Abstract

Large series of cauda equina tumors in adults are seldom reported. This French series retrospectively reviews 231 cases collected for the congress of the Société Française de Neurochirurgie in 1996. The authors first analyze this series and then discuss the pertinent literature. Schwannoma was the most frequent benign tumor in this series, followed by ependymoma. Very few malignant tumors were recorded; these were usually malignant neurinomas nearly always in neurofibromatosis patients. Some other rare tumors were also recorded, including paragangliomas. This series confirms the importance of the pretherapeutic neurological status in functional prognosis. All schwannomas can be cured, while ependymomas and paragangliomas may recur after a very long delay. Surgery must be as complete as possible, since adjuvant therapies are proven to have little efficacy. This type of tumor requires very long follow-up. Prognosis is good for hemangioblastomas. Sphincter dysfunctions carries a poor prognosis and may appear after primary surgery, more often after treatment of recurrences.

Key words Cauda equina tumors · Surgery · Functional outcome

The only previous attempt from that viewpoint was by Freeman and Cahill [1]. As most of these tumors are of benign type, total surgical removal is of utmost importance for these patients. Their reputation for being easy to manage is not sound.

The congress of the Société Française de Neurochirurgie in Poitiers provided an opportunity for reviewing the experience of most neurosurgical units in our country. There were 231 cauda equina tumor cases collected, and the report of this congress was published in 1999 [2].

Definition of cauda equina tumors

These tumors develop from the constituting structures of the cauda equina: filum, nerve sheaths, and other intrinsic tissular structures including embryologic structures. Some authors include other types of tumors arising from other structures – meningeal envelopes, epidural structures, and even bony tumors or intrathecal metastases – because they may be revealed by a cauda equina syndrome. In some cases, the histological diagnosis of “true” cauda equina tumor is made only after pathological examination. True cauda equina tumors are:

1. Benign neurinomas
2. Malignant neurofibroma
3. Ependymomas
4. Paragangliomas
5. Hemangioblastomas
6. Lipomas
7. Lymphomas
8. Epidermoid cysts and teratomas
9. Capillary hamartomas

Some other tumors can present as cauda equina tumors: those arising from envelopes (which are a differential diagnosis), astrocytomas (which are similar but in most cases involve only the conus medullaris), spinal epidural metastases, bony tumors, and rarely chordomas.

Introduction

Cauda equina tumors are rare and have not been reviewed for a long time. In neurosurgical literature, these tumors are generally considered from a pathological rather than anatomical or topographical point of view.
Methodology

A questionnaire was sent to our neurosurgical colleagues regarding clinical symptoms, delay from first symptom to diagnosis, imaging workup, treatment modalities, difficulty of treatment, follow-up, and recurrences. Among the responses to the questionnaire were 77 descriptions of tumors causing cauda equina compression, the final diagnosis coming only after pathological examination.

Details of operative techniques were not investigated and nor was CSF study since, with the general use of MRI and CT scan, lumbar puncture is not routinely performed. Statistical analysis was done by Professor Ingrand (Poitiers Medical School of Medicine) using SAS system software.

The most numerous tumors in this series were neurinomas (114) and ependymomas (79). The large numbers of these two most frequent tumors led the authors to define clinical profile, imaging features, ideal management, and prognosis for both. The authors first detail each type and then present the general features of the whole series.

Profile of neurinomas (n=114)

Revealing symptoms

Low back pain was suffered by 60.4% of all patients but only 55.26% of neurinoma patients, and 92.9% of these suffered from radiculopathy, instead of 80.7% for the whole series. Only 2.63% of neurinoma patients had recent constipation. Urinary retention or overflow incontinence existed in 8.77% of these patients, and sexual dysfunction was noted in 4.39% (13.3% for the whole series).

Physical examination

Of all the neurinoma patients, 46.49% presented with limited spinal motion. Paravertebral spasm was found in 24.5%. Abolition of one or several reflexes was noted in 29.82% of cases and motor deficit in 34.18%. Saddle anesthesia was rare in neurinomas – 2.63% bilaterally and 0.88% unilaterally. In the presence of unilateral amyotrophy, there was no statistically significant difference between neurinomas and ependymomas.

Neuroradiologic findings in neurinomas and neurofibromas

Plain lumbosacral X-rays can show a scalloping at the posterolateral aspect of the vertebral body. Posterior wall erosion was found in 21% of neurinomas. When the neurinoma is of hourglass shape and develops simultaneously in the intra- and extradural spaces, enlargement of the homologous foramen may be visualized.

Myelography shows a curved lacunar image or a curved arrest. Tumor boundaries can be multilobulated.

On computed tomography (CT) scan, the densities of neurofibromas and neurinomas are close to that of muscle, but contrast medium alone is responsible for enhancement in schwannomas, thus helping to distinguish them from neurofibromas, which exhibit almost no enhancement. In many cystic variants, one can note densities close to those of CSF. On myelo-CT scan, neurinomas present lateral development from a root and shift the other roots of the cauda equina contralaterally. This appearance allows the distinction from tumors arising from the filum, which displace the roots of cauda equina on the right and left symmetrically.

In magnetic resonance tomography (MRI), neurinomas and neurofibromas both exhibit homogeneous enhancement, in contrast to CT. Contrast medium injection allows better discrimination of necrotic or cystic regions. In particular, MRI visualizes lesions in the upper lumbar region (primarily L2), the most frequent localization of lumbar neurinomas (Figs. 1, 2).

Treatment

Complete removal was achieved in 99.12% of neurinomas.

Outcome

An aid for walking (cane) was necessary in 3.5% of neurinoma cases. Sphincter dysfunctions were present in 5.62% of patients at 6 months and 3.5% at 12 months, worsening to 5.25% by the end of follow-up. Sexual dysfunctions were involved in 0.8% of neurinomas and appeared during recurrences.

Recurrences

The frequency of recurrences varied according to tumor type: only one neurinoma recurred (0.88% of neurinomas), in a type I neurofibromatosis patient. This patient was reoperated.

Neurinomas, affecting adults of both sexes about equally, carry an excellent prognosis. Mean patient age was 50 years. The delay from onset of first symptom to diagnosis was 24 months. There is no specific clinical presentation, but low back pain and radiculopathy with a mild deficit are in the foreground. Night-time predominance is not always present. Sphincter dysfunctions are uncommon. Often, CT scan ignores the lesion because it explores the spine under it, exploring a more frequent L4, L5, or S1 radiculopathy, and neurinomas generally are at the L1–L2 level. Conversely, MRI has a 100% sensitivity. Myelography is indicated only if MRI is contraindicated.

Malignant neurinomas generally occur in von Recklinghausen’s patients and carry a poor prognosis.