and inhomogeneous enhancement[9] (Fig. 2c). On T2-weighted images the high-intensity regions in the periphery correspond to myxoid degeneration and a low intensity to collagenous fibrous tissue[9].

Surgery is the usual therapy of these tumours. Controversy exists concerning the degree of aggressive tumour resection[4]. Early surgery of a benign tumour may lead to permanent damage of nerve tissue but late excision could be incomplete and may result in missed eradication of a coexistent malignant tumour.

Resection of important nerve roots in direct contact with the tumour can be attempted, because nerve fibres involved in a neurofibroma probably have no function at all[10]. In our case, the resection of the L2 root was not followed by any loss of sensory or motor deficit. This can be explained by a functional compensation as a result of epispinal axons[11] or an overlapping innervation. However, in small tumours of really important roots (e.g. L4, L5 and S1) immediate resection is questionable. In our opinion, asymptomatic lesions that exhibit only foraminal encroachment rather than compression of important roots or of the spinal cord, should be closely monitored radiologically (MRI). Resection is favoured for lesions that are

either enlarging rapidly or causing progressive symptoms.

We strongly advocate interdisciplinary efforts and a radical tumour resection with meticulous dissection of incorporated roots.

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