v. 4, n. 99, 2024

International Journal of Health Science

Acceptance date: 18/11/2024

DIFFERENTIAL DIAGNOSIS BETWEEN INTRAMEDULLARY TUMORS AND SPINAL CORD MALFORMATIONS

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Abstract: Spinal cord arterio-venous shunts represent 5-9 % of all vascular pathologies of the central nervous system (CNS). Spinal cord arteriovenous malformation (SAVM) are much rarer. Intramedullary tumors (IMT) account for 10% of all spinal cord tumors. Most of them are glial tumors. IMT represent 2-4 % of all CNS tumors. Ependymoma is the most common in adults and astrocytoma is the most frequent in children and adolescents. IMT occurs most frequently in the cervical region, thoracic and lumbar regions respectively. Both pathologies may produce similar symptoms and sometimes it may be very difficult to differentiate one pathology from the other. These symptoms and signs will be discussed in this article and how to differentiate these two entities when possible according to them. This is the objective of this article.
Keywords: spinal cord- malformations- Intramedullary tumors- spinal cord tumors. – differential diagnosis

INTRODUCTION

Spinal cord tumor are classified as extradural which correspond approximately for 60% of all spinal cord tumors and are located outside the dura mater, intradural-extramedullary which are located within the dural but outside the spinal cord and represent 30% of all spinal cord tumors and while IMT represent 10% of all spinal cord tumors[1]. IMT account for 2-4% of all CNS tumors, ependymoma is the most common in adults and astrocytoma is the most common type in children. The most common type overall is ependymoma[2]. Metastatic IMT are rare. Most IMT are benign, however 7-30% of astrocytomas may be malignant[3]. IMT are most common at the cervical region (33%), followed by thoracic (26%) and lumbar (24%) levels[4].

A multicentric retrospective study showed that symptoms are variable and 39,2% of patients complain of spinal pain, 12.5% occurring at the cervical level, while thoracic 16.7%; and 10% lumbar. Motor troubles were present in 77,5% of patients and lower limbs being affected in 50% of them. Sensory symptoms affected 29,2% of the patients, and paresthesia being the most common (10%). Sphincter and vesical disorders occurring in 46,7% of the patients. Genital disturbances were seen in 17,5 of the patients[5].

Spine arterio venous shunts are 3-4% of all intradural lesions and 5-9% of CNS vascular pathologies[6]. Spinal cord vascular lesions can be classified as neoplastic (hemangioblastoma and cavernous malformation), aneurysms and spinal cord arteriovenous lesins which can be subdivided into arteriovenous fistulas and arteriovenous malformation[7]. This is a complex subject and another classification considers three types of spinal cord shunt. Intramedullary arteriovenous malformation, perimedullary arteriovenous fistula and dural arteriovenous fistula[8]. In this article intramedullary arteriovenous malformation is referred to as SAVM. Another similar classification is proposed into four categories: type I dural arteriovenous fistula, type II intramedular glomus arteriovenous malformation type III juvenile or combined arteriovenous fistula and type IV intradural perimedullary arteriovenous fistula[9]. Type II is described as SAVM in this article.

Most of the SAVM are found in the thoracic and lumbar levels (70%)[6]. Patients with SVAM have no gender predilection, typically are in their second to fourth decades and present with acute or subacute symptoms due to subarachnoid hemorrhage or venous trhombosis[10]. Variable clinical presentations may occur like myelopathy, back pain as well sensory-motor symptoms. Sudden neurological deterioration may occur even in the absence of hemorrhage probably to venous congestion and thrombosis (Foix Alajounanine syndrome).

DISCUSSION

Intramedullary tumors or arteriovenous malformation may produce similar symptoms like pain, sensory-motor disturbances and differential diagnosis may be really difficult. Images are very helpful and must be considered. Symptoms depend on their size and location.

Intramedullary tumors are associated with cysts in 70% of the cases. These cysts may be intratumoral (within the tumor, with peripheral enhancement and they occur especially associated with ganglioglioma, ependymoma and astrocytoma) or reactive (non tumoral) due to dilatation of the central canal, rostral or dorsal to the tumor itself, do not enhance and are present in 60% of all intramedullary tumors[11].

Spinal cord expansion may occur, as well as high signal intensity on T2 images. Even low grade IMT enhance to some degree however tha absence of enhancement does not exclude an IMT[12].

SAVM are associated with dilated vessels, signal voids from high velocity flow, increased cord signal due to edema. Angiography is the exam of choice[13].

The objective of the treatment for IMT is gross microsurgical total surgical resection while preserving spinal stability and neurological status. Complete tumor resection is higher at cervical levels, which exhibits better functional outcome than thoracic or lumbar located IMT. Nevertheless IMT extending more than 3 spinal segments have postoperative worsening significantly increased[14]. Some cases may require fusion instrumentation. Evoked potential may also be helpful.

Gross resection can improve patient survival but it may worsen neurological déficits. The decision of complete or subtotal resection will depend on pre and intraoperative findings. Considering that chemotherapy and radiotherapy are of limited efficacy for IMT[15].

SAVM often requires a microsurgical and embolization approaches. It also requires a team wih expertise in angiogaphy. Both IMT and SCVM are chalenging entities.

CONCLUSION

Some cases it is very difficult to establish an accurate preoperative diagnosis of an intramedullary lesion, even with a neurological exam, and images. Magnetic resonance image mimics IMT[16] and a preoperative angiography is very useful nevertheless it is technically challenging. Rational decisions must be taken by the team which is in charge taking into consideration possible intraoperative and postoperative risks.

DISCLOSURE

The author has no financial support with any organization or any financial conflict with the subject matter

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