Subependymoma of the spine

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ABSTRACT

أن الأورام تحت البطانة العصبية هي أورام حميدة تحدث في الجهاز العصبي المركزي CNS و تصنف من الدرجة الأولى طبقاً لتصنيف منظمة الصحة العالمية (WHO grade I). الأماكن الشائعة لحدوث هذه الأورام هو البطينات الجانبية والبطين الرابع. يندر حدوث هذه الأورام في الحبل الشوكي. تتشابه هذه الأورام الحميدة في صفاتها الإشعاعية مع أورام أخرى خبيثة شائعة. نستعرض في هذا التقرير حالة لمريض يبلغ من العمر 61 عام مصاب بورم تحت البطانة العصبية وألم أسفل الظهر منذ 3 أعوام. ظهر في الحبل الشوكي عند مستوى الفقرات الظهرية العاشرة وحتى القطنية الأولى. تم عرض المظاهر السريرية والإشعاعية ودراسات التشريح المجهري. تمت مراجعة الأدبيات التي تستعرض حدوث مثل هذه الأورام في الحبل الشوكي.

Subependymoma is a rare benign CNS tumor (WHO grade I). The common sites of origin for this type of tumor are the lateral and the fourth ventricles. A spinal intramedullary location is rare. Radiological features of intramedullary subependymoma can mimic more common and more aggressive tumor types. We report a case of a 61-year-old male who presented with a 3-year history of low back pain, associated with right lower limb progressive weakness and sensory numbness. An MRI of the lumbar spine revealed an intradural intramedullary lesion extending from the level of T10-L1. Clinical presentation, radiological, and pathological studies of this case of subependymoma are presented. The incidence of spinal subependymoma was also discussed in light of a literature review.

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Subependymoma is a rare neoplasm of the CNS. It composes less than 1% of all brain neoplasms. These tumors are slowly growing indolent benign lesions, corresponding histologically to WHO grade I.[1] They are usually found in the floor of the fourth ventricles (50-60% of cases) followed by the lateral ventricular wall (30-40% of cases).[1,2] Around 50% of cases of these tumors are clinically silent and found incidentally at autopsy.[3] Rare symptomatic subependymoma accounts for approximately 0.7% of all symptomatic intracranial neoplasms,[1,2] and 8.3% of all tumors of ependymal origin.[3] The symptomatic ones are rare in childhood and tend to occur in the fifth and sixth decades of life.[1,2] They occur very rarely in the spinal cord, accounting for only 2% of all symptomatic subependymoma cases in the literature. To date less than 50 cases of spinal subependymoma are reported in the literature.[4] Spinal intramedullary subependymoma lesions are reported more commonly in the cervical region. However, both thoraco-lumbar and extra-medullary examples have been described as well. Our objective in presenting this particular case is to describe the clinical, radiological, and histopathological studies of this rare but benign tumor that can mimic more aggressive intramedullary tumors that commonly occur in this location.

Case Report. A 61-year-old man presented with a 3-year history of low back pain and right lower limb progressive weakness and numbness. He denied any sphincteric control symptoms. He was previously diagnosed to have an L3-L4 disc herniation based on a CT scan of the lumbar spine and was treated elsewhere using per-cutaneous laser discectomy with no clinical benefits. Neurological examination revealed a right lower limb weakness grade 4/5 in the major muscle groups, hyper-reflexia, increased muscle tone, and up-going planter response. He also had slightly reduced pinprick sensation at both lower limbs with a T12 sensory level. The left lower limb exam was within normal limits. Radiological investigation included a detailed thoraco-lumbar MRI study, which revealed a diffuse intradural intramedullary lesion extending from the level of T10 until the level of L1 expanding the spinal cord tissue

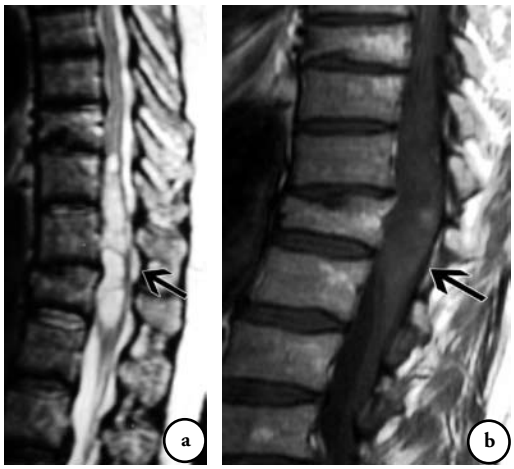

Figure 1 - Sagittal MRI **a)** T2 showing an intramedullary lesion extending from T10-L1. The lesion is hypo-intense, lobulated, and well demarcated. **b)** T1 with iso-intense consistency of the tumor.

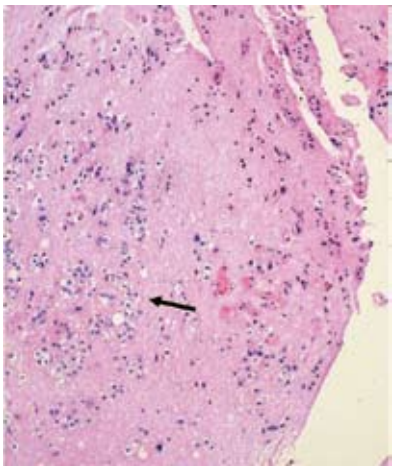

Figure 2 - Histopathology slide showing part of the neoplastic tissue, characterized by highly fibrillary lesion with prominent clustering of nuclei (arrow). The nuclei are markedly uniform with moderate pleomorphism (Hematoxylin & Eosin x 200).

more deviated toward the right side of the cord. The lesion did not enhance after intravenous gadolinium injection. He also had some degenerative changes at multiple lumbar levels (**Figure 1**). He underwent bilateral complete T11, T12, and partial T10 and L1 laminectomies. Intra-operatively the dura was tense. The spinal cord appeared swollen. Dorsal myelotomy was carried out under microscopic magnification. The lesion was yellowish in color, firm in consistency, and multi-lobulated without clear lines of demarcation between the tumor and the spinal cord tissue. It was mildly vascular. The patient had an uneventful peri-operative course with no post-operative complications. His motor weakness improved considerably at a 6 months postoperative evaluation. The histopathological examination of the lesion reported a highly fibrillary lesion with prominent clustering of nuclei, the nuclei are markedly uniform with moderate nuclear pleomorphism, and with relative lack of perinuclear cytoplasm (**Figure 2**).

Discussion. We describe a rare case of a 61-year-old male harboring a spinal subependymoma affecting the thoraco-lumbar junction region of the cord. On reviewing the literature, subependymoma was reported in different age groups ranging from 6-76 years.[1-3] Most patients were in the fifth and sixth decades of life.[1-3] Around two thirds of the patients were male.[1-3] The most common sites within the spinal cord affected by this rare pathology were cervical followed by cervicothoracic then upper lumber region.[4] A case of a holocord cord subependymoma was also reported.[5] The duration of symptoms before diagnosis ranged from a few months to several years.[1-4] The clinical presentation is similar to other intrinsic spinal cord tumors with an admixture of sensory, motor, pain, and sphincter disturbances. The MRI features of subependymoma include spinal cord enlargement, intra-spinal cysts, hypo-intense signal changes on T1 weighted images, and occasionally faint enhancement after gadolinium injection.[6,7] A distinctive feature of subependymoma as compared to classical spinal cord ependymoma is the eccentric location within the spinal cord.[6,7] Most tumors were sharply circumscribed with a clear line of cleavage at surgery. However, cord tissue infiltration was reported in some cases.[8] Histologically, subependymoma is classified within ependymal tumors as it contains rosettes of definite ependymal type. It shows evidence of ependymal differentiation at the ultra structural level and presents transitional or mixed neoplasms harboring regionally distinct elements, morphologically indistinguishable from endymoma.[1-3] The spinal subependymoma originates near the central canal where the subependymal cells exist. These tumors are biologically benign with a very low proliferative index.[9] Thus, prognosis depends largely on surgical factors since only one case of tumor recurrence was reported in the literature after gross total resection.[2] No CSF seeding has been reported to date. Some authors reported occasional cases with sarcomatous transformation.[10] A mixed variant of subependymoma and ependymoma (WHO: grade II) as described above, accounts for around 15% of symptomatic subependymoma.[1,10] This variant was described in younger aged patients with more rapid and infiltrative growth, thus, the ependymal component determines the outcome of these tumors and should be searched for in all subependymoma specimens. The importance in diagnosing these neoplasms lies in the fact

that subependymoma is completely benign and cured by surgical resection without the need for radiation therapy.[1-10]

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