

Intramedullary Spinal Cord Tumors and Pseudotumors. How to Deal With?

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Abstract

Surgery is the best treatment to offer patients with an intramedullary spinal cord tumor. In experienced hands, radical surgery can be performed with acceptable surgical risks in most ependymomas, hemangioblastomas, and cavernomas and in 40% of astrocytomas. However, some inflammatory lesions may mimic an intramedullary tumor. That is the reason why, one should pay attention to spinal cord morphology on MRI to avoid unnecessary surgery on pseudotumors that may benefit from a medical treatment.

Introduction

With MRI diagnosis, more and more intraspinal cord lesions are found with, sometimes, difficulties to assess the differential diagnosis between tumoral and non-tumoral lesions. Therefore, the practical decision on the best therapeutic attitude to recommend is not always easy. Clinical examination remains an important data that should not be forgotten by people who are faced with intramedullary lesions. Of course, there are well-known radiological aspects suggesting vascular or demyelinating myelitis. We shall not emphasize them. We would only remind a case we had several weeks ago. Our opinion was requested for a 77y-old man who had an intramedullary lesion at TH8 level that was thought to be responsible of a left pyramidal syndrome at lower limbs. History started two years before by a progressive left drop-foot. The patient complained of increasing difficulties for walking. MRI showed a small intramedullary signal on T2W images located at TH8 level on right side. There was no gadolinium enhancement of that eccentric lesion. Therefore, the diagnosis of ependymoma was excluded. But astrocytoma and inflammatory disease were kept in the differential diagnosis. However, in the absence of sensory or sphincter deficits with a lesion located on the opposite site to the clinical signs, an MRI investigation of the brain was requested allowing the discovery of a huge rolandic parasagittal meningioma on right side. That tumor was the cause of the clinical complains. So, the patient got surgery for the brain meningioma and not at TH8 spinal cord level. Of course, the question of the nature of the spinal cord lesion remains open and the patient has been proposed to get a clinical and radiological follow-up. That case illustrates the difficulty to make a correct diagnosis in non-specific MRI lesions and the danger of pitfall.

Many papers have been published on the surgical treatment of intramedullary spinal cord tumors. We have selected some. Surgery is the gold standard treatment. Radiation therapy should not be applied anymore, except in malignant tumors as a palliative treatment.

Few papers have emphasized non-neoplastic intramedullary spinal cord lesions mimicking tumors. That pathology is not well known. We have also selected several papers on that topic.

1) RADICAL EXCISION OF INTRAMEDULLARY SPINAL CORD TUMORS: SURGICAL MORBIDITY AND LONG-TERM FOLLOW-UP EVALUATION IN 164 CHILDREN AND YOUNG ADULTS.

J Neurosurg (Spine 2)(2000) 93: 183-193.

Information

The authors give their wide experience on 164 patients 21 years of age and younger in whom an intramedullary spinal cord tumor (IMSCT) was resected. They succeeded in performing what they call a gross-total resection (>95%) in 76.8% and a subtotal resection (80-95%) in 20.1%. All the patients had gliomas, the majority of low-grade (79.3%), most of astrocytic nature due to the pediatric population. There were no deaths related to surgery. When comparing the preoperative and 3-month postoperative functional grades, they observed: 60.4% stable, 15.8% improvement and 23.8% deterioration. Surgery was the only anti-tumor treatment used for the majority (73.5%) of patients, even if the tumor recurred, as long it was low grade. In patients with low-grade intramedullary lesions, long-term results of radical surgery without routine postoperative radiation treatment compare favourably with the results of modest surgery followed by radiation treatment. The major determinant of long-term patient survival was histological composition of the tumor. The 5-year progression-free survival rate was 78% for patients with low-grade gliomas and 30% for those with high-grade gliomas. Patients in whom an IMSCT was only partially resected (<80%) fared significantly worse.

Analysis

There are many important facts in that paper dealing essentially with intraspinal cord glioma in pediatric population. The first and perhaps the most important is that in experienced surgeons hands, radical surgery for IMSCTs can be performed with acceptable surgical risks. As said before by others, the authors confirm that IMSCTs should be recognized as potentially excisable lesions, both at presentation and if they recur. They give arguments discrediting adjunctive radiation therapy. It is important since one may still find in the literature papers recommending radiation therapy after complete or partial removal of intraspinal cord low-grade ependymomas or astrocytomas. That concept is no more acceptable and I fully agree with the authors. Another good message is on the timing of surgery. Functional postoperative results are closely related to preoperative situation and the presence or absence of preoperative deficits. When no plane of dissection is found, it is safer to do a subtotal removal. It is interesting to see that in their experience, subtotal tumor removal may be sufficient for long-term progression free survival.

2) INTRAMEDULLARY EPENDYMOMAS: CLINICAL PRESENTATION, SURGICAL TREATMENT STRATEGIES AND PROGNOSIS.

J Neuro-Oncol (2000) 47: 211-218.

Information

This is a general overview by an experienced team on intramedullary ependymomas, which comprise the majority of intramedullary glial neoplasms in the adult. There is no typical clinical presentation but dysesthesias are the earliest symptoms to present in upwards of 70% of patients. These tumors are benign slow-growing lesions, which are optimally treated with gross total surgical resection without adjuvant therapy. This objective can be attained safely in a majority of patients. Post-operative functional outcome is related to pre-operative functional status. Early diagnosis, prior to symptomatic progression, is critical to the successful treatment of these tumors. Adjuvant therapy is indicated for the rare malignant or disseminated tumors or following sub-total resection.

Analysis

Surgical technique is very well detailed in that paper. In adult population, standard laminectomy is performed. Laminoplasty, which is mandatory in children, is not required in the adult population. The authors rightly insist on the importance of strict hemostasis prior to dural opening to prevent ongoing blood contamination. Surgery of intramedullary tumors needs a very clean operative field. They open the arachnoid separately, as we do. They recommend posterior midline opening extending over the entire rostrocaudal extent of the tumor. That helps finding the polar cysts, very frequent in ependymomas. Most of the times, ependymomas are sharply demarcated from the surrounding spinal cord. In huge tumors, debulking is recommended with ultrasonic aspirator or laser. Once significant tumoral internal decompression has been performed, dissection of the lateral and ventral margins may be accomplished, taking care hurting anterior spinal artery that gives the feeding arteries, which should be identified, cauterised and divided. If no plane is apparent between tumor and surrounding spinal cord, biopsy is obtained to confirm the histological diagnosis of an infiltrative tumor. That is the classical way to remove an intraspinal cord ependymoma. The authors don't perform a closure of the myelotomy. We do it, so as the closure of the arachnoid whenever possible. The authors also emphasize the importance of preservation rather than expected restoration of neurologic function in surgery for intramedullary tumors.

3) SPINAL CORD ASTROCYTOMAS: PRESENTATION, MANAGEMENT AND OUTCOME.

J Neuro-Oncol (2000) 47: 219-224.

Information

This is a general overview by an experienced team on intramedullary astrocytomas, which is slightly less frequent than ependymomas in adult series. There is no typical clinical presentation. But local or radicular pain is the earliest and most frequent presenting complaint. Astrocytomas have less well-defined margins on MRI and are more likely to be eccentrically located within the spinal cord unlike ependymomas, which are always central in location. Ideal candidates for surgery are patients who are still ambulatory. The infiltrating nature of some spinal cord astrocytomas may make total removal impossible without unacceptable neurological deficits. In such instances, subtotal resection may still be worthwhile to reduce tumor mass in preparation for adjunctive therapy. Intraoperative somatosensory and motor evoked potentials are used. They provide information to guide the surgeon in the resection but their intraoperative utility is limited by many factors. So, their use is still a matter of debate. Technique of surgery is similar to what is described for ependymomas in the above paper. The outcome is worse than that of ependymomas, except for pilocytic astrocytomas, more frequent in paediatric population, and which can be totally removed. High-grade astrocytomas have a bad prognosis with an average survival of 6 months in adults and 13 months in children.

Analysis

We agree that MRI cannot be used confidently to distinguish between astrocytomas and ependymomas. MRI cannot predict the presence or the absence of a plane of separation between tumor and spinal cord. That is a sufficient reason to recommend surgery, especially in patients with few neurological deficits. Finding a plane at surgery will make the tumor removable. Otherwise, it is safer to limit to biopsy and to decompressive surgery. Malignant astrocytomas have a grim prognosis, as do intracranial anaplastic gliomas, even with adjunctive therapy.

4) NON-NEOPLASTIC INTRAMEDULLARY PATHOLOGY. DIAGNOSTIC DILEMMA: TO BX OR NOT TO BX?

J Neuro-Oncol (2000) 47: 283-292.

Information

That is a very good synthesis on a difficult diagnosis to which the consulting neurosurgeon, pressured to operate on a rapidly deteriorating patient, may be faced with the uncomfortable decision of whether to operate on potentially nonsurgical pathology or recommend medical management. Multiple sclerosis can mimic an active neoplasm leading to mistaken biopsies. Granulomatous

disease in the spinal cord has been reported for sarcoidosis, tuberculosis, brucellosis and histoplasmosis. These lesions often have systemic manifestations but sometimes, they may be the first manifestation of the illness. A biopsy may be the only way to get a diagnosis, which is important since in sarcoidosis, corticosteroids therapy dramatically improves the patient, cleaning the MRI. Isolated pseudotumors have also been described. Infections may also happen, especially in HIV patients where they are not infrequent. Intramedullary cysticercoids and toxoplasmosis have been occasionally mistaken for intramedullary tumors. Vascular lesions have also been described: amyloidosis, isolated intramedullary vascular lesions of ischemic origin, arterial infarcts. Finally, other lesions like subacute necrotizing myelopathy or radiation myelopathy may also be encountered.

Analysis

When something about the MRI (spinal cord not significantly enlarged, minimal enhancement) or clinical course raises suspicion of a non-neoplastic lesion (symptoms have progressed rapidly), we recommend a cautious attitude. If the illness is slowly progressing, one may suggest performing a new MRI 3 months later. When there is a suspicion of multiple sclerosis, a brain MRI can bring the correct diagnosis, avoiding an unnecessary biopsy. CSF diagnosis may be of great help. In suspected granulomatous diseases, systemic manifestations should be searched since the diagnosis may be assessed on those systemic manifestations or serum analyses. Primary sarcoidosis may happen. Several such a cases have been reported. In the absence of diagnosis, a cautious biopsy of the intraspinal cord lesion is to be done but not resection as deterioration following nodule removal has been reported. Cautious MRI analysis may reveal a ring enhancement, which is not often seen with intramedullary tumors. The later demonstrate homogeneous or heterogeneous patchy enhancement, exceptionally a ring enhancement that is more suggestive of cord infarction, radiation myelopathy or subacute necrotizing myelopathy. In certain circumstances, non-neoplastic lesions will require open biopsy or surgical treatment as a part of the optimal management: bacterial, fungal, tuberculous or parasitic lesions may benefit of surgery in addition to medical treatment.

5) MEDICALLY TREATED INTRASPINAL "BRUCELLA" GRANULOMA.

Surg Neurol (1999) 52: 570-576.

Information

This is a case report of a 40-year-old female presenting with a two months history of progressive weakness of the right leg and numbness of the left leg with urinary and fecal incontinence. Four months before, she had been diagnosed with systemic brucellosis with a period of radiculomeningoencephalitis successfully treated with medical treatment. MRI revealed a 10 x 30 mm intramedullary heterogeneous mass at T5 level with considerable enhancement and surrounding edema. Due to the past medical history, a Brucella granuloma was suspected and CSF analysis performed with finding of lymphocytic pleocytosis and elevated levels of albumin, immunoglobulins and antibody titers for Brucella. The

medications were modified to rifampicin 1200mg daily, doxycycline 400mg, TMP/SMZ (trimethoprim/sulfamethazole) 480/2400 mg daily, and methylprednisolone 100 mg in decremental doses for (for 6 weeks). No surgery was done. After 2 months, the patient was almost symptom-free and her medication doses were decreased. Follow-up MRI at 5 months showed a dramatically decrease of the mass lesion. The treatment was discontinued after 2 years.

Analysis

Brucella infections continue to cause serious problems in developing countries.

That paper is a good warning to neurosurgeons to consider a non-neoplastic lesion before recommending surgical resection when some data are not typical for a tumor. Here, the medical history was short (2 months), the MRI suspicious (eccentric heterogeneous mass) although the authors had the knowledge of a systemic brucellosis 4 months before. Infectious disease may mimic neoplastic mass on MRI. When a medical history keeps the attention, there is few risk of misdiagnosis but when the past history is unknown, it is not easy to take the good decision: medical treatment as an initial treatment rather than surgery.

6) INTRASPINAL SARCOIDOSIS: DIAGNOSIS AND MANAGEMENT.

Surg Neurol (1997) 48: 514-521.

Information

The authors report 3 patients with intraspinal sarcoidosis and absent systemic manifestations of the disease. Two of the three patients had a preoperative diagnosis of a cervicomedullary spinal cord tumor The third one had a cauda equina location. Since MRI with gadolinium did not suggest an inflammatory process, all three patients underwent laminectomy and biopsy. Pathologic examination demonstrated sarcoidosis in all three patients. It is recommended not to attempt complete resection if this granulomatous process is suspected. Treatment is prolonged corticosteroids.

Analysis

One should be aware that there are no distinct MRI signal characteristics for intraspinal cord sarcoidosis. It may mimic an infiltrative glioma. That is one of the reasons why in intramedullary lesions, no adjunctive treatment (especially radiation therapy) should be performed without histologic analysis, even in poorly delineated lesions. In my department, we have experienced two cases of primary intraspinal cord sarcoidosis and we did what the authors have recommended: limited laminectomy, biopsy rather than attempt to resect, and intraoperative frozen sections that in both cases pointed to an inflammatory process. Treatment is corticosteroids, sometimes over several months.

7) NONNEOPLASTIC INTRAMEDULLARY SPINAL CORD LESIONS MIMICKING TUMORS.

Neurosurgery (1998) 43: 788-795.

Information

Among a series of 212 patients undergoing surgery for intramedullary spinal cord tumors, the authors have reviewed a group of 9 patients with atypical, nonneoplastic intramedullary lesions that all underwent surgery. All patients were preoperatively evaluated with MRI. The extent of enhancement with gadolinium varied from homogeneous enhancement to no enhancement. This illustrates well the difficulty to assess the diagnosis of nonneoplastic intramedullary spinal cord lesions. All lesions showed marked T2 changes. There was a lack of significant spinal cord expansion associated with the lesions in all cases. The histology of the surgical specimens showed demyelinating lesions in four patients, sarcoidosis in two patients, amyloid angiopathy in two patients, and a mass of nonneoplastic inflammatory cells of unknown origin in one patient. Although it was difficult to antecedently distinguish these lesions from neoplastic spinal cord tumors by case history and physical examination, the most consistent clue was absent or minimal spinal cord expansion on the preoperative MRI.

Analysis

The surgical management of intramedullary spinal cord tumors is based on oncological principles. The goal is complete resection or significant cytoreduction of the tumor. But in nonneoplastic diseases, these oncological principles do not apply. We need a biopsy only to offer the patient the best medical treatment. Among the preoperative signs that could warn us, the only reliable one is MRI feature. Like the authors, we have observed that spinal cord expansion is always present on MRI of patients with spinal cord tumors. On the other hand, MRI images of patients with nonneoplastic lesions showed no lesional spinal cord expansion, except for mild expansion associated with edema. That is the most important message from that paper.

Synthesis

More and more papers are published on intramedullary spinal cord tumors and lesions mimicking that pathology. In experienced neurosurgeons hands, radical surgery for these tumors can be performed with acceptable surgical risks. It is also clear that the surgical outcome is directly related to the patient's preoperative conditions. The greater are preoperative neurological deficits, the greater are surgical risks and postoperative sequelae.

However, one should avoid operating unnecessary lesions that may be better medically treated. Pseudotumors, inflammatory lesions, sarcoidosis and others may mimic on MRI intramedullary tumors. But if one pay attention to spinal cord morphology, it is also clear that in the absence of spinal cord widening, the first diagnosis to think for is not a tumor but a nonneoplastic lesion.

Papers reviewed

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