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Challenges in diagnosis and management of adult spinal cord gliomas

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INFO ARTICLE

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ABSTRACT

Intramedullary spinal cord gliomas have very low incidence rates. They are associated with difficulties in diagnosis and treatment, and cause significant morbidity. Their clinical presentation and their appearance at magnetic resonance imaging are not specific. They can mimic inflammatory, infectious, vascular disorders or other neoplastic lesions. Primary treatment is surgery. Surgical resection can often be total for ependymomas, but difficult for infiltrating astrocytomas. Radiotherapy is indicated for malignant tumors, but remains controversial in some indications. Chemotherapy is reserved for recurrence, but small retrospective series are available. Genetic studies have revealed genetic alterations which could have a potential impact on treatment in the near future.

1. Introduction

Primary spinal cord tumors are rare, about 4–8% of all tumors of central nervous system (CNS) [1,2], that is, 10 to 15 times less common than their cranial counterparts. They are conventionally divided according to their anatomic location into three categories: extradural, intradural extramedullary and intramedullary. Intramedullary spinal cord tumors (IMSCTs) are the rarest type, and represent 20% of all intraspinal tumors. Their incidence has been reported as 0.22 per 100,000 [3]. Spinal cord gliomas in adults include ependymomas and astrocytomas, representing 90% of IMCSTs. Ependymomas account for 60%, and represent 30% of CNS ependymomas [4]. Astrocytomas account for 30%, but represent only 3% of CNS astrocytomas [5], with the opposite for children under 10 years of age, where astrocytomas are more common than ependymomas. The remaining IMCSTs include hemangioblastomas (3 to 8%), metastases (2%), primary CNS lymphomas and miscellaneous tumors. Despite their rarity, knowledge of these tumors is essential. Without early accurate diagnosis, and thus treatment, they lead to neurologic deterioration, poor quality of life and death. Difficulties occur at every stage of diagnosis and management.

2. Diagnostic difficulties for intramedullary gliomas

2.1. *Clinical history and physical examination are not specific*

Clinical presentation depends on location and malignancy degree, but remains fundamental, alerting to the spinal cord

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location, and prompting early magnetic resonance imaging (MRI). Clinical elements are essential in differential diagnosis, especially when MRI findings cast doubt on other myelitis. Pain is the most common presenting symptom reported, regardless of the location (50–70%) and may manifest as back pain, diffuse pain, or radicular pain; it is more suggestive when worsening at night. The other presenting symptoms are sensory (50–60%) and motor (50–60%) deficits, syringomyelitic syndrome, and autonomic and sphincter impairment [6,7]. No symptoms are pathognomonic. Some elements however point to a tumor: insidious symptom onset, long duration before diagnosis (23 months for astrocytomas versus 27 for ependymomas), average age of onset (33.7 years for astrocytomas, 39.8 years for ependymomas) and progressive evolution without relapse.

2.2. *Magnetic resonance imaging*

MRI is the diagnostic modality of choice for intramedullary gliomas. Whole spine MRI is required, with at least T1-weighted and T2 weighted imaging, with and without contrast. Brain MRI may be essential in some cases. The typical MRI appearance of intramedullary glioma shows a lesion extended over several cord levels, with enlargement of the spinal cord, and variable gadolinium enhancement. A hemorrhagic component, some tumoral or non-tumoral cysts, an area of necrosis, or an infiltrating pattern are frequent, depending on the kind of tumor. But, despite the high quality of available imaging modalities, the diagnosis of spinal cord lesions remains difficult in some cases.

Distinction between astrocytoma and ependymoma is often difficult, though it is important for surgical planning. There are some helpful diagnostic tools but none are specific. Ependymomas are typically centrally located, with symmetrical cord expansion, and most are cervical (in 44%). By contrast, astrocytomas are eccentrically located, and more often thoracic. Associated hemorrhage and cysts are less common in astrocytomas than in ependymomas. Ependymomas enhance homogeneously and have clearly defined margins, unlike astrocytomas. Diffusion tensor imaging (DTI) and diffusion tensor tractography (DTT) may be useful for surgical planning to attempt to differentiate infiltrating spinal cord tumors such as astrocytomas and non-infiltrating tumors such as ependymomas, and, as reported in recent review, to try to assess the extent of the tumor [8].

Evaluation of tumor grade is also difficult on the MRI. Enhancement in brain tumors is due to disruption of the blood-brain barrier, and is synonymous to high-grade, (except in pilocytic astrocytoma (Fig. 1). That is not the case for the spinal cord where enhancement is not a trustworthy criterion of malignancy (Fig. 2). Some spinal cord tumors enhance, while the majority are low-grade: 70% of astrocytoma are low-grade however enhancement is found in 70% of them. Similarly, 90% of ependymoma are low-grade (Fig.Fig. 3), but 90% are enhanced on MRI (Figs. 3 and 4). Clinical elements, such as patient age (30–50 years for ependymoma, 20–30 for astrocytoma), time before diagnosis, longer in ependymomas (8–36 months) than in astrocytoma (24 months), may be helpful. Functional status and quality of life at diagnosis are of interest. They are more preserved in case of ependymoma than astrocytoma [7].

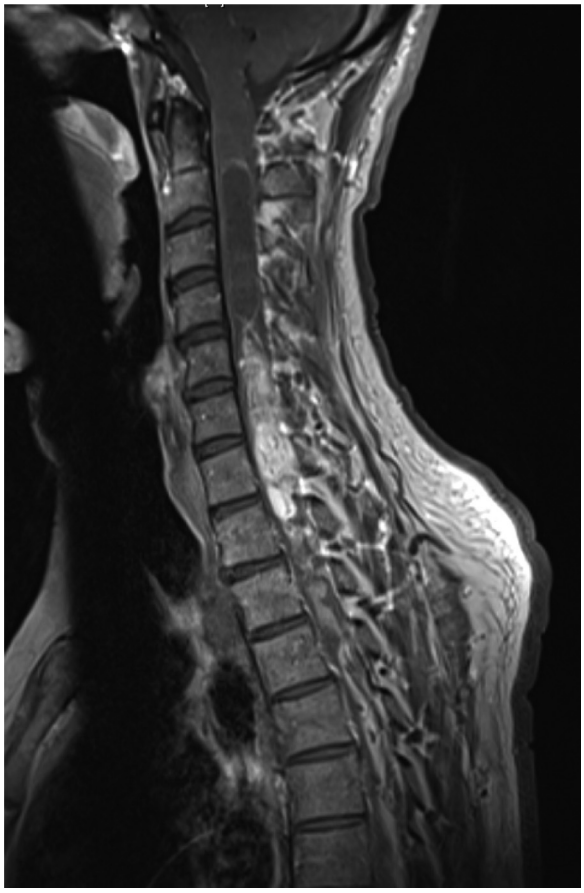

Fig. 1 – Pilocytic astrocytoma in a 30-year-old man who underwent MRI for gait disturbance, for 1 year, and recent bladder dysfunction. Sagittal T1-weighted with contrast MRI revealed an enlargement of the cervical cord on several vertebral segments, with an intense enhancement of a solid component and cyst walls.

2.3. *Histopathological diagnosis*

The diagnosis is based on the WHO 2016 classification of CNS tumors, which fundamentally changes the classification, especially of diffuse gliomas [9]. The previous grading is maintained. Ependymomas are divided in three grades: grade 1 (subependymoma and myxopapillary), grade 2 (low-grade), and grade 3 (anaplastic). Astrocytomas remain divided in four grades: pilocytic (grade 1), more common in children, low-grade (grade 2), anaplastic (grade 3) and glioblastoma (grade 4). However, this new classification integrates histopathological and molecular features. Furthermore, a radiological approach by a neuropathologist, especially in case of biopsy, is needed to achieve accurate diagnosis. This classification seems to improve interobserver reproducibility and evaluation of prognosis compared with a purely histological classification.

Diffuse gliomas (previous grades 2, 3 and 4) are classified according two molecular alterations: presence (mutant), or absence (wildtype) of *IDH1* and *IDH2* mutation, and presence or absence of 1p/19q codeletion. The isocitrate dehydrogenase (*IDH*) gene mutation is frequently found in WHO grade II and III astrocytomas, oligodendrogliomas and secondary glioblasto-

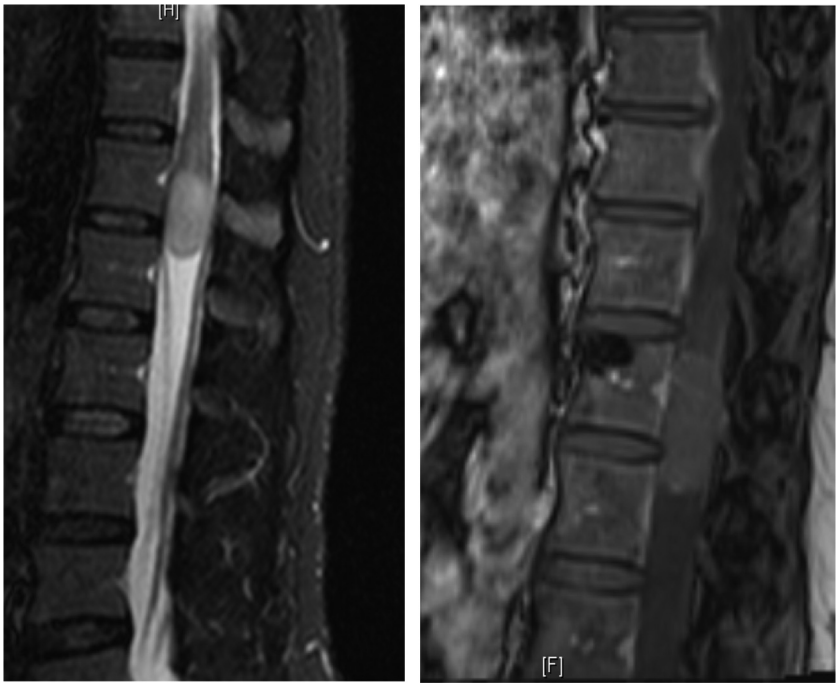

Fig. 2 – Anaplastic astrocytoma in a 50-year-old woman who presented back pain for one month. MRI showed, on sagittal T2-weighted sequences, a hyperintense lesion in the conus, associated with cord expansion. On the T1-weighted sequence post-contrast, the tumor was mildly enhanced.

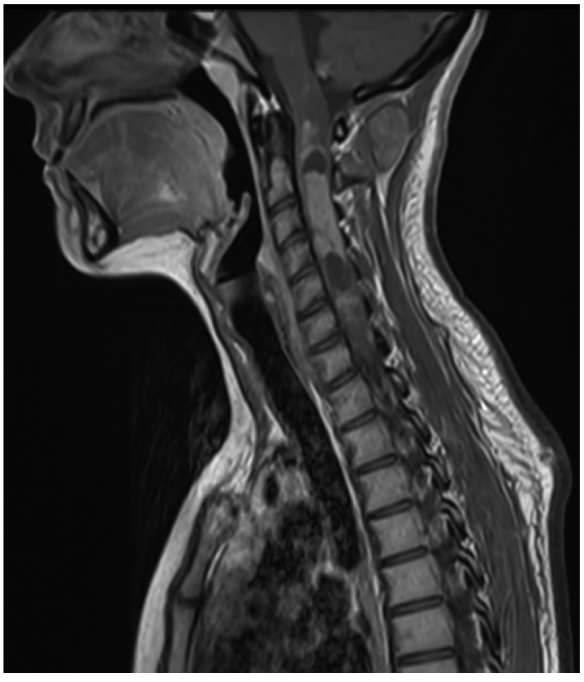

Fig. 3 – Ependymoma grade 2, in a 24-year-old woman with a diagnosis of neurofibromatosis type 2. She presented progressive lower-limb, then, upper-limb weakness. Sagittal T1-weighted MRI post-contrast shows a homogeneously enhancing intramedullary mass, with caudal and rostral cysts.

mas of the brain. *IDH* mutations have a more favorable prognosis compared to *IDH*-wildtype, independently of the glioma grading. 1p/19q codeletion is a strong good prognostic factor, detected in oligodendrogliomas, and is associated with response to chemotherapy. According to presence or absence of *IDH* and 1p/19q deletion, three separate prognosis groups are defined: good, intermediate and poor prognosis.

Other molecular findings are included in the diagnosis of these tumors, such as abnormalities of oncogene *BRAF*, and histone mutations.

Abnormalities of oncogene *BRAF* (belonging to the mitogen-activating protein kinase pathway) include either fusion (*KIAA 1549-BRAF*), or mutation (*BRAF 600*), and are common, especially in pilocytic astrocytomas (grade 1).

When histopathological features are lacking to discriminate astrocytomas, molecular findings can provide the diagnosis. *IDH* mutation signs diffuse astrocytoma, *BRAF* trends to pilocytic astrocytomas [10].

Histone mutations are harbored in a distinct subgroup of gliomas, which is a new entity: diffuse midline glioma *H3 K27M* mutant. This mutation has been detected in malignant astrocytomas with midline location (thalamus, brainstem, spinal cord), and occurs in children and young adults. Its identification means poor prognosis and grade 4. Meyronet et al. [11], reported that in nearly half of their 21 patients, histopathology was suggestive of a diffuse low-grade tumor, and three of them were reclassified as WHO grade 4 after molecular analysis. This mutation, found in glioma of midline location, impacts diagnosis and prognosis.

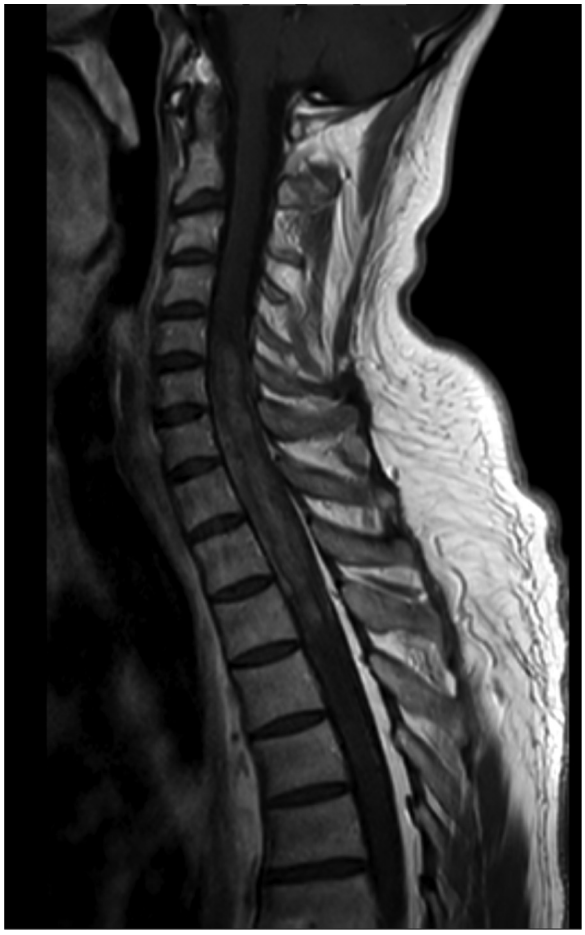

Fig. 4 – Anaplastic Ependymoma in a 44-year-old man with a history of back pain and progressive sensory loss at one year. Sagittal T1-weighted MRI shows mild cervicothoracic contrast uptake on several levels, with swelling of the spinal cord.

But it may be difficult to determine whether prognosis, given by this classification, is the same in brain tumors as in spinal cord tumors, except for diffuse midline glioma. Intramedullary gliomas are different regarding molecular markers. Thus, fusion of *BRAF* is reported more often in brainstem and spinal cord, and mutation, reported more often in brain [10]; likewise, *IDH* mutation seems rarer in spinal cord tumors than in brain tumors. In a recent survey of 83 patients, spinal astrocytomas were mainly *IDH* wildtype [12].

The difficulty of this new classification seems to require several samples, often difficult in the spinal cord, and rapid access to a molecular biology platform.

3. Differential diagnosis of intramedullary gliomas

3.1. *Other neoplastic tumors*

Hemangioblastomas are benign tumors that account for 3 to 8% of spinal cord tumors and are reported in 75% of cases in an intramedullary location [13]. Most are sporadic, but some of them are associated with von Hippel Lindau disease (VHL). VHL is an autosomal dominant disease caused by a genetic mutation of the *VHL* gene on chromosome 3 and may provide multiple benign and malignant tumors. Usually, such tumors do not present a problem for differential diagnosis with IMSCT, depending on their pattern: they are associated with peri-tumoral cysts, are located at the pial surface of the cord, and not in the center like ependymomas. They show homogenous enhancement, and, in some cases, a cyst with an enhancing nodule as in the cerebellar location (Fig. 5). When VHL is suspected, genetic testing must be performed and MRI of the entire craniospinal axis is required.

Primary lymphoma of the CNS represents less than 1% of CNS tumors. T1-weighted MRI shows homogeneous contrast enhancement and spinal cord enlargement (Fig. 6). Diagnosis is essential because high-dose methotrexate therapy has been shown to be an effective treatment in elderly patients [14].

Gangliogliomas account for 1% of all CNS tumors. Most of them are intracranial. MRI findings are not specific, showing a mass lesion, often larger than IMSCT, with contrast enhancement. Diagnosis is histopathological.

Intramedullary spinal cord metastases, from lung cancer, breast cancer, systemic lymphomas and melanoma, represent about 2% of intramedullary neoplastic lesions, and affect 0.4% of all patients with cancer. On MRI there is a peritumoral edema formation and contrast enhancement. However, the clinical presentation may be helpful: progression of neurologic symptoms is rapid with high incidence of complete deficits [15].

Intramedullary spinal cord metastasis of high-grade brain gliomas, also called ‘‘drop metastasis’’ are rare events, less than 2% [16]. They are more often leptomeningeal than intramedullary, and occur throughout the evolution of high-grade glioma or medulloblastoma.

Primary CNS melanomas are extremely rare, and account for about 1% of all melanomas. The progression of symptoms is more rapid than IMSCT. Typically, they are characterized by hyperintensity on T1-weighted sequences, and hypointensity on T2- weighted sequences, due to the presence of melanin, and by homogeneous enhancement by contrast. Additional investigations are required to exclude metastatic melanoma [17].

3.2. *Non-neoplastic lesions*

The appearance at MRI can mimic inflammatory disorders, infectious, systemic or vascular diseases. A glioma in early stage may not be enhanced, with poor enlargement, and short length. On the other hand, a ‘‘big spinal cord’’ with heterogeneous contrast uptake may be an inflammatory myelitis. Many etiologies are possible, but some of them are very rare, and need systematic investigations according to the background.

Multiple sclerosis (MS), the main cause of myelitis in young adults, may be problematic at the beginning of the disease, without a notion of relapse, and no evidence of dissemination in time and space. About 30% of clinically isolated syndromes are only located in the spinal cord with an acute or subacute onset [18]. On MRI, lesions have an increased signal on T2-

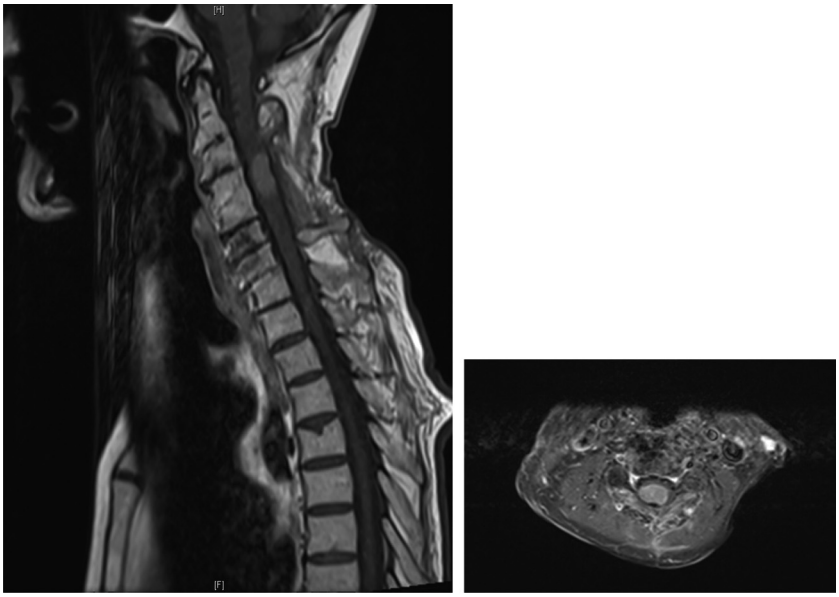

Fig. 5 – Primary CNS lymphoma, in an 80-year-old man with radicular pain and motor deficit. Sagittal T1-weighted and axial sequences identified a mass with intense and homogeneous enhancement. The patient underwent chemotherapy and was alive two years later.

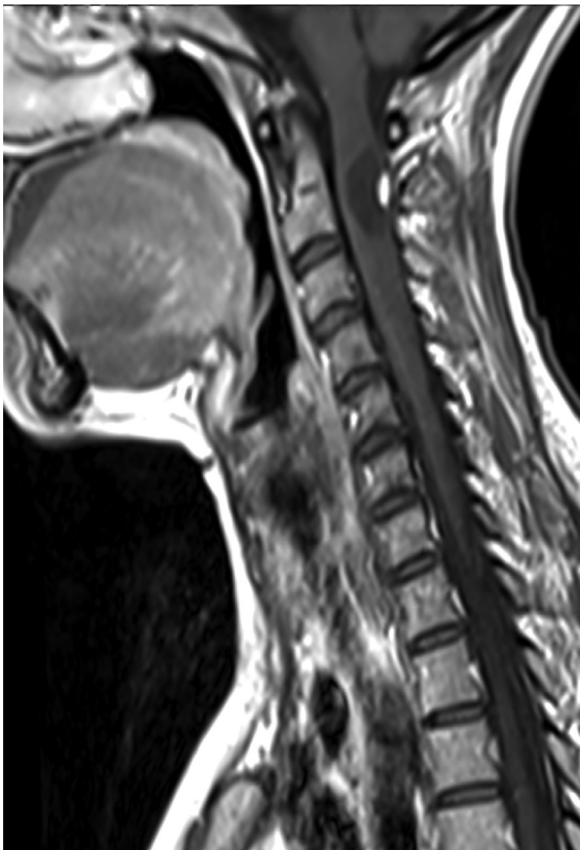

Fig. 6 – Hemangioblastoma. Spinal cord magnetic resonance imaging (MRI), T1-weighted post contrast sagittal sequences, shows an enhancing nodule inside a cyst, with an enlargement of the cervical spinal cord.

weighted images, sometimes focal swelling, with enhancement on active MS lesions, but its length (less than two cord levels) and its localization, at the dorsal portion of spinal cord, are suggestive. Investigations include serological exams and cerebrospinal fluid (CSF) exploration. An intrathecal synthesis of immunoglobulins (positive oligoclonal bands) may be identified and confirm diagnosis. If other investigations such as immunologic tests are not contributive, management consists of corticosteroids at high dosages, and a follow-up with serial MRI. Arguments for MS are decreased swelling and disappearance of contrast enhancement on interval MRIs.

Neuromyelitis optic (NMO) is a severe autoimmune inflammatory disease defined by optic nerve involvement and spinal cord lesions. The diagnosis is made by detection of antibodies against aquaporine 4 (AQP4)-immunoglobulin G which is a sensitive and specific biomarker, 76.7% and 99.8% respectively [19]. This disease occurs between 32 and 45 years of age [20]. The onset is acute or subacute. The MRI aspect may be tumoral, showing longitudinal involvement on the spinal cord (longitudinally extensive transverse myelitis) at three or more vertebral segments. Usually cervical, thoracic or cervicothoracic spinal segments are involved. In acute phases, it is possible to find cord swelling and irregular enhancement, (hypointense patchy areas and ring enhancing lesions) on T1-weighted images. If spinal cord lesions occur alone at first presentation, without optic lesions, and if NMO is seronegative, difficulties are significant. The severity of prognosis imposes emergency treatment, high-dose corticosteroids and additional immunosuppressant treatment. Follow-up is essential. In the event of inadequate response, if lesions do not improve or progress one month after symptom onset, the question of an early biopsy is crucial. It must be considered to

prevent delays and deteriorating neurologic status in the management of misdiagnosed cases.

Sarcoidosis (SA) is a granulomatous, multisystem disease of unknown etiology which occurs in adults about 30–40-years old. Neurosarcoidosis (NS) can occur with other sarcoidosis forms, but, in 1% of cases, leads to diagnostic problems when it involves only the CNS. NS can affect the spinal cord in 4-28% of cases [21] and can mimic a tumoral presentation with a long duration of symptoms. Lesions are intramedullary with fusiform enlargement of the spinal cord, and involve the leptomeninges. Location is often at the thoracic level, affecting three or more spinal segments. Some imaging findings are useful, such as a thickening of the leptomeninges and basilar leptomeningeal involvement. Sets of criteria for SA have been proposed [22]. If diagnosis cannot be established in extra-neural locations (lungs, conjunctiva etc.), a biopsy of the cerebrospinal meninges can be considered.

Infectious myelitis is very rare. The disease presents with an acute or subacute onset, extensive lesions and has a severe prognosis. Infectious myelitis may occur in immunocompromised individuals. Viruses involved include herpes virus, Epstein-Barr virus, cytomegalovirus, human immunodeficiency virus, hepatitis A, B and C viruses etc. [18]. Bacterial etiologies are possible (*Staphylococcus*, tuberculosis etc.), as well as parasitic (bilharziasis...) or fungal (*Cryptococcus*...).

Spinal cord abscesses are extremely rare, with imaging presentation similar to a brain abscess: ring-shaped enhancing lesion that expands the cord and surrounding edema. Rapid progression implies an early diagnosis.

3.2.1. *Spinal vascular dural malformations*

Their incidence is reported to be 3–16% of spine space-occupying lesions. They are not intramedullary, but may include venous congestion, leading to cord edema, producing progressive myelopathy. Spinal dural arteriovenous malformations (SDAVF) are the most frequent. They occur mostly in men aged 40–60-years old, in the lower thoracic level, and consist of a dural shunt between a radiculomeningeal artery and a radicular vein [23,24]. Characteristic features are hyperintensity on T2-weighted sequences, with cord expansion and serpentine low-signal intensity flow voids of tortuous vessels. However, they can be obscured by a CSF pulsation artefact. If doubt remains, the definite diagnosis is based on catheter angiography.

3.2.2. *Spinal cord cavernous malformation (SCM)*

SCMs are angiographically-occult slow-flow vascular lesions. Their incidence is 5% of intra-medullary lesions in adults. The clinical presentation may be acute or chronic. On T2-weighted MRI, SCMs demonstrate a peripheral rim of low signal intensity due to hemosiderin. Differentiation between SCM and hemorrhagic glioma may be absence of cord edema and minimal enhancement.

3.2.3. *Syringomyelia*

Syringomyelia is a chronic disorder involving the spinal cord, characterized by the presence of a longitudinally cystic intramedullary cavity, located around the central canal of the spinal cord, most commonly at the cervicothoracic junction. It may be congenital, such as Chiari malformation, or secondary to trauma or tumor [25]. The cysts do not enhance on post-contrast studies. If any doubt, clinical follow-up and serial MRI are sometimes required to look for an enlargement of the spinal cord or contrast uptake to ensure there is no tumor.

4. Management difficulties for intramedullary gliomas

The risk of secondary surgery-related neurologic deterioration implies a multidisciplinary reflection before making any decision about an appropriate time for surgery. If the patient is mildly symptomatic, serial imaging follow-up is usually recommended to evaluate disease evolution (MRI every 2 or 3 months). Prudence is nevertheless the rule since preoperative neurologic function is one of the major prognostic factors. No improvement over the preoperative status can be expected postoperatively. Decisions concerning the type of surgical procedure (biopsy with duraplasty, resection) depend on the localization, the degree of infiltration, and the presence of a surgical plane. An optimized MRI is needed. Surgery should be performed under intraoperative electrophysiological monitoring.

In the postoperative period, MRI evaluation of the spinal cord glioma will be one of the difficulties. Evaluation methods are similar to those used for brain gliomas, but measurements are difficult owing to the small size of these tumors.

4.1. *Ependymoma*

4.1.1. *Surgery*

The standard of care for grade 2 sporadic symptomatic ependymoma, which occurs in 70% of cases, is surgery. A surgical plane of dissection can often be identified, allowing gross total resection (GTR). It is the most effective treatment, and well codified [26]. No further treatments are needed. Extent of resection, confirmed by postoperative MRI, is a strong prognostic of overall survival.

4.1.2. *Radiotherapy*

In 2017, the European Association of Neuro-Oncology (EANO) guidelines for the diagnosis and treatment of ependymal tumors [27], recommended radiotherapy (focal or craniospinal), in case of anaplastic ependymoma regardless of the extent of resection.

Management is more debated for low-grade ependymomas, when GTR is not achieved. Two strategies are available. The first one is closer surveillance and new surgery at clinical and imaging recurrence, if the patient remains mildly symptomatic. The second is adjuvant radiotherapy following partial resection, or differed when recurrence. A review of the literature [28], in 2013, has been performed for 348 ependymomas. The conclusion was that radiotherapy prolonged progression-free survival (PFS) in patients receiving STR (subtotal tumor resection): median PFS 48 months in patients treated with STR alone and 96 months for these treated by STR and followed by radiotherapy. But all studies have been retrospective and management is currently based on evaluation of tumor localization (possibilities of new surgery), age, and functional status.

4.1.3. *Chemotherapy and target therapy*

Chemotherapy is an option at recurrence of grade 2 ependymoma after surgery and radiotherapy, and for grade 3 ependymoma. Data supporting chemotherapy for spinal localizations are very limited: Chamberlain in 2002 [29] reported a study of 10 patients with recurrent spinal ependymoma given topoisomerase-2. He related some efficacity of chemotherapy in 20% of patients with a partial response. Chemotherapy has been studied mostly in recurrent intracranial ependymoma, and by extension is used for spinal ependymoma. Chemotherapeutic agents include platinoid salts and temozolomide, an oral alkylating agent. Lapatinib, an epidermal growth factor antagonist, has also been studied. These treatments seem to have a very modest effect. More studies with larger numbers of patients must be conducted with various chemotherapeutic agents or target therapies before concluding.

4.1.4. *Ependymomas and neurofibromatosis 2 (NF2)*

Another problem is the management of ependymomas associated with NF2. NF2 affects one in 30,000 individuals worldwide, and *NF2* mutation is the most common mutation found in spinal cord ependymomas. The majority of NF2-associated spinal ependymomas are asymptomatic, only 20% of them are symptomatic. There is no well-established management protocol, but follow-up is important.

Surgical resection may be curative, but the main difficulty is timing the resection. A detailed neurologic surveillance is warranted to assess clinical evolution of the ependymoma. For some authors, surgery can be indicated only when patients become symptomatic [30], but for others, should be undertaken as soon as radiological progression is demonstrated. Sometimes, the difficulty is to demonstrate the relationship between the symptoms and the tumor.

A retrospective study of NF2 patients who were treated with bevacizumab for schwannomas is reported by Morris et al. [31]. Bevacizumab is an antiangiogenic agent which targets vascular endothelial growth factor. A clinical and radiological improvement was noted in 58% of patients with spinal cord ependymoma.

4.2. *Astrocytoma*

4.2.1. *Surgery*

Surgery is an effective treatment only for grade 1 pilocytic astrocytoma when resection can be complete. Management of diffuse astrocytoma is more difficult. Astrocytomas are low-grade in 75% of cases, but they infiltrate the spinal cord without characterized boundaries. Surgery is almost always partial. The challenge is to attempt maximal resection without permanent neurological deficit. The optimal extent of surgery remains controversial. Some publications support maximal resection [32]. A number of publications have not found any benefit. Treatment consists of maximal and safe resection, but often a biopsy alone is performed with duraplasty, to allow tumor growth or swelling. But partial resection increases recurrence risk of astrocytoma and needs adjuvant treatments.

4.2.2. *Radiotherapy*

Radiotherapy is the next course of treatment for patients with high-grade astrocytomas, biopsied-only tumors, or progressive disease. However, time of radiotherapy for low-grade astrocytoma remains controversial, even after partial resection has been done. Minehan et al. [33], reported a retrospective study of 136 patients. Radiotherapy improved survival for grade 2, 3 and 4 astrocytomas, but not pilocytic astrocytoma. A recent meta-analysis [34], on 3022 cases of ependymoma and astrocytoma, revealed that radiotherapy reduces time of recurrence, but improves overall survival only in high-grade tumors. Some authors recommend closer observation after subtotal resection, and differ radiotherapy until recurrence, considering the young age of patients. Indeed, radiation necrosis is one form of toxicity. It may be difficult to differentiate tumor recurrence from radiation necrosis, especially when swelling or contrast uptake are located in the radiation field. MRI is not specific. Reports using positron emission tomography (PET) to image primary intramedullary tumors are limited. PET, with FDG (fluorodeoxyglucose), and MET (methionine), seems to help in the differentiation of these diagnoses [35]. The development of new techniques could reduce complications associated with traditional radiotherapy, and could broaden indications.

4.2.3. *Chemotherapy and target therapy*

Chemotherapy is reserved for recurrence, after surgery and radiotherapy. By extrapolation with brain gliomas, the same agents are delivered with the same scheme. Only small series have been published about spinal cord gliomas, so the relevance of treatment is unproven.

In low-grade astrocytoma, Chamberlain [36] reported a retrospective study of 22 patients with recurrent low-grade gliomas, previously treated by surgery and radiotherapy. This study suggested a modest, but effective, efficacity with median survival of 23 months, without major adverse reactions.

In high-grade astrocytomas, radiotherapy combined with concomitant chemotherapy with temozolomide, has been often delivered for high-grade gliomas, especially spinal cord glioblastomas, but toxicity of spinal cord radiation has not been evaluated with concomitant chemotherapy.

This grade 4 tumor is extremely rare, accounting for 1.5% of all spinal cord tumors, but occurs in earlier decades of life, with a poor prognosis (survival 10–12 months after diagnosis). Some studies reported improvement of prognosis with multimodality treatment (combined radiotherapy and chemotherapy with bevacizumab at recurrence etc.). The overall survival switched to 17.9 and 32.5 months [37,38] but apparently, without improving the neurological status. Furthermore, a small number of patients have been included, respectively four and six.

Target treatment with *BRAF* inhibitors has been attempted with some responses in brain *BRAF* mutant gliomas [39]. Clinical trials are on-going to evaluate concurrent use of *BRAF* and *MEK* inhibition which could provide a better response [40].

5. Conclusion

The scarceness of IMSCTs is the first challenge. The lack of specific clinical symptoms, and imaging features at the time of diagnosis, is also problematic. Furthermore, management is hampered by surgical accessibility to the tumor, difficulties of

imaging evaluation and heterogeneity of current treatments. Published literature consists mostly of retrospective studies. This leads to a significant lack of statistical power. Cooperation between neurologists, neuro-radiologists, neurosurgeons, neuropathologists, and neuro-oncologists could improve management. We need larger series, and better knowledge of genetic alterations, specific to the spinal location, to access appropriate treatment guidelines.

Disclosure of interest

The authors declare that they have no competing interest.

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