



Lumbar Intradural Epithelioid Sarcoma: Case Report

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Abstract

Background: Intradural spinal tumors are rare lesions as they represent 4 - 10% of all central nervous system tumors and intramedullary tumors have an incidence of less than 1/100,000. The clinical presentation of primary spinal cord tumors is determined by their location and size with pain being the most common symptom. Mesenchymal tumors are coincided very rarely in intradural space. Initial therapy should involve radical resection of the tumor, followed by adjuvant therapy if malignant features are involved.

Case Presentation: Hereby; we are presenting a 22 years old male patient with intradural intraaxial lesion at L3 spinal level. Histopathological examination of the tumor resulted in mesenchymal sarcoma with epithelioid differentiation. Total surgical resection was preceded by adjuvant radiotherapy.

Conclusion: Although prevalence of epithelioid sarcomas are rare, they should be included in the differential diagnosis of intradural spinal tumors. Surgical resection is the treatment of choice and since local recurrence rates are high, adjuvant radiotherapy should be considered.

Introduction

Intradural spinal tumors are rare lesions with a prevalence of 1/100,000 (1). Mostly dependent on location and size; these tumors present with radicular pain and motor or sensory deficit. Although consensus on management of these tumors have not been reached yet; gold standart treatment is surgical resection. Epithelioid sarcoma is a slow-growing, morphologically benign soft tissue tumor and rare with most common locations being limbs, pelvis, perineum, genitalia and mediastinum. In this report; we are presenting a 22 years old patient with epithelioid sarcoma located in intradural intra-axial compartment at L3 level.

Case Presentation

A 22 years old male patient presented with a complaint of sacral pain with extension of pain to groin and both legs since 7 months. In his medical history; he did not reveal any diseases.

Neurological examination was normal without any motor and sensory deficit. Deep tendon reflexes were normoactive and there were no pathological reflexes. He had axial pain while flexion and extension. Initially an MRI of the lumbar spine was performed (Figure 1). MRI scan revealed an intradural intra-axial lesion with a size of 11*8*16 mm at L3 spinal level was located ventral to equinal fibers. In contrast weighted T1 imaging, lesion was homogenously contrasted with regular borders. Patient was operated in prone position with a vertical midline incision. L3 laminectomy was performed. Tumor was exposed after dural incision. Tumor was in grey color, soft and encapsulated. It was adherent to equinal fibers and was carefully dissected microscopically with neuromonitorization of lumbar and sacral nerves. It was totally excised (Figure 2). Patient was neurologically intact after tumor removal. He was ambulated 24 hours postoperatively and discharged on day four without further peroperative complications.

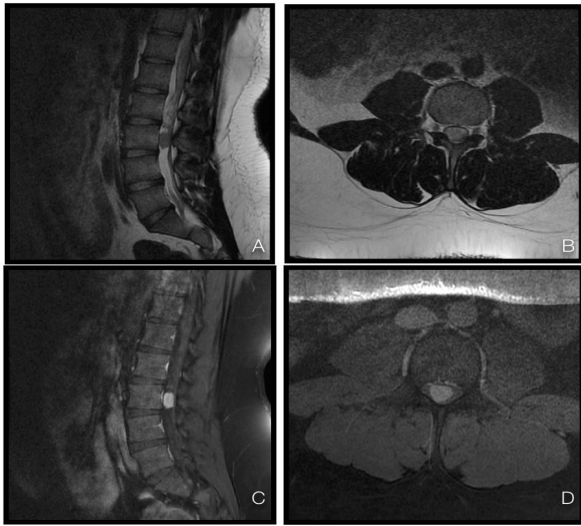


Figure 1: Preoperative imaging of the patient 1A. Sagittal T2 weighted MRI of the patient showing an isointense lesion with well defined borders, located intradurally in L3 segment. 1B. Axial T2 weighted MRI of L3 segment. 1C. Contrast weighted T1 sequence MRI showing homogeneously contrasted lesion. 1D. Axial T1 contrast weighted imaging of L3 segment.

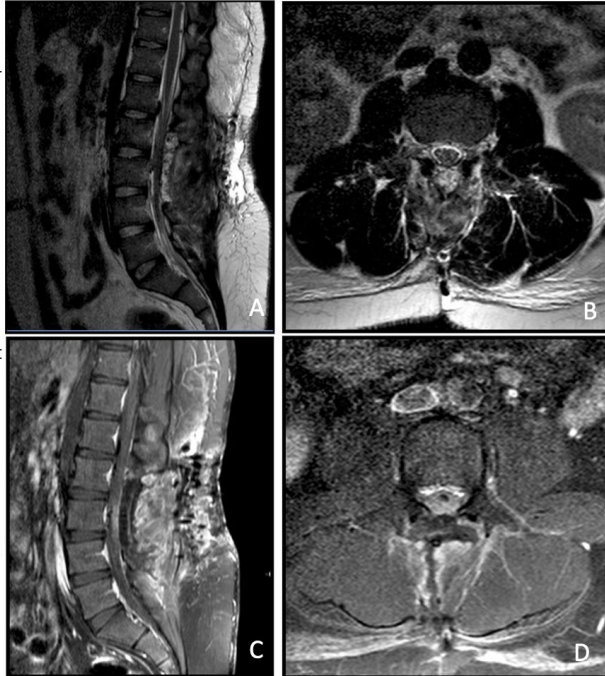


Figure 2: Postoperative imaging of the patient shows total removal of the tumor. 2A. Sagittal T2 weighted MRI. 2B. Axial T2 weighted MRI of L3 segment. 1C. Contrast weighted T1 sequence MRI. 1D. Axial T1 contrast weighted imaging of L3 segment.

Histopathological examination revealed mesenchymal sarcoma in epithelioid morphology with neural differentiation. Ki-67 (Cell-Marque/SP6) index was 7-8%. It was S100, Vimentin, NSE (neuron specific enolase) and CD56 positive. It was GFAP and Synaptophysin negative. Patient was initiated on radiotherapy after the histopathological diagnosis.

Discussion

In this case report, we present a 22 years old patient with epithelioid sarcoma located in L3 intra-axial compartment. Intradural spinal tumors are rare lesions as they represent 4-10% of all central nervous system tumors; approximately 2/3 are located extramedullary; the rest of them are intramedullary tumors, with an incidence of less than 1 in 100 000 habitants [1]. They are less common than intracranial tumors and most of them are in benign morphology [2]. Seventy percent of intradural tumors localize in the cervical or thoracic spine [1,3,4].

Central nervous system sarcomas are rare with most common sites being extremities, pelvis, genitalia and trunk [5]. Subgroups consist of malignant fibrous histiocytoma, chondrosarcoma (classical and mesenchymal type), rhabdomyosarcoma, liposarcoma, fibrosarcoma, angiosarcoma and leiomyosarcoma [6]. Recurrence and metastasis rates are high [6]. It is thought to arise from synovioblastic mesenchyme [5]. Histopathologically tumors may consist of epithelioid and/or spindle cells with centrally necrotic nodules^o. Immunohistochemically vimentin and cytokeratin expressions are frequent and EMA which is an epithelial cell marker may also be expressed [6]. In a study, S100 protein positivity was observed in 1 of 88 epithelioid sarcomas. However; scattered S100 positive dendritic cells may be observed [7]. In the case that we are presenting; the tumor is both vimentin and desmin positive with cooccurring positivity in S100 protein. Subgrouping of the mesenchymal tumor was challenging because of these indistinct molecular patterns. On the other hand, EMA positivity supported the diagnosis of an epithelioid sarcoma.

Epithelioid sarcoma usually occurs in young adults between 15-35 years of age. Most commonly they are located in fingers, hand, forearm, lower leg and knee. Despite high recurrence rates, 5-year survival may vary from 65 to 100% and depends on tumor infiltration, pulmonary metastasis, male gender and rhabdoid cytomorphology. In our case, tumor was isolated with no local or distant metastasis [8]. It was confirmed via a PET (positron emission tomography) scan. The tumor in our report has intradural intraaxial location. Differential diagnosis includes epidermoid cysts, lipoma, ependymoma, astrocytoma and metastasis. Lipomas are usually hyperintense in T1 and T2 weighted imaging without contrast enhancement. Ependymomas mostly show heterogenous contrast enhancement and astrocytomas have peritumoral edema and frequently shows exophytic growth pattern. Even though

sarcomas are usually known to be located in soft tissue and osseous compartments, it may rarely be seen intradurally and should be included in differential diagnosis of intradural intraaxial tumors.

Conclusion

Mesenchymal epithelioid sarcomas may be seen in spinal intradural space with rare occurrence, therefore they should be included in the differential diagnosis of intradural spinal tumors. Initial therapy should involve radical resection of the tumor, followed by adjuvant therapy if malignant features are involved.

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