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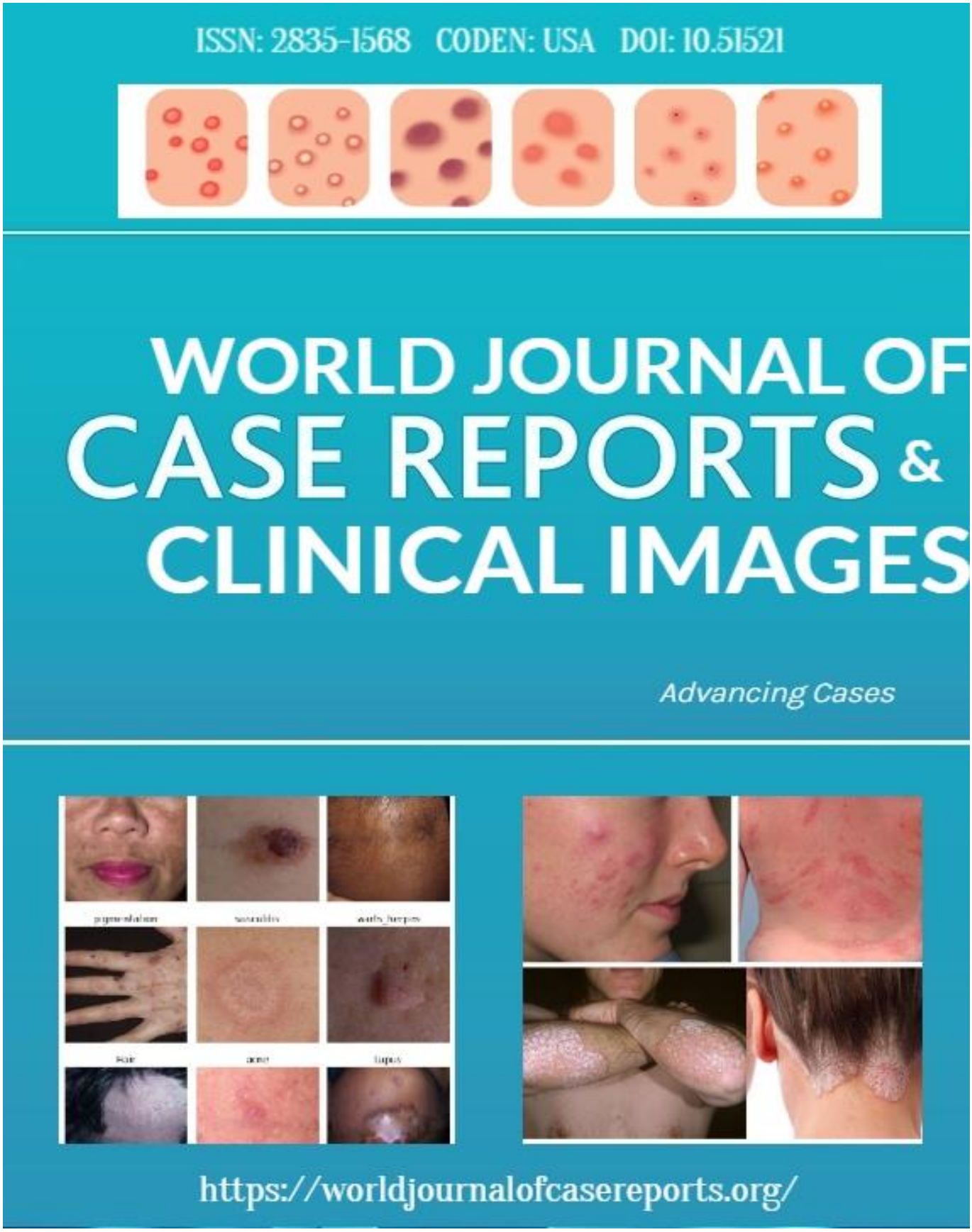

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CASE REPORT

Cauda Equina Cavernoma at level L1 Concurred with Intervertebral Disc Herniation at Level L5-S1: A Case Report of Misdiagnosis.

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Abstract

Vascular tumors show an extremely wide spectrum of morphologic appearances and clinical behavior. Hemangiomas or cavernomas occupy a gray zone and, because of their mass effect and localized nature, are designed frequently as tumors. Cavernous angiomas are uncommon vascular malformations found in all locations within the central nervous system. Incidence reported lie between 0.02 and 0.9%. Within the spinal cord they are most likely intramedullary and they account for 5-12% of all vascular lesions [1], [3]. Cauda equina is the least frequent site [3]. We show a case of misdiagnosis of a 45-year-old man complained of low back pain six months prior to surgery. A magnetic resonance imaging (MRI) scan revealed an L5-S1 bulky disc herniation extruding to the center of the lumbar spinal canal and compressing the thecal sac and the L5 left nerve root. An intracanal structure, at L1 level, was found and a tumor was suspected. A contrast enhanced MRI showed a round mass of 4mm diameter, slightly enhanced by gadolinium on T1W scans. Radiological diagnosis was a schwannoma. After neurosurgical and radiological evaluation, the patient underwent to the surgery. Tumor was pathologically confirmed to be cavernoma. Post-surgery, symptoms decreased and the hypoesthesia recovered completely. There were no post-operative complications. There are very few descriptions (to our knowledge, < 30 cases [2] are documented in the literature) of intradural extramedullary cavernomas of the cauda equina. The most frequent presentation of cavernomas of cauda equina is between L1 and L3 [2,3], sometimes extended to more than one level. In our case the level was T12-L1, extended more in L1, on the right side, the presentation was sub-acute. Surgical intervention was direct to the disc herniation, secondary to the exeresis of the tumor. Because of intra-operating and histological findings, the acute presentation was more to be appointed to the cavernomas lesion, so the surgery we performed was precise and effective due the radiological misdiagnosis and sub-acute clinical manifestation. In our opinion their management should be a complete microsurgical removal of the tumor not only to obtain a neurological improvement in symptomatic cases, but also to achieve a definitive diagnosis.

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Introduction

Vascular tumors show an extremely wide spectrum of morphologic appearances and clinical behavior in which the line between neoplasia and malformation remains undefined. Hemangiomas or cavernomas occupy a gray zone and, because of their mass effect and localized nature, are designed frequently as tumors. Cavernomas are soft, spongy masses composed of large cavernous blood-filled vascular spaces, separated by small connective tissue stroma [1,2]. Cavernous angiomas are uncommon vascular malformations found in all locations within the central nervous system. Incidence reported lie between 0.02 and 0.9%. Within the spinal cord they are most likely intramedullary and they account for 5-12% of all vascular lesions [1,3]. Cauda equina is the least frequent site [3], so they are rare in this location. Cavernomas follow an autosomal-dominant pattern of inheritance and have no gender predilection [3] Gene mutations involving endothelial junctions (particularly of the CM genes) contribute to the multiple cavernous malformations variant [2]. Clinical symptoms may occurs due to sub-acute-chronic or acute intratumoral hemorrhage which may cause an increase in tumor volume, or due to extra-lesion hemorrhage such as sub-arachnoid hemorrhage [2,3]. In the first case, a compression of cauda equine may occurs, resulting in cauda equina syndrome with asymmetric lower limb symptoms and/or sphincter deficits. Cavernomas are angiographically occult lesions. Diagnosis gold standard is MRI with some characteristic radiologic features, such as: well-defined lesion with mixed intensity on T1W and T2W images for the presence of the already noted intra-lesion hemorrhage [3,4] The differentials diagnosis that can be taken into consideration are: disc pathology, neurofibroma's, meningioma's or schwannomas. [2] We describe here the case of misdiagnosis with a concur presence of disc herniation.

Case Presentation

A 45-year-old man complained of low back pain six months prior to surgery. Pain was irradiated to the left hip, the postero-lateral side of the left inferior limb and down to the sole of the left foot. There was no history of trauma, he underwent to conservative treatment with cortisone and FANS medications with slightly symptoms improvement. Two months before surgery his pain worsened with hypoesthesia to the left foot sole and toes. When the patient came to our attention, a physical examination was performed. Straight leg raising test (Lasegue test) was normal bilaterally. Muscle strength test demonstrated score of 5/5 in iliopsoas, quadriceps femoris, tibialis anterior, gastrocnemius and hamstring muscles. Hypoesthesia and dysesthesia were present to the left foot sole. Babinski test was absent bilaterally. Deep tendon reflexes were diminished bilaterally. Bladder and bowel functions were unaffected. A magnetic resonance imaging (MRI) scan revealed an L5-S1 bulky disc herniation extruding to the center of the lumbar spinal canal and compressing the thecal sac and the L5 left nerve root. An intracanal structure, at L1 level, was found and a tumor was suspected. A contrast enhanced MRI showed a round mass of 4mm diameter, slightly enhanced by gadolinium on T1W scans (Figures 1, 2).

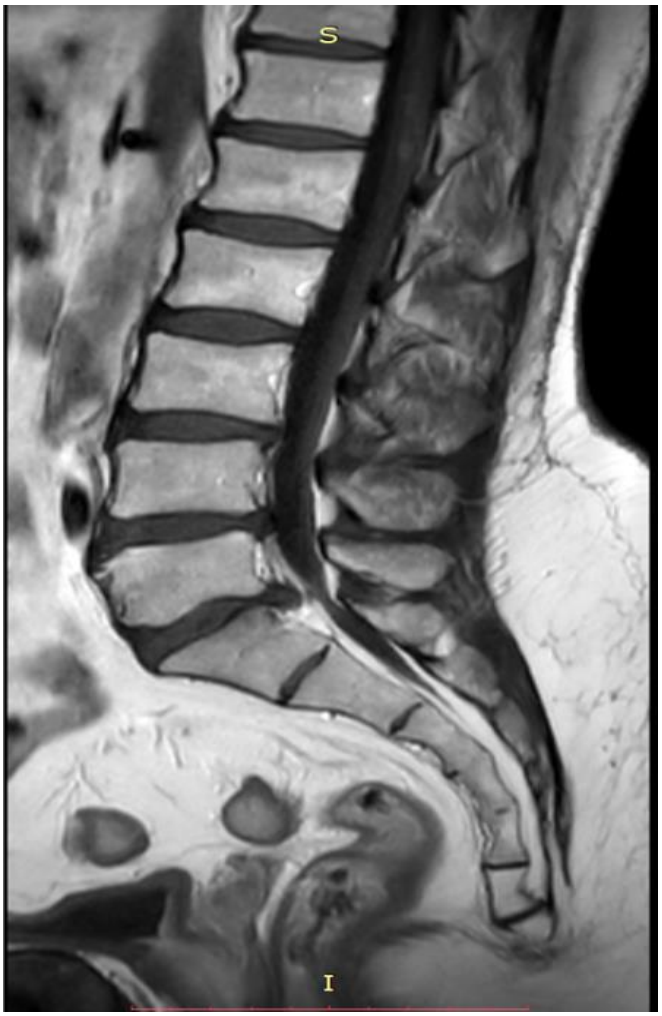

Figure 1: TW1 TSE weighted sagittal with mdc sequence showing intradural lesion with slightly enhanced image with gadolinium

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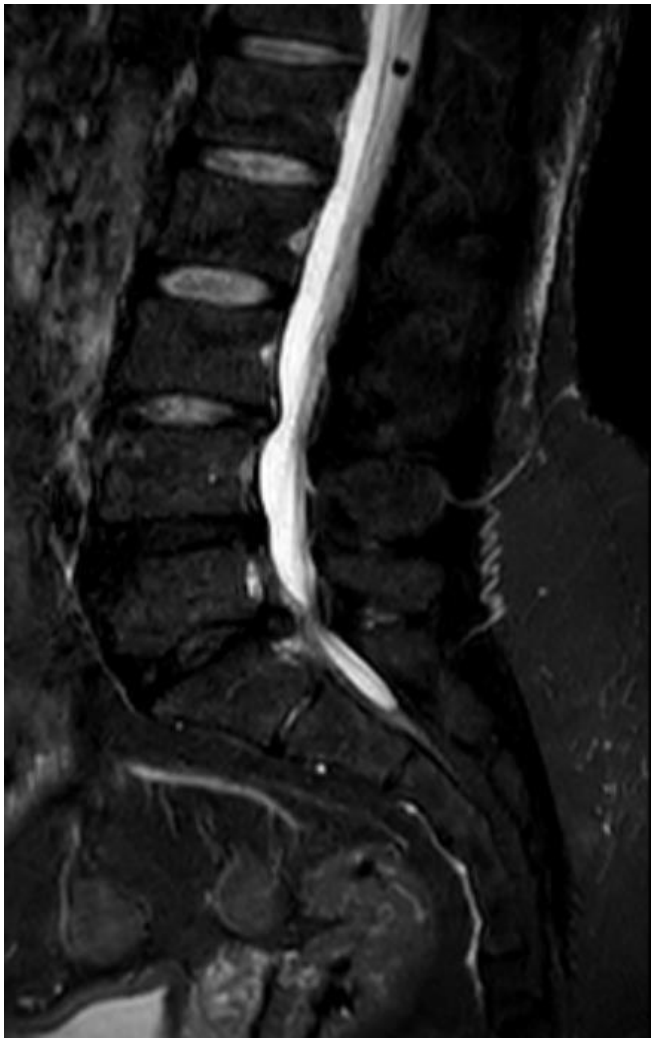

Figure 2: TW2 weighted sagittal STIR sequence MRI showing an intradural T12-L1 lesion and a discal herniation L5-S1.

The mass was intracanalar and intradural, located on the right side, among cauda fibers. Radiological diagnosis was a schwannoma. Prior to surgery the symptoms worsened and a bilateral hyposthenia and bilateral inferior limb pain appeared. After neurosurgical and radiological evaluation, the patient underwent to the surgery. Prone position and general anesthesia were used, two medial incisions at the level of T12-L1 and L5- S1 were performed with a right-sided and left-sided fenestration respectively. Through partial L1 laminectomy, microscopic resection of the tumor was performed. The mass did not appeared to be consistent with the radiological diagnosis. Instead, tumor was not afferent to a nerve root but extremely adherent to it. A L5-S1 lumbar interlaminotomy and a microscopic herniectomy and discectomy were performed. During surgery, intraoperative neurophysiological monitoring (IONM) was performed. MEP, SEP, EMG free running and EMG triggered showed no change during and after surgery. Tumor was pathologically confirmed to be cavernoma. Post-surgery, symptoms decreased and the hypoesthesia recovered completely. There were no post-operative complications. A three months MRI control scan was performed, the compression of thecal sac disappeared, the nerve root was free, and the tumoral mass was radiologically resected.

Discussion

There are very few descriptions (to our knowledge, <30 cases [2] are documented in the literature) of intradural extramedullary cavernomas of the cauda equina. Cavernomas can appear as space-occupying or hemorrhagic lesions, independently from their location. They can grow and increase their volume in time, either as consequences of repeated intralesional hemorrhage which can lead to thrombosis, fibrosis and enlargement of cavernoma; or as result of intrinsic grow capacity of endothelium that compose the cavernomatous tissue or as result of neo-angiogenesis [5]. The most common clinical syndrome is an acute/sub-acute compression of cauda equina, known as cauda equina syndrome which can present itself as complete (back pain with sciatic bilateral pain, lower motor neuron deficit or sensory loss, sphincter and sexual dysfunctions) or incomplete (sciatic pain with lower limb deficit lacking of sphincter/sexual dysfunctions) [1,2,3,5]. Another onset can be the subarachnoid hemorrhage (SAH) syndrome (headache, nuchal rigidity and vomiting) which can be the only clinical expression or combined with cauda equina syndrome [1]. The most frequent presentation of cavernomas of cauda equina is between L1 and L3 [2,3], sometimes extended to more than one level. In our case the level was T12-L1, extended more in L1, on the right side, the presentation was sub-acute. The presence of disc herniation can create diagnosis discrepancy, as this case, maybe the full clinical presentation was occult by disc herniation symptoms. An accurate physical examinations an essential tool to avoid not identifying signs of the presence of a mass lesion. Secondary clinical presentation was an incomplete cauda syndrome, lacking of sexual or sphincter dysfunctions, unlikely for a diagnosis of schwannoma, but more frequent with a disc pathology. Surgical intervention was direct to the disc herniation (Figure 3), secondary to the exeresis of the tumor. Because of intra-operating and histological findings, the acute presentation was more to be

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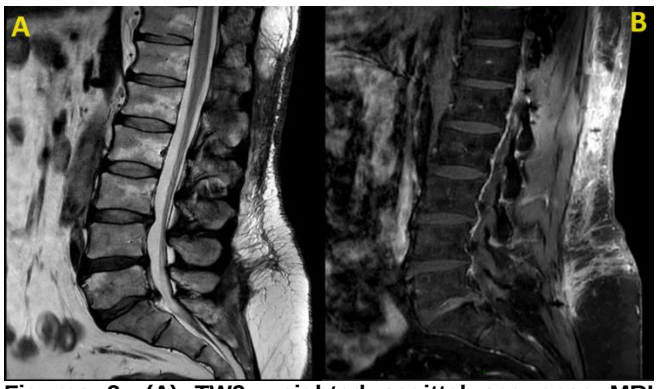
appointed to the cavernomas lesion, so the surgery we performed was precise and effective due the radiological misdiagnosis and sub-acute clinical manifestation. Post-op imaging shows the complete removal of the lesion, the symptomatology disappeared, and, in one year follow-up, there are no clinical presentation, nether radiological manifestation of a new growing mass lesion.



Figures 3: (A) TW2 weighted sagittal sequence MRI showing the results of interventions. (B) TW1 TSE weighted sagittal sequence showing the results of interventions

Conclusion

Given the benign nature of cavernomas or schwannomas of this particular site, given the presence of symptomatic disc pathology and excellent outcomes reported in literature for both pathologies , in our opinion their management should be a complete microsurgical removal of the tumor not only to obtain a neurological improvement in symptomatic cases, but also to achieve a definitive diagnosis of a lesion not always easy to interpret based only in radiological finding and physical examination preoperatively especially in cases in which the symptomatology is unseen by a disc pathology and the radiological diagnosis in unsure, as in this case.

Conflict Of interest: Authors did not provided any conflict of interest.

Ethical Consideration: None

Acknowledgements: Bot he the authors are equally contributed for the project and manuscript preparation

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