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CASE REPORT

Intradural cervical chordoma with diffuse spinal leptomeningeal spread: case report and review of the literature

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Abstract

Purpose Chordoma is a low-grade malignant bone tumor derived from embryonic notochord remnants along the axial skeleton. About 50% of chordomas occur in the sacral vertebrae and 35% in the skull base. Most chordomas are extradural and cause extensive bone destruction. Intradural spinal tumors without bone involvement are rare.

Methods We herein describe the clinical features of a patient with a chordoma as well as the imaging and pathological manifestations of the tumor.

Results We encountered an unusual presentation of a C6 and C7 spinal intradural chordoma in a 23-year-old man. He presented with a 5-day history of discomfort over the lumbosacral region. Magnetic resonance imaging and enhanced scanning of the cervical spine showed an intradural soft tissue mass at C6 and C7 and linear enhancement of the spinal meninges. The tumor was excised because the patient had been previously misdiagnosed with an intraspinal neurogenic tumor with spinal meningitis. Postoperative pathological examination confirmed the diagnosis of chordoma. On postoperative day 7, the patient underwent brain magnetic resonance imaging because of severe headache. The images showed multiple soft tissue nodules in the skull base cistern. To the best of our knowledge, this is the first case report of an entirely extraosseous and spinal intradural chordoma with diffuse spinal leptomeningeal spread. The patient died 2 months postoperatively.

Conclusions An intradural spinal chordoma is difficult to distinguish from a neurogenic tumor by imaging. When the lesion is dumbbell-shaped, it is easily misdiagnosed as a schwannoma. In the present case, the tumor was intradural and located at the level of the C6 and C7 vertebrae. Preoperative diagnosis was difficult, and the final diagnosis required pathological examination.

Keywords Chordoma · Notochord · Intradural · Spinal leptomeningeal spread · Magnetic resonance imaging

Introduction

Chordomas are rare, slow-growing primary malignant tumors arising from the embryonic cells of the primitive notochord. They are most commonly seen in the clivus and sacrococcygeal regions. Chordomas account for 1–4% of all primary malignant bone tumors and are usually diagnosed after the age of 30 years, most commonly between 60 and 70 years [1]. Intradural chordomas without bone involvement are uncommon and may occur intracranially or in the spinal canal. Spinal intradural chordomas are extremely rare. We herein present what we believe to be the first reported case of a spinal intradural chordoma with diffuse spinal leptomeningeal spread. We also provide a review of the relevant literature.

Case report

A 23-year-old man was admitted to our hospital with a 5-day history of discomfort over the lumbosacral region. He also had weakness of the upper and lower extremities, especially the lower extremities; urinary retention; difficulty raising his limbs; and an inability to stand and walk. He had no other significant medical history. Physical examination revealed decreased muscle strength in the upper and lower extremities. Muscle strength was grade 4 in both upper extremities, grade 3 in the left lower extremity, and grade 1 in the right

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lower extremity. Hyperalgesia was present below C3–C4 of the cervical spinal cord.

Conventional magnetic resonance imaging (MRI) showed a spinal intradural mass located at the C6 and C7 vertebrae. The lesion appeared isointense on T1-weighted imaging (T1WI) and T2WI. The mass measured 13.8 × 8.8 × 28.5 mm (Fig. 1a). The spinal cord was significantly compressed. After MRI enhancement, the mass showed inhomogeneous enhancement. Our preliminary diagnosis was a neurogenic tumor. The soft meninges in the base of the skull and soft spinal meninges exhibited multiple nodular enhancements (Fig. 1b), which were misdiagnosed as inflammatory lesions. The intradural tumor was completely resected.

The patient underwent endotracheal intubation. He was placed in the left decubitus position. After successful induction of general anesthesia, the patient's head was placed in 15° of anteflexion and fixed with a Mayfield holder. A straight midline incision of about 10 cm in length was marked from C5 to C8. The skin, subcutaneous tissue, and muscular layer were incised with an electric knife along the midline. During the operation, we exposed the spinous processes of the C5–C7 vertebrae and the laminae of the C6 and C7 vertebrae. The laminae and spinous processes of C6 and C7 were removed with a rongeur and electric grinder, respectively. The tension of the spinal meninges was high and showed no fluctuation. The dura was opened under microscopic magnification, and the basivertebral veins were found to be compressed. The tumor tissue was predominantly soft but mixed with areas of toughness. The lesion was located on the right side of the spinal cord. The mass had a rich blood supply and was accompanied by a solid, chronic clot. The tumor was resected under a microscope and was closely adhered to the spinal nerve root sheath. Outflow of cerebrospinal fluid occurred upon tumor removal. After tumor resection, the fluctuation of the cerebrospinal fluid was resolved and venous filling was restored. After sufficient hemostasis, an approximately 5- × 5-cm section of artificial dura was applied. A drain was left in the incision. After hemostasis, the muscle layer, subcutaneous tissue, and skin were sutured closed. The drain was connected to a mild negative-pressure drainage device, and dressing was placed over the incision. The specimen was sent for histopathological examination.

Microscopically, the tumor cells were round and uniform in size. The nuclei were round and slightly biased, and the stroma was mainly myxoid. Immunohistochemical examination revealed positivity for CD99 and epithelial membrane antigen (EMA), with scattered positivity for vimentin and S100; negativity for cytokeratin, myogenin, myogenic differentiation 1, and desmin; and a 30% Ki-67-positivity rate. The pathological examination findings were consistent with a chordoma (Fig. 2). MRI showed that the tumor was located at the level of the C6–C7 subdural tissue. Therefore, the tumor was classified as a cervical spinal intradural chordoma.

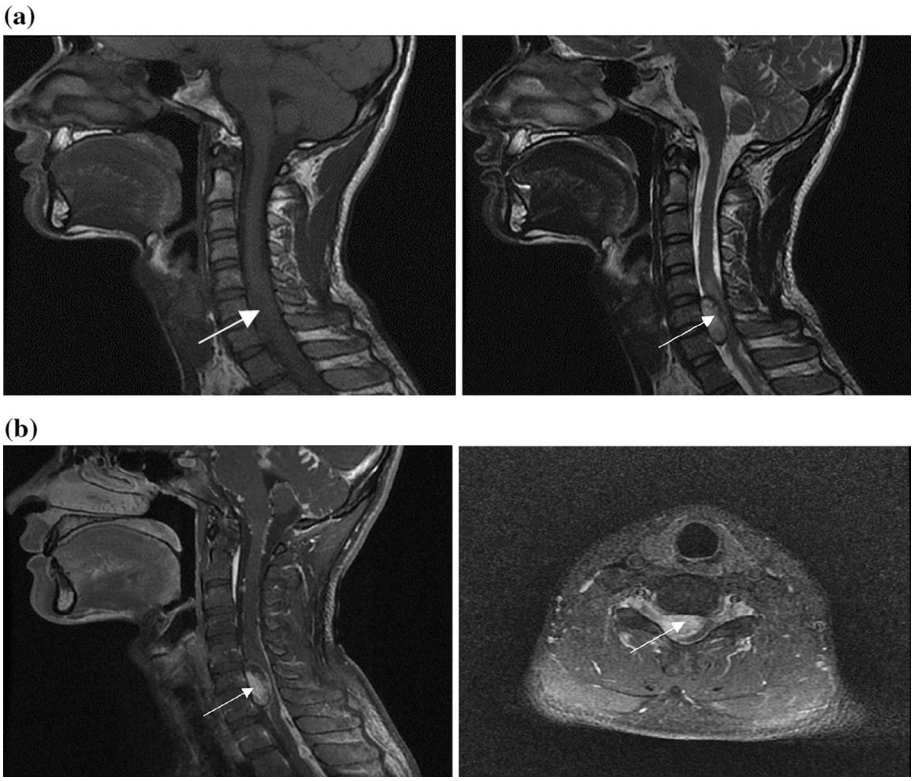

Fig. 1 **a** Conventional magnetic resonance imaging (MRI) showed a spinal intradural mass located at the C6 and C7 vertebrae. **b** The lesion appeared isointense on T1-weighted imaging (T1WI) and T2WI. The soft meninges in the base of the skull and soft spinal meninges exhibited multiple nodular enhancements

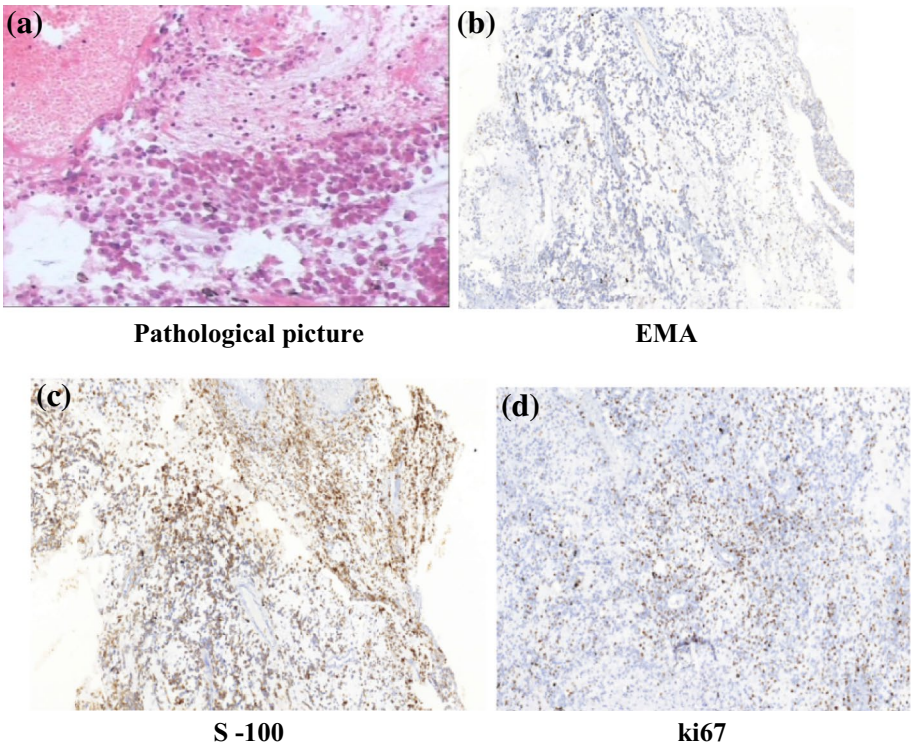

Fig. 2 Microscopically, the tumor cells were round and uniform in size. The nuclei were round and slightly biased, and the stroma was mainly myxoid (**a**). Immunohistochemical examination revealed positivity for epithelial membrane antigen (EMA) (**b**), with scattered positivity for S100 (**c**); and a 30% Ki-67-positivity rate (**d**)

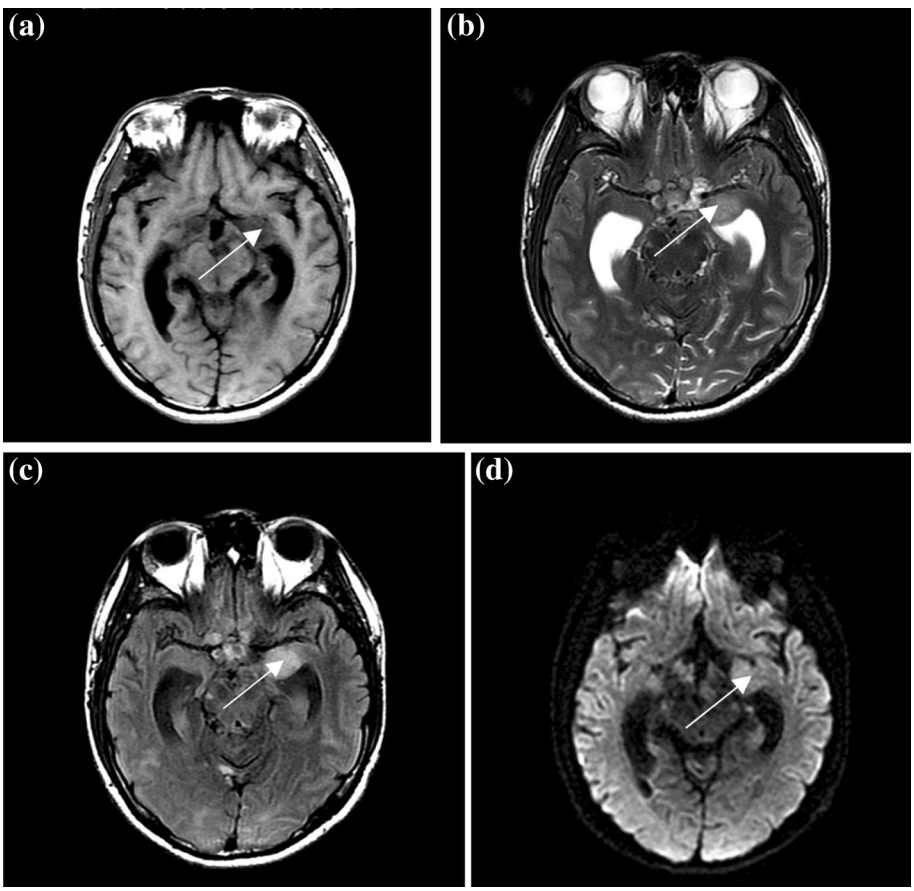

Fig. 3 Multiple soft tissue nodules were noted in the skull base cistern, and the lesions were hypointense on T1WI (**a**) and hyperintense on T2WI (**b**). Fluid-attenuated inversion recovery showed high signal intensity (**c**), and diffusion-weighted imaging showed slightly high signal intensity (**d**)

The patient developed a headache on postoperative day 7 and underwent MRI. Multiple soft tissue nodules were noted in the skull base cistern, and the lesions were hypointense on T1WI and hyperintense on T2WI. Fluid-attenuated inversion recovery showed high signal intensity, and diffusion-weighted imaging showed slightly high signal intensity (Fig. 3). The pathological diagnosis and imaging findings were consistent with an intradural chordoma with spinal leptomeningeal dissemination. The patient showed progressive deterioration and died 2 months after the operation.

Discussion

Chordomas are rare, locally aggressive primary bone tumors of notochordal origin, and they almost exclusively involve the axial skeleton. Nearly 90% of cases occur in the sacrococcygeal region and base of the skull. The remaining cases develop in the mobile spine, predominantly in the cervical and lumbar vertebrae (approximately 10 and 4% of cases, respectively). Chordomas uncommonly develop in the thoracic vertebrae.

The notochord is composed of a column of cells ventral to the neural tube and arises in week 3 of embryonic life [2, 3], disappearing by week 7. The general consensus is that chordomas arise from these persistent notochordal remnants. These remnants exhibit intraosseous localization; therefore, chordomas are usually extradural and cause local bone destruction.

An intradural chordoma may be associated with ectopic localization of residual notochord tissue along the axial skeleton from the coccyx to the dorsum sella during embryonic development; however, its pathogenesis remains unclear [4]. Luschka [5] first described his finding of pathological ectopic notochordal tissue at the posterior clivus in 1856. In another study, ectopic notochord tissue was found in an intradural location anterior to the pons in 2% of all autopsies [6]. Warnick et al. [7] speculated that the tumor may arise from a primitive notochordal remnant during embryogenesis or from the ectopic position of notochordal tissues following an early traumatic injury.

An intradural chordoma may occur in the tentorium cerebelli, pontocerebellar region, foramen magnum, or filum terminale. Occurrence in the intradural cervical, thoracic, lumbar, and epidural lumbar regions have been reported to date [2, 8, 9]. Primary spinal intradural chordomas are extremely rare. An intradural chordoma without bone involvement is primarily seen in middle-aged

Table 1 Reported cases of spinal intradural chordomas without bone involvement

No.	Age (yr)	Sex	Duration	Symptom and sign	Treatment	Recurrence/follow-up	Author and year
1	33	F	3 mo	Shoulders pain, gait disturbance Spastic palsies, T6 sensory level	Resection	No/1 yr	Ramiro et al. [11]
2	37	F	9 yr	Paraparesis, abnormal tendon reflexes	Subresection	No/18 mo	Vaz et al. [14]
3	65	M	1 mo	Lower extremities weakness, spasticity Tendon reflexes increased, T4 sensory level	Resection	No/18 mo	Gelabert-Gonzalezet al. 1999 [15]
4	69	M	4 mo	Pain radiating into the left hand and involving second to fifth fingers	Resection	NA	Gunnarsson et al. 2001 [16]
5	38	F	3 mo	Bilateral buttock pain	Resection	No/6 mo	Sumi et al. 2001 [17]
6	50	F	NA	Back pain, headache, sensomotor paresis of the right leg	Subresection	No/18 mo	Steenberghs et al. [12]
7	28	F	2 yr	Low-back pain radiated over the buttocks, thighs, and calves bilaterally	Resection	No/5 mo	Bayar et al. 2002 [18]
8	36	M	1 mo	Sensomotoric paraparesis up to L1 Saddle anesthesia	Subresection	Dead/3 wk	Badwal et al. 2006 [13]
9	38	M	1 yr	Left neck and shoulder pain	Resection	No/7 yr	Bergmann et al. 2010 [19]
	44	F	Several mo	Neck and right side pain	Resection	No/1 yr	
10	63	F	2 wk	Left upper limb weakness, grade 4/5 both upper extremities, hyperreflexia of lower extremities	Resection	No/20 mo	Kawanabe et al. 2014 [20][a]
11	23	M	5 d	Discomfort over the lumbosacral	Resection	Dead/2 mo	Current case

yr year, *mo* month, *wk* week, *d* day

[a]Simultaneous discovery of cranial and spinal intradural chordomas

Table 2 Imaging characteristics of spinal intradural chordomas

No.	Location	T1WI	T2WI	Enhancement	Disseminate
1	T6	No	No	No	No
2	C5-T2, ventral	Hypointense	NA	Homogeneous	No
3	C6–C7, ventral	Isointense	Isointense	Homogeneous	No
4	C1–C2, right intradural and neuroforamen	NA	NA	Homogeneous	No
5	S1	Isointense	Hyperintense	No	No
6	Multiple, Cervical to sacral	NA	NA	NA	Spinal cord, paraspinal muscles
7	L5-S1	Hypointense	Hyperintense	No	No
8	Multiple, C2, C7, T12-L1	NA	NA	Homogeneous	Spinal cord
9	C2–C3, left intradural and neuroforamen	NA	Hyperintense	No	No
	C2–C3, right intradural and neuroforamen	NA	NA	Isointense	No
10	C1–C2, posterior	Isointense	Hyperintense	Homogeneous	No
11	C6–C7, ventral	Isointense	Hyperintense	Heterogeneous	Spinal leptomeningeal

and elderly patients [10]. Forty-five cases of intracranial and spinal intradural chordomas have been reported in the English language literature. In 1986, Ramiro et al. [11] diagnosed the first spinal intradural chordoma by myelography; the tumor was located at the T6 level. To the best of our knowledge, 12 patients with spinal intradural chordomas have been reported in the English language literature (Table 1). A multicentric spinal intradural chordoma was reported by Steenberghs et al. [12] and Badwal et al. [13] in 2002 and 2006, respectively.

Rare cases of extraosseous intradural chordomas have been reported [21, 22]. The absence of bone involvement and a different position relative to the dura mater are not the only characteristics that distinguish intradural chordomas from typical chordomas. Katayama et al. [23] and Vaz et al. [14] reported that spinal intradural chordomas have a slower growth pattern and clear boundaries that contribute to their complete removal and that they never metastasize. En bloc resection was performed in 8 of the 12 reported cases of spinal intradural chordomas. The patients were followed up for 5 months to 7 years without recurrence or metastasis. In one case, the capsule adhered to the spinal cord, which made total resection difficult, and partial resection was instead performed. The patient was followed up for 15 months, and no tumor recurrence or metastasis was found. Two patients with multiple spinal intradural chordomas underwent local resection. One patient was followed up for 18 months and was still alive at the time of this writing. In the other patient, the lesions extended into the paraspinal muscles, and metastatic lesions developed in the cervical cord; the patient died 3 weeks postoperatively [12]. The primary tumor was completely removed in the present case; however, spinal leptomeningeal spread occurred, and the patient died 2 months after surgery.

Chordoma metastases are usually found at advanced stages of the disease. The metastases occur generally in lung, bones, liver and lymph nodes [3]. The patient in the present report showed diffuse spinal leptomeningeal spread upon initial admission. Steenberghs et al. [12] and Badwal et al [13]. reported a case of spinal and meningeal metastasis in 2002 and 2006, respectively. The findings of these studies indicate that spinal intradural chordomas are prone to metastasis despite the absence of bone destruction.

In the present case, all histological sections examined showed tumor proliferation comprising cells with vacuolated cytoplasm and areas resembling cartilage. Tumor cells were arranged in nests, cords, or sheets within a myxoid stroma. Cytokeratin immunohistochemistry was strongly positive and focal but definite positivity was present for both S-100 and EMA. The Ki-67 index was low (< 0.5%), with only a few positive nuclei. Positive staining for brachyury, galectin-3, and Ki-67 would be helpful to establish a differential diagnosis.

The imaging characteristics of spinal intradural chordomas are listed in Table 2. The maximal tumor diameter varies from 1.5 to 6.0 cm [24]. MRI contributes to discrimination between the tumor and surrounding structures. In some cases, an intradural retroclival chordoma is difficult to distinguish from an epidermoid cyst in the prepontine cistern and a petroclival meningioma by imaging features alone, and the correct differential diagnosis depends on a combination of imaging and clinical features [25]. In the present case, the tumor appeared isointense on T1WI and hyperintense on T2WI. The lesion showed a heterogeneous signal with significant contrast enhancement.

Rare cases of extraosseous intradural chordomas have been reported. These chordomas are generally considered to have a more favorable prognosis than typical chordomas [21, 22]. Surgery is the most effective first-line treatment for patients with chordomas. Complete resection is feasible for a single spinal intradural chordoma that is not adhered to surrounding structures. Among the 12 reported cases of

spinal intradural chordomas, 2 involved multiple tumors with metastasis to the spinal cord and paraspinal muscles. In the present case, the cervical spinal intradural chordoma presented with diffuse spinal leptomeningeal spread. Accordingly, spinal seeding might emerge as a potential complication during long-term follow-up. Close observation and timely screening for spinal seeding are required.

Conclusion

In conclusion, spinal intradural chordomas are difficult to distinguish from neurogenic tumors. These chordomas are difficult to diagnose preoperatively and must be confirmed histopathologically. Surgery is the first-choice treatment for this disease. The occurrence of cerebral dural spread or spinal cord metastasis of an intradural spinal chordoma has a poor prognosis.

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Compliance with ethical standards

Conflict of interest None of the authors has any potential conflict of interest.

Informed consent The patient and/or his family were informed that data from the case would be submitted for publication and gave their consent.

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