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RESEARCH ARTICLE

PRIMARY PARASPINAL LEIOMYOSARCOMA INVADING THE DORSAL SPINAL CANAL:A CASE REPORT AND REVIEW OF THE LITERATURE

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Abstract

Leiomyosarcoma (LMS) is one of the most common subtypes of soft tissue sarcoma in adults and can occur in almost any part of the body, which presents a high propensity to metastasize to various locations. Primary leiomyosarcomas of the paraspinal soft tissues are exceedingly rare. We report a case of a primary leiomyosarcoma of the dorsal paraspinal musculature invading the spinal canal in a 36-year-old man, successfully treated with surgery.

Introduction:

Leiomyosarcomas (LMSs) are variably aggressive tumors of smooth muscle cell origin, with a high propensity to metastasize to various locations. They fall under the broad category of soft tissue sarcomas (STS) that constitute 0.7% of all malignancies. They have been reported to constitute 5–16% of all soft tissue sarcomas[1]. They commonly arise in the uterus and deep soft tissues, especially of the retroperitoneal space, but may also arise in the dermis, subcutaneous tissues, gastrointestinal tract, and medium- to large-sized veins[2].

Primary leiomyosarcomas of the paraspinal soft tissues are exceedingly rare[3]. We report a case of a primary leiomyosarcoma of the dorsal paraspinal musculature invading the spinal canal in a 36-year-old man. Successfully treated with surgery.

Case Report

he is a 35-year-old man, with no notable pathological history, who was admitted to our institution for the management of chronic back pain with gait disturbance, the clinical examination found a dorsal paraspinal mass measuring 6 x 4 cm with dorsal spinal syndrome and posterior cord syndrome without neurological deficit,

MRI showed dorsal lesion process; measuring 5.8 x 3.7 cm; next to the posterior arch of D3 and D4, the process has a tissue signal, it enhances heterogeneously, infiltrates the paravertebral muscles, and is responsible for lysis of the spinous and blades of D3 and D4, with invasion of the canal spinal cord responsible for posterior spinal cord compression(fig. 1). the patient has benefited a surgical treatment, the procedure included excision of the extra-spinal part, laminectomy from D1 to D5, and excision of the intra-spinal part of the tumor. post-surgical evolution was favorable with disappearance of back pain and improvement in walking after functional rehabilitation.pathological examination showed the appearance of a spindle cell tumor. Immunohistochemically, these tumor cells were positive for desmin, alpha-smooth muscle actin (SMA), and h-caldesmon, Immunostaining using Ki 67 revealed a proliferation index inferior than 50%. These features provided the diagnosis of leiomyosarcoma(fig. 2).

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Discussion:

The incidence of LMSs has been reported to be in the range of 5–16% of all soft tissue sarcomas[1].As in soft tissue sarcomas(STS) in general, the overall incidence of LMS increases with age and peaks at the seventh decade of life. The sex incidence greatly depends on tumor location, with women comprising a clear majority of patients with retroperitoneal and inferior vena cava LMS, whereas there is a mild male predominance in no cutaneous soft tissue sites and cutaneous LMS[4].Our patient was a 36-year-old man. Her age was three decades younger than the mean age reported in the literature.

LMSs are evenly distributed over various locations in the body. Retroperitoneal LMSs are the most common lesions and are associated with a grave prognosis considering their metastatic potential. Somatic (or external) soft tissue LMSs are generally considered to be of intermediate metastatic potential and are sub-classified as cutaneous, subcutaneous, or deep soft tissue tumors, being present over the thighs, arms, or trunk[1]. Our patient had a paraspinal LMS, which is considered a deep soft tissue LMS.The 5-year survival rate of somatic soft tissue LMSs has been reported to be 61% compared to the 21% survival rate of retroperitoneal LMS[5].

To our knowledge,this is the sexth report of a primary leiomyosarcoma of the paraspinal musculature invading the spinal canal, three cases involved in the cervical spine, tow in the dorsal spine and one in the dorsal spine **(TABLE 1)**.

The origin of spinal leiomyosarcomas is not clear. Most are presumed to arise from local blood vessels[2], although an origin from pluripotent mesenchymal cells, perhaps of the leptomeninges, has also been suggested[6]. Additionally, leiomyosarcomas may arise in the osseous spine, sellar region, and other locations secondary to therapeutic radiation[7].

Histologically, these tumors consist of intersecting bundles of spindle-shaped cells, with typically elongated nuclei having blunt ends. The cytoplasm is eosinophilic and fibrillar, with occasional vacuoles seen within [8]. The degree of nuclear pleomorphism varies from case to case [9].The majority of LMS are reactive for alpha-smooth muscle actin, desmin, and h-caldesmon on immunohistochemistry, although none of these markers is specific for smooth muscle differentiation.

On MR imaging, intra- or paraspinal leiomyosarcomas are usually isointense or hypointense on T_1-weighted and hyperintense on T_2-weighted images (hypointense to cere- brospinal fluid) and homogeneously enhance, and may be lobulated [6]. These leiomyosarcomas may thus appear very similar to schwannomas[6,3,10] or neurofibromas,especially when they involve neural foramina or nerve roots. despite rare, leiomyosarcomas should be incorporated in the differential diagnosis of paraspinal and spinal masses, including those involving the spinal roots and/or spinal canal.

A comprehensive preoperative physical examination should always be conducted. Any clinical suspicion of a malignancy should be increased by a large tumor size, rapid tumor growth, and involvement of bone and/or paraspinal soft tissues. A preoperative workup for an unknown primary tumor or metastatic disease should be performed in all such possible cases. Computed tomography scans of the chest, abdomen, and pelvis are necessary to determine the presence of metastatic disease or an occult primary tumor. In addition to diagnostic MR imaging, a preoperative CT scan of the spine to evaluate the extent of bone involvement may be useful in surgical planning.

Currently, there is no consensus in the literature on the treatment protocol of these tumors, and there have been no randomized trials in this regard. However, it is generally accepted that a multimodality treatment comprising surgery and adjuvant radiotherapy and chemotherapy would provide the highest chances of survival [10].

Conclusion:

Primary leiomyosarcomas of the paraspinal soft tissues are exceedingly rare, butshould be kept in mind as one of the possible diagnoses when a patient with a vertebral tumor is presented. The ability to perform a complete surgical resection at the time of initial presentation is the most important prognostic factor for survival.

- The authors confirm that they have no conflict of interest to declare

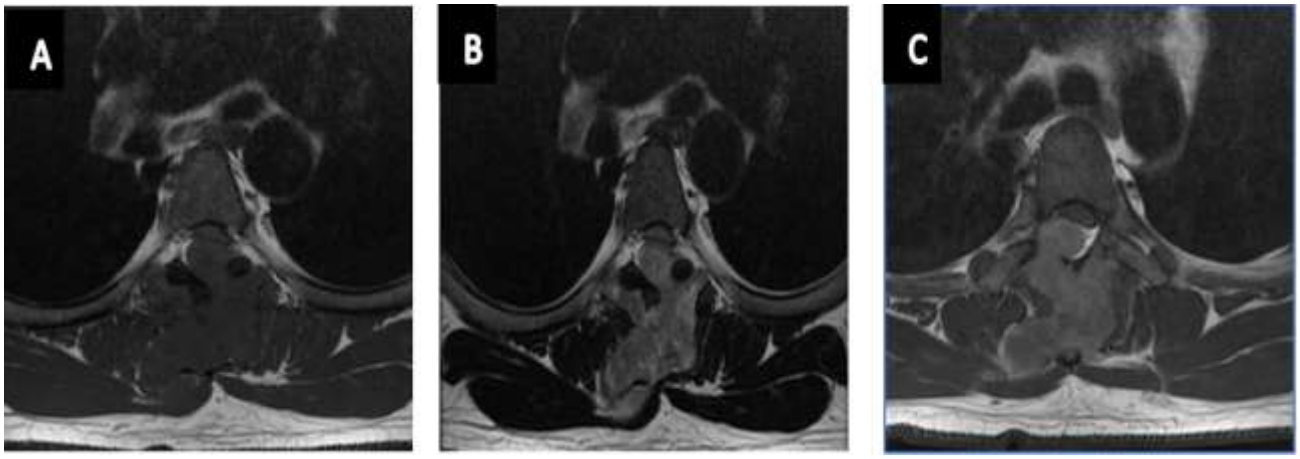

Fig.1:-Magnetic resonance images obtained in the patient. (A) T1-weighted axial magnetic resonance imaging section showing the iso-intense tumor, (B) T2-weighted axial magnetic resonance imaging section showing the hyperintense tumor,(C) T1-weighted axial magnetic resonance imaging section with gadolinium injection, showing homogenous contrast enhancement.

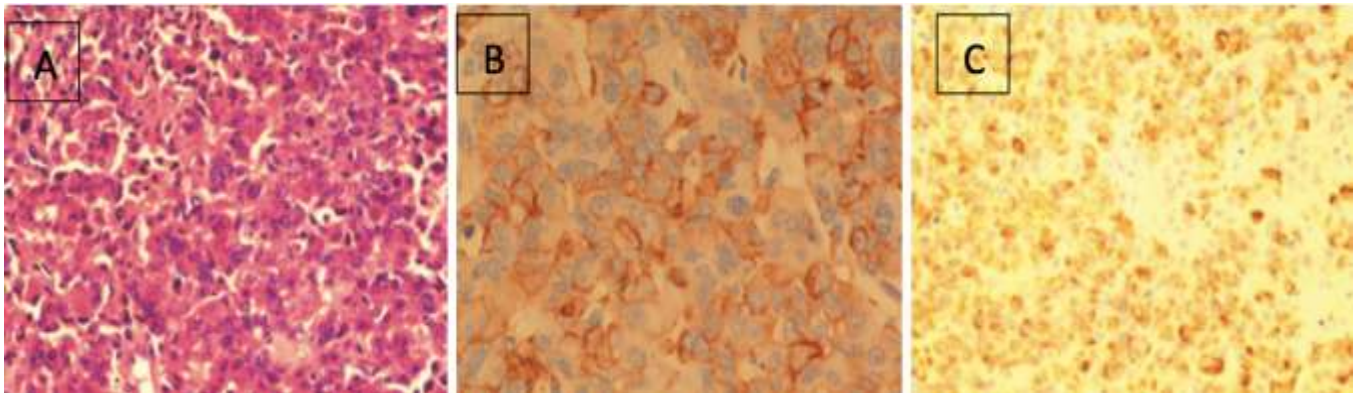

Fig. 2:- Histopathological photomicrotaphs. A: the pleomorphic spindle cells in fascicles. B: positive immunostaining for alpha-smooth muscle actin. C positive immunostaining for desmin.

Table 1:-Overview of previously reported cases on primary paraspinal leiomyosarcomas.

Report	Age/sex	Clinical features	location	treatment	outcome
Sengupta and Nag 1992	43/male	Neck pain, no deficits	Right lateral musculature C7-T1	Surgery + radiotherapy	4 years follow-up recurrence
Marshman et al. 2005)	61/female	Cervical myelopathy	C3-C5 posterior paraspinal musculature and ligamentum flavum	surgery	Lost follow-up
Lehman et al. 2007	45/male	Painful enlarging mass on right side of neck, no deficits	C1-C2 right posterolateral musculature	Surgery + chemotherapy + radiotherapy	6 years no primary site recurrence. Metastases present
Aksoy et al. 2002	70/female	Pain in the neck and right leg and sensory loss on the right side of body	T1-T3 right paravertebral mass	Surgery + chemotherapy	Not know
Ankush and all. 2016	34 /female	Low back arche with radicular pain in the right lower limb	L4-L5 right paraspinal mass	Surgery + external beam radiation therapy + chemotherapy	Metastatic disease at 6 months
Present case	36/mal	gait disorders and back pain	D3-D4 posterior paraspinal	surgery	no primary site recurrence, no metastatic disease at 6 months

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