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Contents

Thrice Monthly Volume 9 Number 10 April 6, 2021

MINIREVIEWS

- 2160 Tertiary peritonitis: A disease that should not be ignored
Marques HS, Araújo GRL, da Silva FAF, de Brito BB, Versiani PVD, Caires JS, Milet TC, de Melo FF
- 2170 SARS-CoV-2, surgeons and surgical masks
Khalil MI, Banik GR, Mansoor S, Alqahtani AS, Rashid H

ORIGINAL ARTICLE

Case Control Study

- 2181 Igaratimod promotes transformation of mononuclear macrophages in elderly patients with rheumatoid arthritis by nuclear factor- κ B pathway
Liu S, Song LP, Li RB, Feng LH, Zhu H

Retrospective Study

- 2192 Factors associated with overall survival in early gastric cancer patients who underwent additional surgery after endoscopic submucosal dissection
Zheng Z, Bu FD, Chen H, Yin J, Xu R, Cai J, Zhang J, Yao HW, Zhang ZT
- 2205 Epidemiological and clinical characteristics of 65 hospitalized patients with COVID-19 in Liaoning, China
Zhang W, Ban Y, Wu YH, Liu JY, Li XH, Wu H, Li H, Chen R, Yu XX, Zheng R
- 2218 Comprehensive clinicopathologic characteristics of intraabdominal neurogenic tumors: Single institution experience
Simsek C, Uner M, Ozkara F, Akman O, Akyol A, Kav T, Sokmensuer C, Gedikoglu G
- 2228 Distribution and drug resistance of pathogens in burn patients in China from 2006 to 2019
Chen H, Yang L, Cheng L, Hu XH, Shen YM

Observational Study

- 2238 Impact of simethicone on bowel cleansing during colonoscopy in Chinese patients
Zhang H, Liu J, Ma SL, Huang ML, Fan Y, Song M, Yang J, Zhang XX, Song QL, Gong J, Huang PX, Zhang H

Prospective Study

- 2247 Effect of suspension training on neuromuscular function, postural control, and knee kinematics in anterior cruciate ligament reconstruction patients
Huang DD, Chen LH, Yu Z, Chen QJ, Lai JN, Li HH, Liu G

CASE REPORT

- 2259 Turner syndrome with positive SRY gene and non-classical congenital adrenal hyperplasia: A case report
He MN, Zhao SC, Li JM, Tong LL, Fan XZ, Xue YM, Lin XH, Cao Y

- 2268** Mechanical thrombectomy for acute occlusion of the posterior inferior cerebellar artery: A case report
Zhang HB, Wang P, Wang Y, Wang JH, Li Z, Li R
- 2274** Bilateral retrocorneal hyaline scrolls secondary to asymptomatic congenital syphilis: A case report
Jin YQ, Hu YP, Dai Q, Wu SQ
- 2281** Recurrent undifferentiated embryonal sarcoma of the liver in adult patient treated by pembrolizumab: A case report
Yu XH, Huang J, Ge NJ, Yang YF, Zhao JY
- 2289** Adult onset type 2 familial hemophagocytic lymphohistiocytosis with *PRF1* c.65delC/c.163C>T compound heterozygous mutations: A case report
Liu XY, Nie YB, Chen XJ, Gao XH, Zhai LJ, Min FL
- 2296** Salvage of vascular graft infections *via* vacuum sealing drainage and rectus femoris muscle flap transposition: A case report
Zhang P, Tao FL, Li QH, Zhou DS, Liu FX
- 2302** Innovative chest wall reconstruction with a locking plate and cement spacer after radical resection of chondrosarcoma in the sternum: A case report
Lin CW, Ho TY, Yeh CW, Chen HT, Chiang IP, Fong YC
- 2312** Changes in sleep parameters following biomimetic oral appliance therapy: A case report
Singh GD, Kherani S
- 2320** Bone remodeling in sigmoid sinus diverticulum after stenting for transverse sinus stenosis in pulsatile tinnitus: A case report
Qiu XY, Zhao PF, Ding HY, Li XS, Lv H, Yang ZH, Gong SS, Jin L, Wang ZC
- 2326** Prolonged use of bedaquiline in two patients with pulmonary extensively drug-resistant tuberculosis: Two case reports
Gao JT, Xie L, Ma LP, Shu W, Zhang LJ, Ning YJ, Xie SH, Liu YH, Gao MQ
- 2334** Low-grade mucinous appendiceal neoplasm mimicking an ovarian lesion: A case report and review of literature
Borges AL, Reis-de-Carvalho C, Chorão M, Pereira H, Djokovic D
- 2344** Granulomatosis with polyangiitis presenting as high fever with diffuse alveolar hemorrhage and otitis media: A case report
Li XJ, Yang L, Yan XF, Zhan CT, Liu JH
- 2352** Primary intramedullary melanoma of lumbar spinal cord: A case report
Sun LD, Chu X, Xu L, Fan XZ, Qian Y, Zuo DM
- 2357** Proliferative glomerulonephritis with monoclonal immunoglobulin G deposits in a young woman: A case report
Xu ZG, Li WL, Wang X, Zhang SY, Zhang YW, Wei X, Li CD, Zeng P, Luan SD

- 2367** *Nocardia cyriacigeorgica* infection in a patient with pulmonary sequestration: A case report
Lin J, Wu XM, Peng MF
- 2373** Long-term control of melanoma brain metastases with co-occurring intracranial infection and involuntary drug reduction during COVID-19 pandemic: A case report
Wang Y, Lian B, Cui CL
- 2380** Solitary bone plasmacytoma of the upper cervical spine: A case report
Li RJ, Li XF, Jiang WM
- 2386** Two-stage transcrestal sinus floor elevation-insight into replantation: Six case reports
Lin ZZ, Xu DQ, Ye ZY, Wang GG, Ding X
- 2394** Programmed cell death protein-1 inhibitor combined with chimeric antigen receptor T cells in the treatment of relapsed refractory non-Hodgkin lymphoma: A case report
Niu ZY, Sun L, Wen SP, Song ZR, Xing L, Wang Y, Li JQ, Zhang XJ, Wang FX
- 2400** Pancreatic cancer secondary to intraductal papillary mucinous neoplasm with collision between gastric cancer and B-cell lymphoma: A case report
Ma YH, Yamaguchi T, Yasumura T, Kuno T, Kobayashi S, Yoshida T, Ishida T, Ishida Y, Takaoka S, Fan JL, Enomoto N
- 2409** Acquired haemophilia in patients with malignant disease: A case report
Krašek V, Kotnik A, Zavrtanik H, Klen J, Zver S

ABOUT COVER

Editorial Board Member of *World Journal of Clinical Cases*, Deb Sanjay Nag, Senior Consultant, Department of Anaesthesiology, Tata Main Hospital, C-Road (West), Bistupur, Jamshedpur 831 001, India. ds.nag@tatasteel.com

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Primary intramedullary melanoma of lumbar spinal cord: A case report

Le-Dong Sun, Xin Chu, Li Xu, Xiu-Zhen Fan, Yi Qian, Da-Ming Zuo

ORCID number: Le-Dong Sun 0000-0003-4129-4204; Xin Chu 0000-0002-7853-1296; Li Xu 0000-0003-2071-6922; Xiu-Zhen Fan 0000-0001-8696-8132; Yi Qian 0000-0003-3779-6973; Da-Ming Zuo 0000-0003-2003-9474.

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Le-Dong Sun, Xin Chu, Li Xu, Xiu-Zhen Fan, Department of Dermatology, The Fifth Affiliated Hospital, Southern Medical University, Guangzhou 510900, Guangdong Province, China

Yi Qian, The Fifth Affiliated Hospital, Southern Medical University, Guangzhou 510900, Guangdong Province, China

Da-Ming Zuo, Department of Medical Laboratory, Southern Medical University, Guangzhou 510515, Guangdong Province, China

Corresponding author: Yi Qian, MD, Associate Professor, The Fifth Affiliated Hospital, Southern Medical University, No. 566 Congcheng Avenue, Conghua District, Guangzhou 510900, Guangdong Province, China. 1328059119@qq.com

Abstract

BACKGROUND

Primary intramedullary melanoma is a very rare tumor, most frequently occurring in the cervical and thoracic spinal cord.

CASE SUMMARY

We present a rare case in which the primary intramedullary melanoma was located in the lumbar spine. A 56-year-old man complained of progressive intermittent pain in the lumbar area. Thoracic magnetic resonance imaging showed a spinal intramedullary tumor between the L3 and S1 levels. The tumor was resected entirely, and the diagnosis of malignant melanoma was confirmed by histopathology.

CONCLUSION

Primary melanoma of the spinal cord, particularly intramedullary localization, has rarely been reported in the previous literature. We describe a primary malignant melanoma of the lumbar spinal cord and discuss the challenges associated with the diagnosis.

Key Words: Intramedullary melanoma; Spinal cord; Lumbar; Magnetic resonance imaging; Histopathological examination; Immunohistochemical staining; Case report

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INTRODUCTION

Primary malignant melanoma of the central nervous system (CNS) accounts for about 1% of all melanoma cases^[1]. Primary CNS melanoma localized to the spinal cord is extremely rare, and most frequently involves the thoracic spine and the cervical spine. The diagnosis of primary intramedullary melanoma was based on histological examination and the exclusion of other lesions outside of the CNS^[2,3]. In this paper, we present the case of a 56-year-old male patient diagnosed with primary intramedullary melanoma of the lumbar spine that proceeded with surgical resection.

CASE PRESENTATION

Chief complaints

A 56-year-old man visited our hospital with radiating pain in the lower limbs for 2 wk.

History of past illness

The patient had been suffering from progressive intermittent pain in the lumbar area for approximately 3 years, and his pain was only partially relieved after bed rest. He then began to feel severe radiating pain in the left lower limb 2 wk before presentation.

Physical examination

The patient had mild kyphosis of the spine and intervertebral tenderness in the L4-L5 and L5-S1 disc spaces. Physical examination showed left hip tenderness and percussion pain. The muscles of the left lower limb were significantly atrophied relative to the right lower limb. Resultantly, the left lower limb was observed to be 3 cm shorter than the right one.

A Faber test of the left lower limb was positive. Straight leg elevation tests of both lower extremities were negative. Pain sensation and temperature sensation were normal in both lower limbs. Muscle tension in both lower extremities was also normal.

Dorsiflexion force of the left ankle joint was 3+, and plantar flexor muscle strength was level 3. The nail of the big toe was grade 0. Right ankle dorsiflexor force was 4, and plantar flexor force was 5. The anal sphincter contracted evenly and forcefully. Bilateral knee and Achilles tendon reflexes were normal. The physiological reflex was present, but the pathological reflex was not elicited.

Imaging examinations

Magnetic resonance imaging (MRI) analysis of the lumbar spine exhibited an intramedullary mass between the L3 and S1 levels. An impressive Schmorl node was visible in the L4 vertebral body (Figure 1).

Histopathological examination

Histopathological examination of the specimen showed that the tissue was composed of most polygonal and some spindle tumor cells. The tumor cells were relatively uniform in size, with rich and transparent cytoplasm. The nuclei of most of the cells were either round or oval, and they had large nucleoli. The tumor cells displayed a high degree of mitotic activity, but no necrosis was seen (Figure 2A). The isolated tumor was also subjected to immunohistochemical examination, which revealed positive staining for S-100, homatropine methylbromide (HMB)-45, and p53, indicating an intramedullary malignant melanoma (Figure 2B-D). The tumor cells expressed vimentin and did not exhibit positivity for the epithelial marker cytokeratin,



Figure 1 Magnetic resonance imaging analysis of spinal cord lesion. A: Sagittal T1-weighted magnetic resonance imaging (MRI); B: Sagittal T2-weighted MRI; C: Homogeneous contrast enhancement on axial T1-weighted images with gadolinium.

which indicates that the tumor had undergone the epithelial-to-mesenchymal transition process (Figure 2E and F). Additionally, immunostaining for Ki67 supported an appreciable proliferative activity, and the Ki67 proliferation index was about 10%-50% (Figure 2G).

FINAL DIAGNOSIS

The patient had no history or clinical manifestation of the primary cutaneous or ocular lesion. Based on the MRI analysis and histological examination, the final diagnosis was primary malignant intramedullary melanoma of the lumbar spinal cord.

TREATMENT

The patient underwent L4-S1 discectomy with total resection of a dark reddish-brown ventrally exophytic intramedullary tumor. The tumor was exposed after the dura was widely opened. It was black and firm, and invaded the L5 spinal nerve root. Titanium mesh implantation and pedicle screw fixation were used for the treatment.

OUTCOME AND FOLLOW-UP

The patient refused the molecular targeted therapy based on the drug sensitivity test. Unfortunately, the patient passed away 6 mo after the surgery.

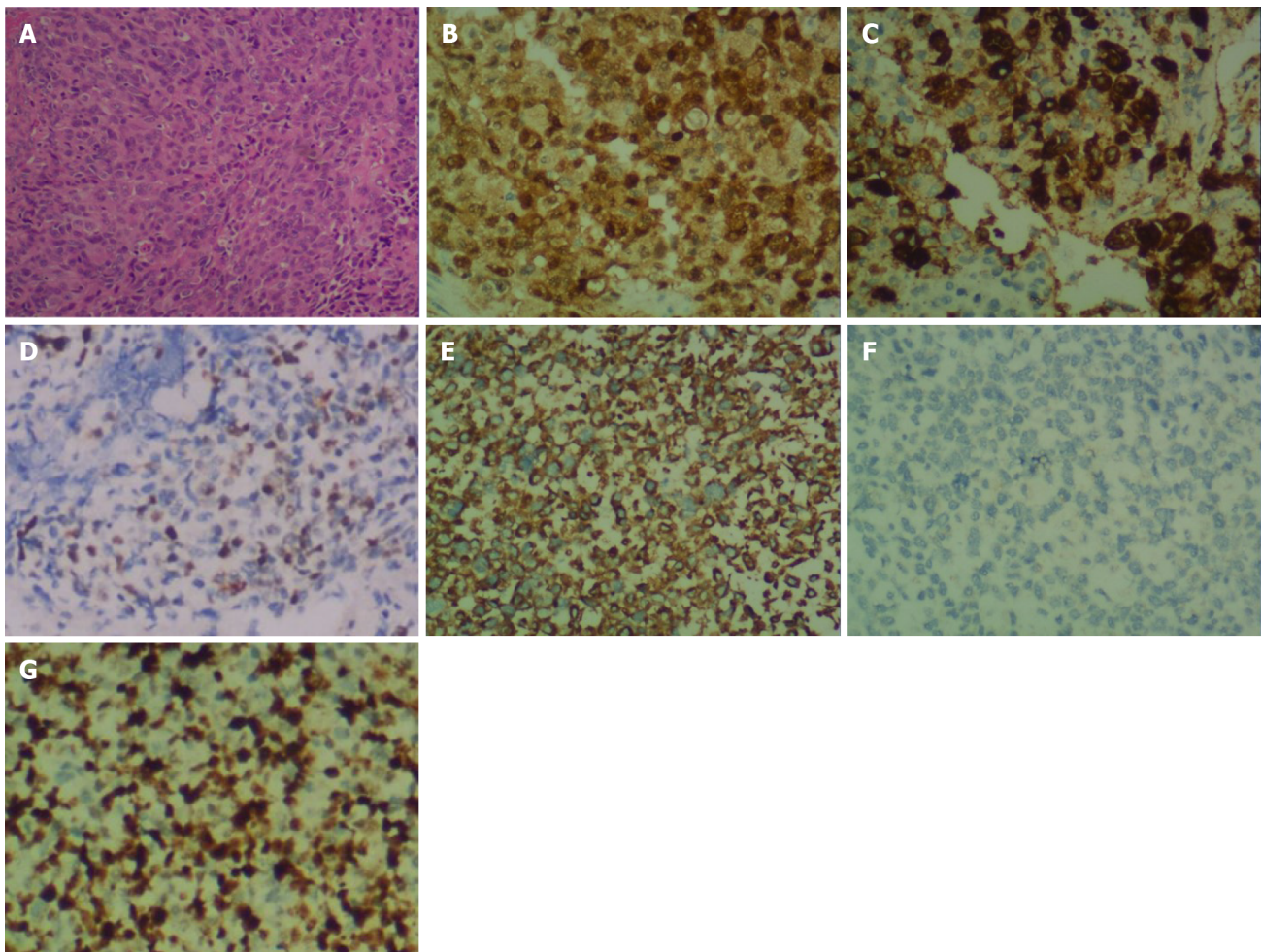


Figure 2 Histological analysis of the tumor tissue. A: Hematoxylin and eosin staining showing the tumor cells ($\times 200$); B-G: Immunohistochemical staining for S-100, homatropine methylbromide-45, p53, vimentin, cytokeratin, and Ki67, respectively ($\times 400$).

DISCUSSION

Our case presents a rare occurrence of primary intramedullary malignant melanoma in the lumbar spine manifested by pathological features and clinical behavior. Primary malignant melanoma of the spinal cord is common in the thoracic region and the cervical region.

Melanoma is an aggressive form of cancer that develops in the cells (melanocytes) that produce melanin and can show up anywhere on the skin^[4]. Less common types may be found in other organs. Primary melanoma in the CNS arises from melanocytes that develop from their precursors. Hayward's classification of primary spinal cord melanoma relies on the absence of a melanoma outside of the spinal cord and histologic confirmation of melanoma^[2]. Primary intramedullary melanoma shows either slow progression or rapid decline, and this lesion is distinct from meningeal melanocytoma and the frequent type of skin melanoma with metastases extending to the CNS.

Notably, surgical criteria are useful when distinguishing primary malignant melanoma from meningeal melanocytoma. At surgery, the adherence to nerve roots by the tumor is suggestive of primary spinal melanoma^[5]. Additional chemotherapy and adjuvant radiotherapy may improve disease-free survival^[3]. Accordingly, it was believed that our case was associated with a primary spinal cord melanoma.

So far, MRI analysis is the best imaging modality for diagnosing spinal cord tumors. Spinal cord melanoma often displays slightly greater signal intensity on the T1-weighted images than the otherwise healthy spinal cord. On the T2-weighted images, however, it can show the same or less signal intensity than the normal cord. The lesion usually shows mild and homogeneous enhancement following the intravenous administration of a gadolinium-based contrast agent. Here, the appearance of the lesion on MRI in our case was consistent with previously reported findings^[6,7]. The final diagnosis should be based on histological and immunophenotyping

examinations. Histopathologic features of malignant melanoma include the formation of tight nests surrounded by well-differentiated melanocytes, which produce pigmented melanin^[8]. Immunohistochemically, S-100 and HMB-45 may contribute to the diagnosis of malignant melanoma^[9].

The various differential diagnoses of primary intramedullary melanoma include metastatic carcinoma, epithelioid schwannoma, and meningeal melanocytoma. In the diagnosis of spinal cord melanoma, the distinction between primary intramedullary melanoma and metastatic melanoma is necessary. As mentioned in the Hayward criteria^[2], histological confirmation and exclusion of melanoma outside the CNS (*e.g.*, the skin, squamous mucosa, and the eyes) are required for the diagnosis of primary intramedullary melanoma. In contrast to melanomas, epithelioid schwannomas exhibit compactly interwoven fascicles of pigmented spindle cells with oval nuclei and low nuclear grade^[10]. Meningeal melanocytoma is a rare pigmented CNS tumor that might be intradural or extradural and was frequently found in the posterior cranial fossa and spinal cord^[11]. Histologically, the tumor cells are arranged in sheets, bundles, nests, and whorls surrounded by a network of reticulin fibers. Mitosis and necrosis are rarely seen in the meningeal melanocytomas.

CONCLUSION

We herein report a case of primary intramedullary melanoma of the lumbar spinal cord, which is a very rare disease, and surgical resection was applied to the patient after careful evaluation.

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