NMC Case Report Journal 2020; 7: 107–111 DOI: 10.2176/nmccrj.cr.2019-0238

Case Report

Intraosseous Melanotic Schwannoma in the Sacrum Mimicking Primary Bone Tumor

Yoshitaka Nagashima,[1] Yusuke Nishimura,[1] Kaoru Eguchi,[1] Takayuki Awaya,[1] Satoshi Yoshikawa,[1] Shoichi Haimoto,[1] Toshihiko Wakabayashi,[1] and Masahito Hara[2]

Primary tumors of sacrum are rarely seen, and the differential diagnosis is extensive, such as chordomas, giant cell tumors, and schwannomas. Sacral intraosseous schwannomas (IOSs) are very rare and encompass approximately 1%–5% of all spinal schwannomas. Melanotic schwannomas (MSs) are categorized as an unusual variant of benign schwannomas; however, they sometimes follow a malignant course. The authors present a case of MS with intraosseous extension into sacrum in a 48-year-old male arising from the left S2 nerve root. Magnetic resonance imaging (MRI) and computed tomography (CT) scan demonstrated a destructive mass in the sacrum. He was made a diagnosis with MS by 18F-fluoro-deoxy-glucose positron-emission-tomography (^{18}F-FDG PET) and open biopsy. The tumor was blackish-colored and vascular-rich fragile tumor covered by fibrous capsule. The floor of the tumor was not encapsulated and invading into the sacral bone. Total removal of the tumor together with the left S2 nerve of origin via posterior approach was achieved. The patient made dramatic recovery of neurological symptoms and tumor recurrence is not seen for 6-month follow-up period. MS is a benign tumor with potential for aggressive behavior and capacity to metastasize. Therefore, total removal of the tumor and careful postoperative follow-up are recommended. Postoperative spinopelvic stability also needs to be taken into consideration. The authors discuss our successful management with a focus on diagnostic process, surgical planning, and histological consideration to provide the most up-to-date guidance on managing this challenging tumor.

Keywords: melanotic schwannoma, intraosseous schwannoma, ^{18}F-FDG PET, open biopsy, spinopelvic stability

Introduction

Primary tumors of sacrum are rare, which account for less than 7% of all intraspinal primary tumors.[1)] Chordoma is the most common, accounting for about 40% of all primary sacral neoplasms followed by giant cell tumor representing 13% of them.[2,3)] Intraosseous schwannomas (IOSs) of sacrum are very rare and encompass approximately 1%–5% of all spinal schwannomas.[4)] Their natural history is generally thought to be benign and slow growing with nonspecific early symptoms.[4)] Melanotic schwannomas (MSs) are rare variants of schwannomas, representing less than 1% of them. MSs arise sporadically or as part of Carney complex in association with other lesions. Although MSs are also generally classified as benign tumor, not a few aggressive MSs with local recurrence or metastasis have been reported[5-7)] and their natural history is completely different from other conventional schwannomas. Therefore, MSs should be managed with careful consideration of surgical planning and postoperative monitoring for recurrence and metastasis. We describe a case of sporadic intraosseous MS with aggressive invasion into the sacrum mimicking primary bone tumor on radiographic imaging, which was successfully resected without complications.

Case Presentation

A 48-year-old man was referred to us presenting with low back pain and left sciatic pain lasting for 6 months. The patient also complained of slight difficulty in urination. He had no muscle weakness of both legs. A lumbar spine magnetic resonance imaging (MRI; Fig. 1) revealed an irregular-shaped and well-defined mass in the spinal canal extending into the sacrum. The lesion was hyperintense on T_1-weighted MRI (T1WI; Figs. 1A, 1D, and 1G), and hypointense on T_2-weighted MRI (T2WI; Figs. 1B, 1E, and 1H). Contrast-enhanced T_1-weighted MRI (T1CE; Figs. 1C, 1F, and 1I) demonstrated remarkable contrast enhancement. A computed tomography (CT) scan (Figs. 2A–2C) revealed irregular bone destruction of the sacrum with a thin sclerotic rim, which prompted us to consider the differential diagnosis includes IOS or primary bone tumor arising from sacrum. 18F-fluoro-deoxy-glucose positron-emission-tomography (^{18}F-FDG PET; Fig. 2D) showed low uptake of FDG with SUV max of 4.21, suggesting denial of highly malignant tumor. We performed an open surgical biopsy under general anesthesia for the histological exploration ahead of surgical procedure, the result of which indicated MS. We proceeded to surgical removal of the tumor via L5 and S1 laminoplasty (Figs. 3A–3D). The tumor consisted of two different components that were the superficial fibrous capsule and blackish vascular-rich fragile component sitting inside (Fig. 4A) the fibrous capsule, which was deemed the main component of

[1]Department of Neurosurgery, Nagoya University, Nagoya, Aichi, Japan
[2]Department of Neurosurgery, Aichi Medical University, Nagakute, Aichi Japan

Received: November 5, 2019; Accepted: November 20, 2019
Online June 26, 2020

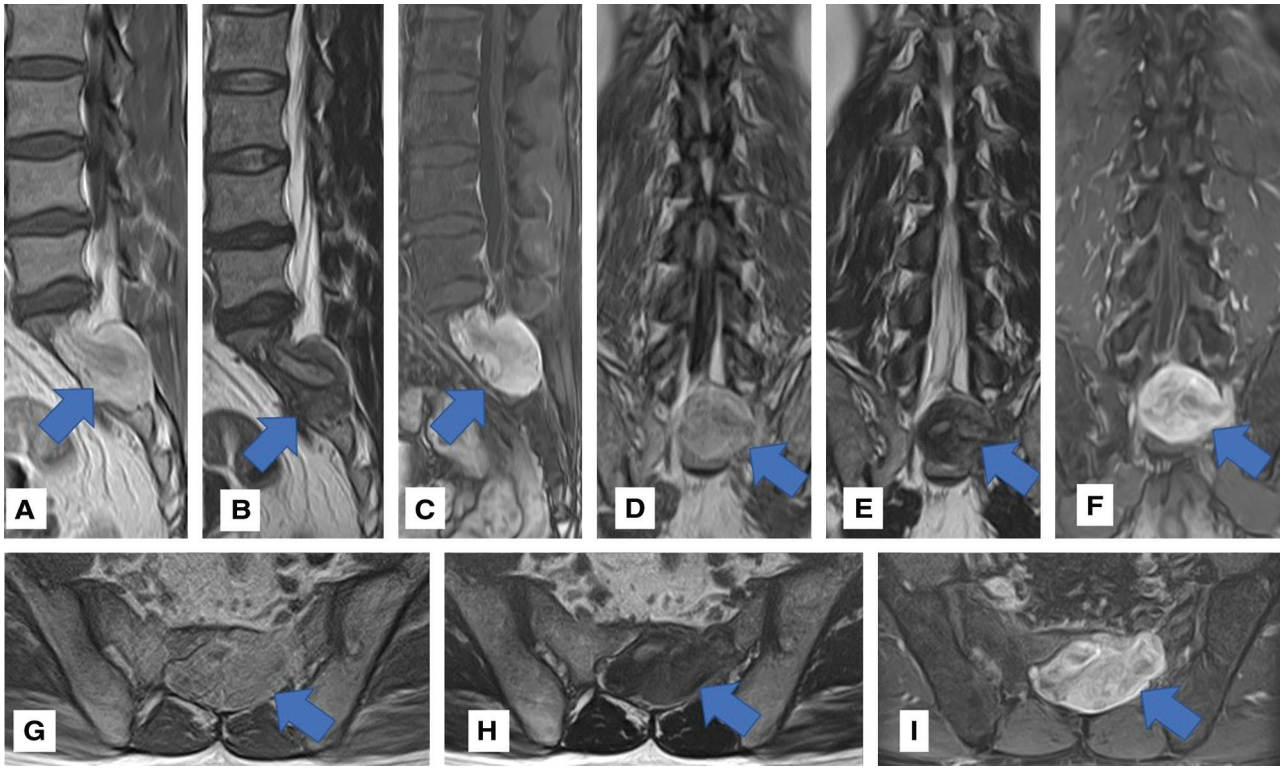

Fig. 1 MRI of lumbosacral spine revealed an irregular-shaped destructive mass occupying spinal canal in the sacrum (arrow). The lesion was hyperintense on T1-weighted MRI (T1WI; **A, D, G**), and hypointense on T2-weighted MRI (T2WI; **B, E, H**), which are characteristic MRI signal intensity for melanotic schwannoma. Contrast-enhanced T1-weighted MRI (T1CE; **C, F, I**) demonstrated remarkable contrast enhancement. MRI: magnetic resonance imaging, T1WI: T1-weighted image, T2WI: T2-weighted image.

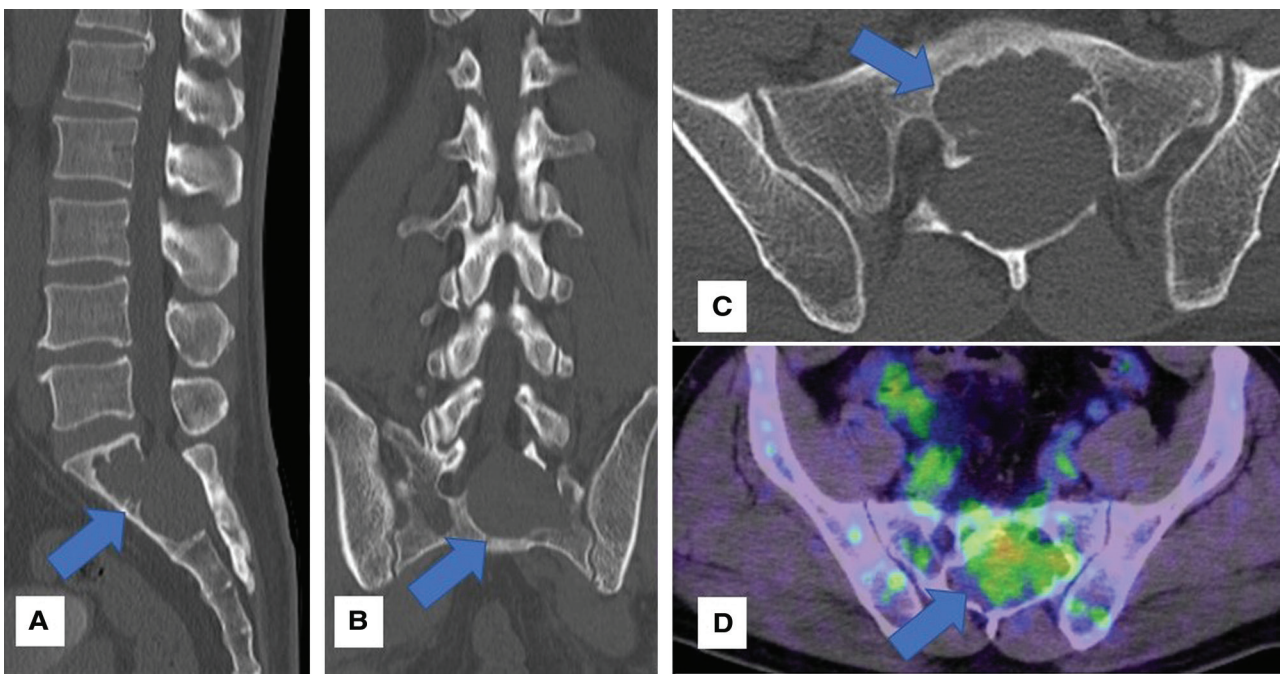

Fig. 2 CT scan revealed irregular bone destruction of the sacrum with a thin sclerotic rim, suggestive of benign nature (A–C, arrow). 18F-FDG PET showed low uptake of FDG corresponding to the CT findings (D, arrow). ^{18}F-FDG PET: 18F-fluoro-deoxy-glucose positron-emission-tomography, CT: computed tomography.

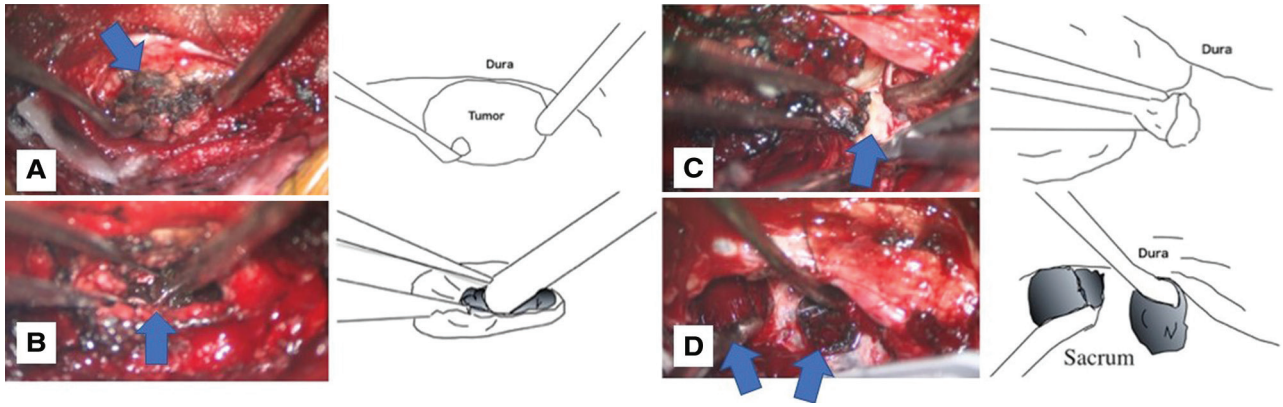

Fig. 3 Intraoperative photographs. The fibrous capsule covering the surface of the tumor. It seemed like a typical schwannoma (**A**, arrow). The blackish vascular-rich fragile component appeared inside (**B** arrow). The tumor arising from the left S2 nerve root (**C**, arrow). After the main part of the tumor was removed, blackish vascular-rich fragile component was remained in the sacrum which is honeycombed with holes (**D**, arrow).

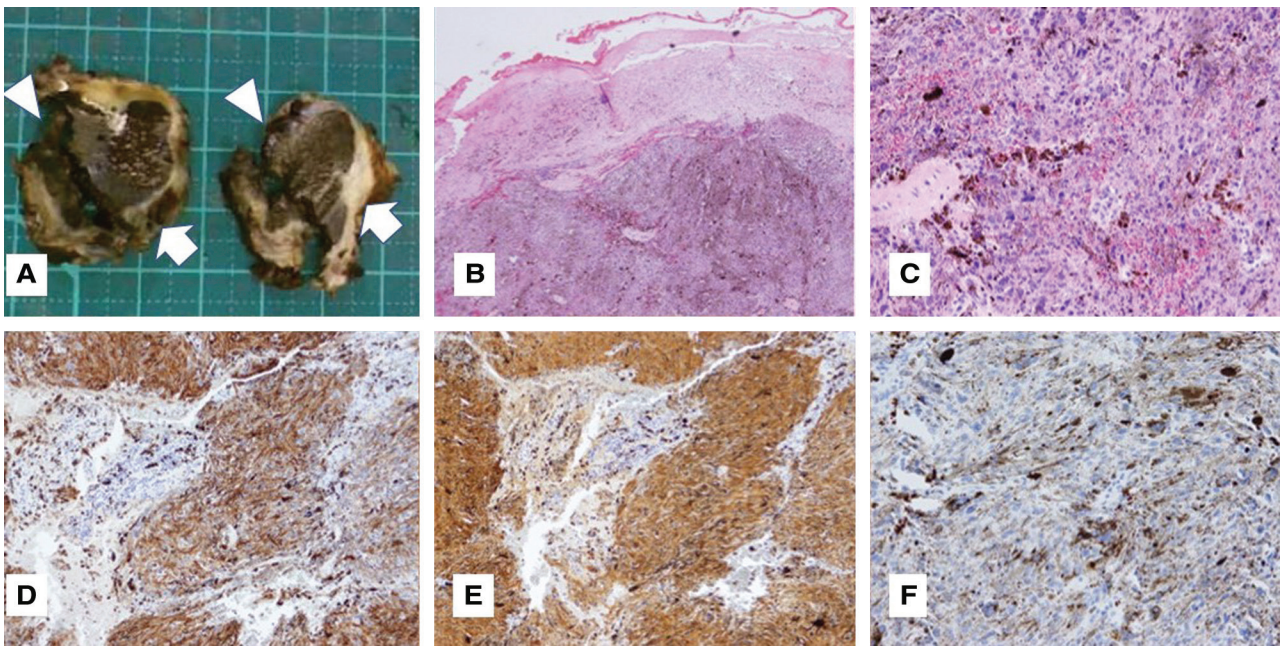

Fig. 4 The cross section of specimen shows the blackish component was covered by fibrous capsule and blackish component penetrated the capsule at the floor (**A** arrowhead). Histopathological examination showed compact fascicles of spindle cells and psammoma bodies (**B** and **C**). HMB-45 (**D**), S-100 (**E**) was positive. Staining of p-53 (F) was very low.

the tumor. The floor of the tumor was not encapsulated (Fig. 4A arrowhead), and the blackish component was directly invading into the sacrum. The tumor was considered originating from the left S2 nerve root (Fig. 3C) and the nerve was completely embedded into the tumor. Therefore, we sacrificed the S2 nerve root of origin for complete removal following direct S2 nerve stimulation, which confirmed no response from leg muscles and anal sphincter muscles on intraoperative motor evoked potentials. After careful circumferential tumor dissection from surrounding nerve tissues and dural sac, en-bloc removal of the tumor was performed together with the left S2 nerve root (Fig. 4A). However, small part of the blackish component invading into the sacrum was left inside the bone and was additionally and completely resected (Fig. 3D). Histopathological examination showed compact fascicles of spindle cells containing dark brown intracellular pigment positive for HMB-45, S-100 and MART-1, with suggestive of MS. The patient made dramatic recovery of neurological symptoms postoperatively. There was no recurrence on follow-up imaging at 6th month postoperative period. Carney complex was denied as a result of further examinations.

Discussion

While schwannomas are the most frequent intradural and extramedullary spinal tumor, sacral IOSs are rare.[8)]

Differential diagnosis of sacral primary tumors includes sacral chordomas and giant cell tumors other than IOSs. Compared with destructive nature of these primary bone tumors, IOSs display no bone residues and permitted themselves be shaped by remodeling around the lesion to form a sclerosis boarder on CT scan as described in the present case.[9-11] MRI has a better contrast than CT for the study of soft tissues and provides a better display in multiplanar views. IOSs have no involvement of surrounding muscles and sacroiliac joints on MRI,[10] which is distinct imaging characteristics separated from chordomas or giant cell tumors.

MS is a rare variant of schwannoma comprising melanin producing schwann cells,[12] which was originally described by Millar in 1932 as a "malignant melanotic tumor of sympathetic ganglion cells."[12] Hodson in 1961 suggested that it was a form of schwannoma.[13] Histogenesis of melanin in schwann cell rests in the fact that the neural crest cells migrate and differentiate into divergent tissues such as melanocytes, schwann cells, neurons of peripheral nervous system, adrenal medulla, and calcitonin-producing c-cells of thyroid. MRI is the investigation of choice to differentiate MS from other conventional schwannomas. Typically, the lesions are hyperintense on T1WI and hypointense on T2WI due to paramagnetic free radicals in melanin while other conventional IOSs tend to be hypointense on T1WI and hyperintense on T2WI.[14] Both of MSs and conventional schwannomas enhance on contrast.[14,15] In contrast to the typical encapsulation of IOSs, MSs are circumscribed but unencapsulated tumors. MSs are enveloped by a thin fibrous membrane that may be infiltrated by tumor infiltration. Often lobulated, these are soft, firm or rubbery with black, brown, or gray cut section; sometimes with areas of hemorrhage or necrosis,[16] which may reflect the potential of more aggressive nature and an invasive growth pattern.[6] Considering the aggressiveness, it is difficult to distinguish MSs from other primary bone tumors preoperatively only on conventional imaging.

18F-fluoro-deoxy-glucose positron-emission-tomography (^{18}F-FDG PET) has been advocated for preoperative assessment of biological activity, malignant capacity, and histological characteristics. ^{18}F-FDG PET has offered the possibility of non-invasive estimation of pathological diagnosis and it also may help the prediction of patient prognosis.[17] Eary et.al. analyzed ^{18}F-FDG PET of 238 patients with diagnosis of sarcoma and reported the median ^{18}F-FDG SUVmax was 6.2, with a mean of 8.2.[18] They concluded SUVmax has been applied widely to predict the prognosis and the presence of spatial heterogeneity of SUVmax could suggest worse prognosis more accurately. In the present case, ^{18}F-FDG PET demonstrated low uptake with a value of 4.21 with little heterogeneity, indicating benign tumor.

The biopsy was successfully performed in the present case. Although the procedure has a potential risk of complications such as infection, hemorrhage, site-related problems, and wound closure issues, it was significantly helpful for the predetermination of the nature of the tumor in consideration of MS due to its atypical imaging findings.[8,19] As we all know, misdiagnosis of the disease will gradually promote tumor growth, making the operating procedure more difficult.

Klimo et al. proposed a classification of sacral nerve sheath tumors based on their location into three types (Types I–III).[20] Type I is confined to the sacrum like the present case and could be resected via a posterior approach alone. Type II locally metastasizes through the anterior and posterior sacral walls into the presacral and subcutaneous spaces, usually requiring combined anterior–posterior surgery. Type III is located primarily in the presacral/retroperitoneal area, which should be treated via an anterior approach. In cases of conventional IOSs, several authors reportedly prefer the piecemeal subtotal excision with sacral nerve roots of origin preserved as much as possible, which could achieve a good outcome without local recurrence and transformation to malignancy.[19] However, all available literatures of MSs recommend complete surgical excision. En-bloc resection may provide excellent local control in this locally aggressive tumor[21] and we attempted to achieve En-bloc resection via posterior approach while small part of the tumor inside the sacrum needed to be additionally resected. Spinopelvic reconstruction is considered on the criteria proposed by Gunterberg et al. such as (1) total sacrectomy in which the whole sacroiliac joint is removed and (2) partial sacrectomy involving more than 50% of sacroiliac joint each side.[21] In the present case, spinopelvic stabilizing factors including sacral alar, L5/S1 facet joint and disc space, and lumbopelvic ligaments[21] were preserved and only a partial laminectomy of eroded lamina was performed, we determined the present case would maintain adequate stability. Periodic follow-up is needed because of high risk of local recurrence, malignant transformation, and metastasis in cases of MSs unlike conventional IOSs.[5,7,22] Zhang et al. demonstrated that the chance of recurrence following resection is 18.2% and metastasis in 9.1%.[15]

Histological investigation shows the cellular lesion composed of spindle-shaped and epithelioid cells arranged in lobules or fascicles, cells containing melanin pigments. Melanosomes in all stages of maturation are found within the cytoplasm of these tumor cells.[12] Immunohistochemistry shows that MSs are immunoreactive for vimentin, S-100, and HMB-45. Melan-A, melanoma cell adhesion molecule and microphthalmic transcription factor reactivity, is also seen.[12,23] Pathologically, differential diagnosis includes meningeal melanocytoma and metastatic melanoma. MSs and melanocytomas may be found to represent a lesion continuum.[12] Of great clinical importance is differentiating MS from metastatic melanoma. Metastatic melanoma is rarely totally black and rarely located at paraspinal site and obviously cytologically malignant with lack of fat.[12] Dendritic appearance of cells in MS is seldom seen in metastatic melanoma.[16]

MS is a benign tumor with potential for aggressive behavior and capacity to metastasize. Therefore, total removal of the tumor and careful postoperative follow-up are recommended. Klimo's classification is helpful to determine the optimal surgical approach and spinopelvic stability also

needs to be taken into consideration. More studies are necessary to understand the natural history, prognosis, and best management strategies for MS.

Conflicts of Interest Disclosure

The authors have nothing to disclose.

References

1) Meister M, Choyke P, Anderson C, Patel U: Radiological evaluation, management, and surveillance of renal masses in Von Hippel-Lindau disease. *Clin Radiol* 64: 589–600, 2009
2) Unni KK: Dahlin's bone tumors: general aspects and data on 11087 cases. 5th ed. Philadelphia: Lippincott-Raven, 1996
3) Turcotte RE, Sim FH, Unni KK: Giant cell tumor of the sacrum. *Clin Orthop Relat Res* 291: 215–221, 1993
4) Khan UA, Ismayl G, Malik I: Giant Sacral schwannoma treated with a 360 approach: a rare case and systematic review of the literature. *World Neurosurg* 115: 65–72, 2018
5) Chandran RS, Patil AK, Prabhakar RB, Balachandran K: Melanotic schwannoma of spine: illustration of two cases with diverse clinical presentation and outcome. *Asian J Neurosurg* 13: 881–884, 2018
6) Choi SE, Cha YJ, Kim J, et al.: A rare case of aggressive melanotic schwannoma occurred in spinal nerve of a 59-year-old male. *J Pathol Transl Med* 51: 505–508, 2017
7) Faria MH, Dória-Netto RH, Osugue GJ, Queiroz Lde S, Chaddad-Neto FE: Melanotic schwannoma of the cervical spine progressing with pulmonary metastasis: case report. *Neurol Med Chir (Tokyo)* 53: 712–716, 2013
8) Wang YQ, Hu JX, Yang SM, et al.: Intraosseous schwannoma of the mobile spine: a report of twenty cases. *Eur Spine J* 27: 3092–3104, 2018
9) Çağlı S, Işık HS, Yıldırım U, Akıntürk N, Zileli M: Giant sacral schwannomas. *J Neurooncol* 110: 105–110, 2012
10) Si MJ, Wang CS, Ding XY, et al.: Differentiation of primary chordoma, giant cell tumor and schwannoma of the sacrum by CT and MRI. *Eur J Radiol* 82: 2309–2315, 2013
11) Turk PS, Peters N, Libbey NP, Wanebo HJ: Diagnosis and management of giant intrasacral schwannoma. *Cancer* 70: 2650–2657, 1992
12) Antonescu CR, Scheithauer BW, Woodruff JM: AFIP atlas of tumor pathology series 4. Tumors of the peripheral nervous system:, Silver spring, Maryland, 2013
13) Hodson JJ: An intra-osseous tumour combination of biological importance-invasion of a melanotic schwannoma by an adamantinoma. *J Pathol Bacteriol* 82: 257–266, 1961
14) Hoover JM, Bledsoe JM, Giannini C, Krauss WE: Intramedullary melanotic schwannoma. *Rare Tumors* 4: e3, 2012
15) Zhang HY, Yang GH, Chen HJ, et al.: Clinicopathological, immunohistochemical, and ultrastructural study of 13 cases of melanotic schwannoma. *Chin Med J (Engl)* 118: 1451–1461, 2005
16) Carney JA: Psammomatous melanotic schwannoma. A distinctive, heritable tumor with special associations, including cardiac myxoma and the Cushing syndrome. *Am J Surg Pathol* 14: 206–222, 1990
17) Matsumoto Y, Baba S, Endo M, et al.: Metabolic tumor volume by ^{18}F-FDG PET/CT can predict the clinical outcome of primary malignant spine/spinal tumors. *Biomed Res Int* 2017: 8132676, 2017
18) Eary JF, O'Sullivan F, O'Sullivan J, Conrad EU: Spatial heterogeneity in sarcoma 18F-FDG uptake as a predictor of patient outcome. *J Nucl Med* 49: 1973–1979, 2008
19) Pan W, Wang Z, Lin N, et al.: Clinical features and surgical treatment of sacral schwannomas. *Oncotarget* 8: 38061–38068, 2017
20) Klimo P Jr, Rao G, Schmidt RH, Schmidt MH: Nerve sheath tumors involving the sacrum. *Case report and classification scheme. Neurosurg Focus* 15: E12, 2003
21) Zhang HY, Thongtrangan I, Balabhadra RS, Murovic JA, Kim DH: Surgical techniques for total sacrectomy and spinopelvic reconstruction. *Neurosurg Focus* 15: E5, 2003
22) Li B, Chen Q: Melanotic schwannoma of thoracic spinal root mimics metastatic melanoma: a potential pitfall for misdiagnosis. *Int J Clin Exp Pathol* 8: 8639–8641, 2015
23) Brat DJ, Giannini C, Scheithauer BW, Burger PC: Primary melanocytic neoplasms of the central nervous systems. *Am J Surg Pathol* 23: 745–754, 1999

Corresponding author:
Yusuke Nishimura, MD, PhD, Department of Neurosurgery, Nagoya University School of Medicine, 65 Tsurumai-cho, Showa-ku, Nagoya, Aichi 466-8550, Japan.
✉yusuken0411@med.nagoya-u.ac.jp