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Case Report

O-arm Navigation-Guided Surgical Resection and Posterior Fixation for a Large Sacral Schwannoma

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Sacral schwannoma is a rare tumor with relatively few symptoms; it thus tends to be large at diagnosis and is challenging to treat surgically. We present the case of a 12-year-old girl with a large sacral schwannoma that was successfully surgically resected using O-arm navigation in a two-stage operation. First, we performed tumor resection from the posterior aspect with assisted O-arm navigation. One week later, resection from the anterior aspect was conducted with posterior spinopelvic fixation and fibula graft. We performed partial resection of the tumor from the anterior and posterior aspects as much as possible. O-arm navigation contributed to precise and safe tumor resection and implant insertion.

Key words: sacral schwannoma, cellular schwannoma, spinal tumor, intradural extramedullary tumor, O-arm navigation

Primary sacral tumors are relatively rare. The most common primary malignant sacral tumor is chordoma, followed by chondrosarcoma; both are low-grade malignant tumors [1]. Of the benign sacral tumors arising from the neural component, schwannomas are the most common [2,3]. Sacral schwannomas represent less than 1-5% of all spinal schwannomas, accounting for 25% of primary spinal tumors [4]. Schwannoma is more common in women between the ages of 20 and 50 years [3]. Most patients with sacral schwannomas have few signs or symptoms, as tumors are slow-growing and rarely painful. Most schwannomas are thickly encapsulated neoplasms amenable to total excision. However, due to the complex anatomy around the sacrum, surgical excision of a sacral schwannoma is challenging. We report a case of a large sacral schwannoma that was treated by staged surgery using O-arm navigation.

Case Report

A 12-year-old girl had complained of back pain for a month, along with urinary incontinence. Two years previously, she had flexion-contracture of the knees and leg pain. These symptoms were diagnosed as cerebral palsy. One year prior to our involvement, myotomy and tendon elongation were performed on both legs at another hospital. After surgery, she was able to walk independently. Shortly thereafter, however, she developed low back pain and spinal deformity, which progressively worsened. Magnetic resonance imaging (MRI) revealed a large sacral tumor. Needle biopsy at another hospital revealed that the tumor cells formed palisading patterns with slightly proliferated chromatin. A slight nucleus malformation was observed.

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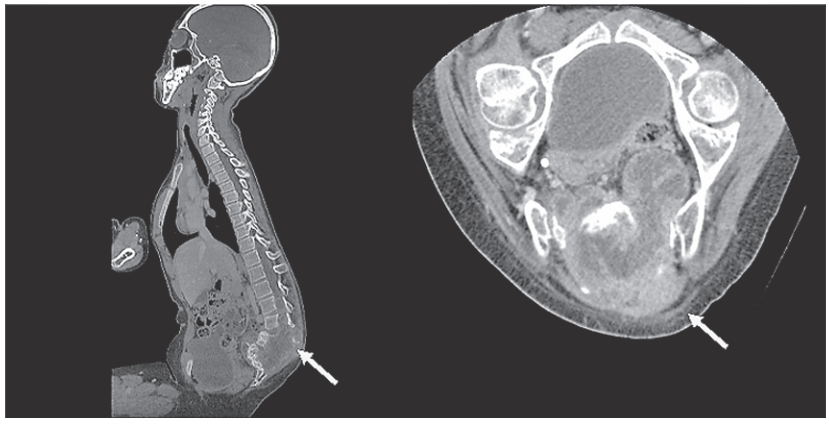

Fig. 1 Computed tomography with contrast revealed a solid sacral mass 12 × 8 × 8 cm in size, with a well-defined contour, high-density wall, and scalloping of the bone from L4 to the sacrum.

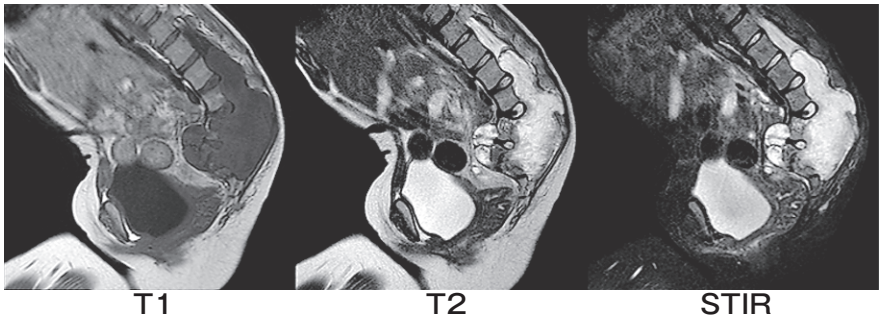

Fig. 2 MRI showed a large sacral tumor with well-defined margins. The tumor's signal intensity on MRI was equal to that of the abdominal muscles on T1-weighted images but was higher on T2-weighted images. The signal intensity of the tumor was not suppressed on STIR images. From the MRI images, the tumor appeared to originate from the right S2 nerve root.

Immunohistochemistry (IHC) showed that the tumor cells stained diffusely positive for S-100 protein and negative for keratin 7. Furthermore, p53 IHC was the wild-type form. These findings were compatible with a diagnosis of schwannoma.

When the patient presented to our department, she had gait disturbance, and could not lie down due to low back pain and swelling of the low back. Neurological status according to manual muscle testing (MMT) grade (right/left) was as follows: iliopsoas muscle 5(R)/5(L); quadriceps muscle 5(R)/5(L); tibialis anterior muscle 0(R)/4(L); extensor hallucis longus muscle 0(R)/3(L); flexor hallucis longus 0(R)/3(L) and gastrosoleus muscle 0(R)/3(L). Sensory deficit was observed bilaterally below the L5 dermatome. She had urinary incontinence and required laxatives for bowel evacuation. No café-au-lait spots or neurofibromas were observed.

Computed tomography (CT) revealed a solid sacral mass about 12 × 8 × 8 cm in size, with a well-defined contour, a high-density wall and scalloping of the sacral bone (Fig. 1). No substantial tumor enhancement or significant vascularity of the tumor was observed in the contrast-enhanced CT images. MRI also showed a large sacral tumor with well-defined margins. The tumor extended cranially to the L4 pedicle level, caudally to the S3 vertebra level, ventrally 6 cm from the S2 vertebral anterior wall within the retroperitoneal space, and 5 cm from the S2 posterior wall to the subcutaneous layer. The tumor's signal intensity on MRI was isointense on T1-weighted images and hyperintense on T2-weighted images. The signal intensity of the tumor was not suppressed in the short TI inversion recovery (STIR) images. MRI showed that the tumor appeared to originate from the right S2 nerve root (Fig. 2).

Because this patient's bony construct was scalloped

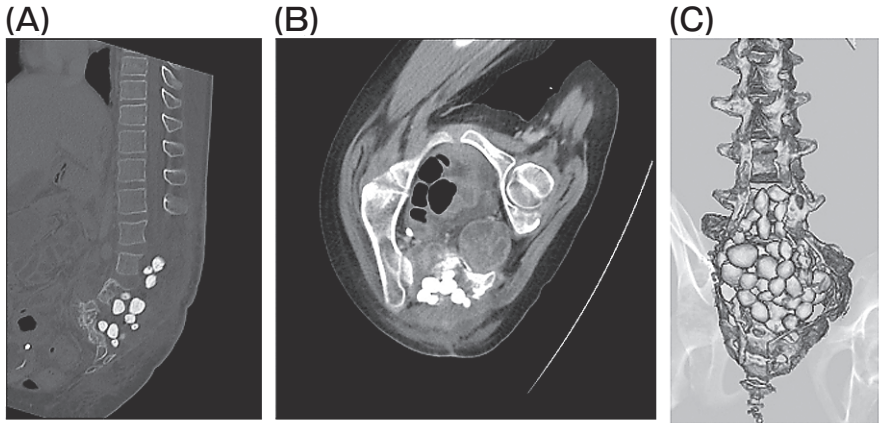

Fig. 3 The tumor cavity after posterior resection filled with cement beads. **(A)** sagittal plane image; **(B)** axial plane image; **(C)** 3D image.

and destroyed, we decided that reinforcement would be needed for her lumbopelvic construct. As the tumor size was large, two staged surgeries with maximum safe partial removal and reconstruction were planned. The second surgery was planned for one week after the first surgery, depending on the patient's recovery. During the first surgery, the tumor was excised as much as possible from the posterior approach. Because the bony structures were only scalloped, the tumor was resected by detachment from the sacrum and lumbar vertebrae. The sacral bone was not resected.

The surgery was performed under navigation using the O-arm. The reference frame was attached to the L3 spinous process, and the border of the tumor was easily recognized under navigation. Therefore, laminectomy was performed from L4 to S3, and the tumor was resected as precisely and safely as possible. Bilateral L5 roots were preserved. Intraoperative neuromonitoring indicated no change of amplitudes in the bilateral quadriceps and tibialis anterior muscles. The cavity after resection was filled with cement beads to prevent infection (Fig. 3). Vascular embolization was not performed before surgery, so the cavitron ultrasonic surgical aspirator (CUSA) was used for tumor resection to reduce bleeding. The operation time was 190 min, and estimated intraoperative blood loss was 3,200 mL. Histology of the resected tumor on the first operation indicated palisading patterns of tumor cells, high cellularity and invasion into part of the tissue around the tumor. IHC was the same as that of the needle biopsy. Therefore, the pathologist indicated the possibility of a high tumor proliferation rate.

We therefore decided to perform further resection of the tumor from the anterior aspect followed by posterior fixation at the second stage. A well-preserved cleavage plane between the tumor and the rectum helped mobilize the rectum away from the tumor. The soft elastic tumor protruded from the sacral promontory. The ureter and major vessels were protected, and the hypogastric nerves were dissected and released. With the vital structures dissected away and preserved, the tumor could be resected completely. The L5 vertebra and sacrum were severely scalloped by the tumor and were too weak to maintain spinopelvic continuity. Therefore, we had to perform a spinopelvic fixation.

The patient was subsequently positioned prone, and posterior fixation from L3 to the pelvis was performed using bilateral non-vascularized fibular bone grafts. Two grafts were applied between the bilateral iliac crests, and the others were placed extending from each iliac crest to the L3 spinous process. The grafts were fixed to the spinous process using a polyethylene cable (NESPLON cable system®, Alfresa, Japan) (Fig. 4). The bony structures were deformed due to the large size of the tumor, so O-arm navigation contributed to precise and safe screw insertion into the pedicles and iliac bones. The operation time was 560 min, and the estimated intraoperative blood loss was 2,400 mL. Intraoperative neuromonitoring revealed no change of amplitudes in the bilateral quadriceps and tibialis anterior muscles. Histopathological examination of the tumor specimen revealed a spindle cell tumor and high cellularity. IHC for S-100 protein indicated high positivity. In addition, IHC revealed reactivity for Ki67

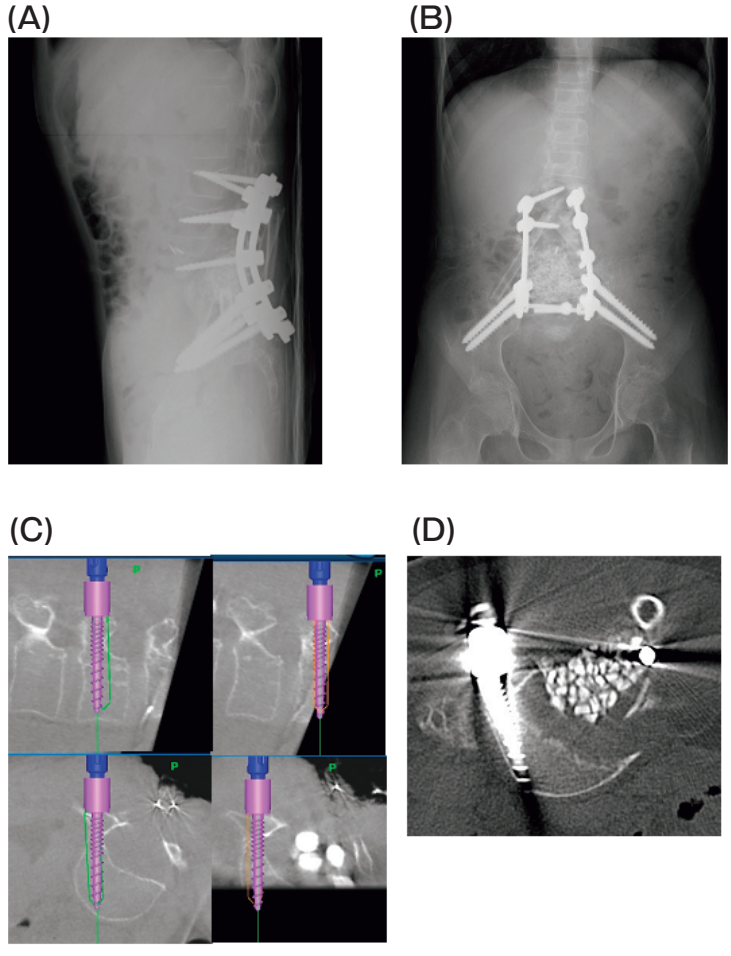

Fig. 4 **(A,B)** X-ray images after posterior fixation from L3 to the pelvis using bilateral fibular bone grafts; **(C)** intraoperative navigation images (left column, L4 vertebra; right column, L5 vertebra); **(D)** CT image of the pedicle screw inserted into the L5 vertebra using O-arm navigation.

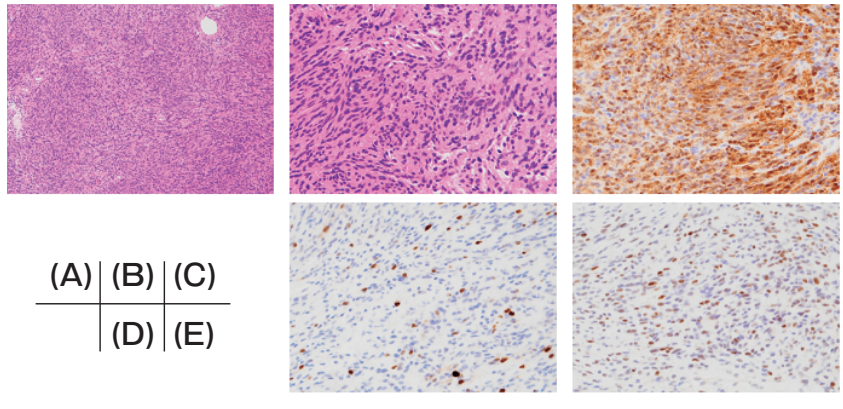

Fig. 5 Histopathological examinations of tumor specimens. **(A,B)** Hematoxylin-eosin-stained sections revealed a spindle cell tumor with high cellularity (a: low magnification; b: high magnification); **(C)** Immunohistochemistry (IHC) for S-100 protein indicated high positivity; **(D)** IHC for Ki67 protein revealed 5% reactivity; **(E)** IHC for p53 protein showed 20% positivity.

protein of 5%, and for p53 protein of 20%. These results confirmed the diagnosis of cellular schwannoma (Fig. 5).

There were no complications, such as postoperative infection, spinal fluid leakage, hematoma or delayed wound healing. Twelve days after the operation, gait training was initiated using parallel bars. The low back pain disappeared, and the patient was able to lie down. At 26 months of follow-up, the patient could walk independently with bilateral ankle orthoses. MMT grades (right/left) for the iliopsoas, quadriceps, tibialis anterior, extensor hallucis longus flexor hallucis longus, and gastrosoleus muscles were 5(R)/5(L), 5(R)/5(L), 1(R)/3(L), 1(R)/3(L), 1(R)/3(L), 1(R)/1(L) and 1(R)/1(L), respectively. The sensory deficit was the same as before surgery, and the patient required self-catheterization and laxatives as the bowel and bladder functions were not substantially improved. Follow-up MRI showed no recurrence of the tumor.

This study was conducted in accordance with the Declaration of Helsinki, and institutional ethical clearance was obtained before writing this report (No. 271). The subject provided written informed consent for the publication of this report.

Discussion

Schwannoma is observed in 30% of intradural extramedullary tumors, and rarely occurs in the sacrum [5]. Only 0.3-3.2% of schwannomas are retroperitoneal [6]. Because retroperitoneal schwannomas are usually slow growing and less symptomatic, diagnosis and treatment are delayed. In our case, this patient had been treated for cerebral palsy, and it was difficult to notice the signs and symptoms caused by the tumor as they were masked by neurological deficits and spasticity. A cellular schwannoma is often observed as a painless mass, and its origin is mostly retroperitoneal, including the pelvic extraperitoneal space and posterior mediastinum. Microscopic examination revealed that the features of cellular schwannoma differ from those of typical schwannoma because of increased cellularity and the absence of Verocay bodies. Due to its histological features, such as increased cellularity, hyperchromasia, and mitotic activity, and its clinical features, including bone erosion and local recurrence, cellular schwannoma may be misinterpreted by the clinician as a malignant tumor [7].

Sacral schwannoma requires surgical resection, but the operation is complicated by the anatomic location, with a tendency for local recurrence. Several surgical approaches have been reported [2]. For tumors located in front of the sacrum, the anterior approach is preferrable [3]. Posterior approaches can be performed to remove larger tumors involving the sacrum and presenting posteriorly [8]. When the tumor's extraspinal component is more extensive than the intraspinal and vertebral body components or when bony erosion is present, a combined approach is preferable [3]. In our case, we selected the combined approach for tumor resection. We then performed a posterior fixation with implants using bone graft from bilateral fibulae, since the tumor had complex anteroposterior involvement of the sacrum. There have been many reports of the utility of O-arm navigation for precise tumor resection by recognizing the tumor's orientation and for precise implant insertion into deformed bony structures [9-11]. In our case, the O-arm was very useful for placing pedicle screws in a narrow pedicle and resection of the tumor. Fortunately, no recurrence has been observed so far. We will continue long-term follow-up to check for tumor recurrence.

In this case, the patient's flexion-contracture of the knees and leg pain, which occurred 2 years prior to our involvement, were considered to be due to cerebral palsy, and myotomy and tendon elongation were performed. She was better for a short period, but started again to experience low-back pain and gait disturbance. She was later diagnosed with sacral schwannoma. We believe that the patient's initial symptoms were probably due to schwannoma but were masked by cerebral palsy. Therefore, if there is doubt in diagnosis, it is important to always consider other pathological possibilities and conduct neurological examinations or diagnostic imaging.

Conclusion

We performed a two-stage resection for a large sacral cellular schwannoma in a 12-year-old girl with cerebral palsy. After the surgery, the patient gradually became ambulatory with orthoses. MRI at 26 months of follow-up showed no recurrence of the tumor, and further long-term follow-up will be necessary.

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