Giant Sacral Schwannoma Causing Bilateral Hydronephrosis: Case Report and Review of the Literature

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Key words
- Giant
- Hydronephrosis
- Sacrum
- Schwannoma
- Surgical treatment

Abbreviations and Acronyms
MRI: Magnetic resonance imaging
T1WI: T1-weighted images
T2WI: T2-weighted images

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BACKGROUND: **Giant sacral schwannomas are very rare, and less than 1%—5% of spinal schwannomas are found in the sacral region. These frequently grow to considerable size because of permissive anatomic location and benign, slow growth of tumor. They can be unnoticed before reaching a huge size.**

CASE DESCRIPTION: **We report a rare case of a giant sacral schwannoma in a 46-year-old man. The patient presented with difficulty in passing urine, episodic constipation, and swelling of the right lower extremity for 6 months. Magnetic resonance imaging revealed 160 x 110 x 110 mm encapsulated heterogenous solid mass originated from left S1 spinal nerve extending into the pelvis and abdomen. Sigmoid colon and rectum were displaced to the right side, and bladder was displaced anteriorly. Left side of the S1 and S2 vertebral bodies, left S1 and S2 neural foramen were also eroded. It also compressed ureters causing bilateral hydronephrosis. The patient underwent a 2-stage procedure in which complete resection was achieved.**

CONCLUSIONS: **We report the second case of a completely resected giant sacral schwannoma with bilateral hydronephrosis in the literature. Performing a 2-stage procedure is important in giant sacral schwannomas. Morbidity can be minimized, and extent of resection can be maximized with the help of combined anterior/posterior approach.**

INTRODUCTION

Tumors developing from Schwann cells of the peripheral nerve sheath are called schwannomas and they are mostly benign. These can arise de novo or as a result of genetic conditions, such as neurofibromatosis type II. Most of these are found in the head and neck.[1] Spinal schwannomas are common tumors accounting for 25% of all primary spinal tumors,[2] and commonly occur in the thoracic region, followed by cervical and lumbar regions.[3] Schwannomas located in the sacral region are very rare, and less than 1%—5% of spinal schwannomas are found in the sacral region.[4] Sacral nerves found outside the spinal canal are usually the source of sacral schwannomas.[1] These frequently grow to considerable size because of permissive anatomic location and benign, slow growth of tumor.

Patients can present with mild symptoms, such as sciatica, low back, and abdominal pain. Motor weakness is rare and generally associated with malignant schwannoma.[5] Urinary retention and/or constipation can be caused by mass effect of the tumor on abdominal organs and neural tissues.[6] Hydronephrosis is also a rare situation in sacral schwannomas. Only 3 cases are present in the literature,[7-9] and just 2 of them are bilateral.[7,9] From these 2 cases, complete removal of the tumor was accomplished in 1 of them.[7] Despite the rareness of these tumors, early diagnosis is important to prevent serious complications.

Surgical treatment of these tumors is challenging because of the complex anatomy of the sacrum and surrounding structures. The aim of the treatment should be total surgical removal because adjuvant treatments, such as chemotherapy and radiotherapy, are not effective. Anterior, posterior, or 2-staged anterior/posterior approach can be used.

In this report, we describe a giant sacral schwannoma that originated from left S1 nerve root causing bony destruction and pelvic extension causing bilateral hydronephrosis owing to pressure on ureters. Tumor was totally removed via combined anterior and posterior approaches. We report the second case of a completely resected giant sacral schwannoma with bilateral hydronephrosis in the literature.

CASE DESCRIPTION

A 46-year-old man presented to our department with abdominal distention, difficulty in passing urine, episodic constipation, and swelling of the right lower extremity for 6 months. His complaints had increased during the past month. On physical examination, a huge mass was palpated in the lower abdomen. The patient had no focal neurologic deficit. There was no motor weakness and diminishing of reflexes. He did not have a family history of neurofibromatosis or spinal/brain tumors. He underwent magnetic resonance imaging (MRI) of the pelvis and lumbar spine for further evaluation. MRI

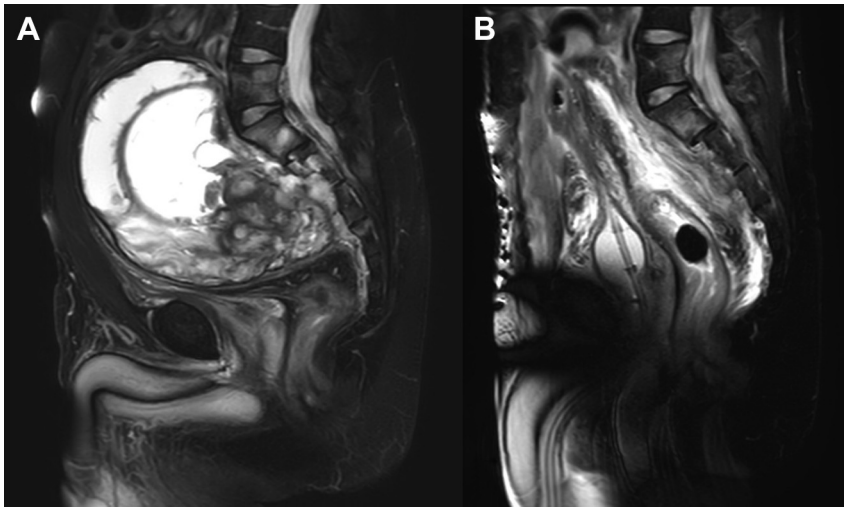

Figure 1. (**A**) Preoperative sagittal T2-weighted (T2WI) magnetic resonance imaging (MRI) showing a large retroperitoneal tumor involving the left hemipelvis and compressing the bladder and bowels. (**B**) Postoperative sagittal T2WI MRI showing complete removal of sacral schwannoma at 1 week after second surgery.

demonstrated 160 x 110 x 110 mm encapsulated heterogenous solid mass originating from the left S1 spinal nerve extending into the pelvis and abdomen. The central part of the mass was cystic. Sigmoid colon and rectum were displaced to the right side, and bladder was displaced anteriorly. Left side of the S1 and S2 vertebral bodies, left S1 and S2 neural foramen were also eroded. This also compressed ureters causing bilateral hydronephrosis (Figure 1). According to Klimo et al.[10] it was diagnosed as a type II tumor.

Two-stage surgery was decided. Nephrostomy tube was placed in the left side with fluoroscopic guidance by urologists. First, a midline laparotomy was performed by a general surgeon and neurosurgeon. After the incision was made on the mass, the subacute/chronic hematoma in the cystic cavity was evacuated. After shrinkage of the mass, it was stripped from the colon and rectum around which it was attached. Bilateral ureters and common iliac vessels were identified and preserved. Tumor was excised until anterior wall of the sacrum (Figure 2A). Methylene blue was injected into the rectum and there was no leakage. After strict hemostasis, drains were placed in the pelvis. The patient woke up with motor weakness (2/5) on the distal side of the left lower extremity. The second approach was performed by a neurosurgeon after 1 week. Sacrum was exposed via midline incision. Sacral bilateral laminectomy was performed. Left S1 and S2 nerve roots were identified. Anterior to these nerves, the tumor was attached to the left S1 nerve root with bone erosion in the left side of the body of S1-S2 vertebrae. However, left sacroiliac joint was intact. Piecemeal total tumor resection with capsule excision was done. Rectum was identified anterior to resection cavity. S1 and S2 nerve roots were well preserved (Figure 2B). Pathological examination revealed that the most portion of the specimen was composed of bundles of elongated cells with spindle-shaped nuclei, diagnosed as schwannoma (Figure 3). There was no additional neurologic deficit after the second approach and patient was discharged after 7 days. After 3 months, all patient's symptoms improved, including motor weakness on the lower extremity, with the help of physiotherapy. Also, no problem with sphincters were detected. The patient now has no complaint and neurologic deficit, and there is no recurrence 1 year after total resection of giant sacral schwannoma.

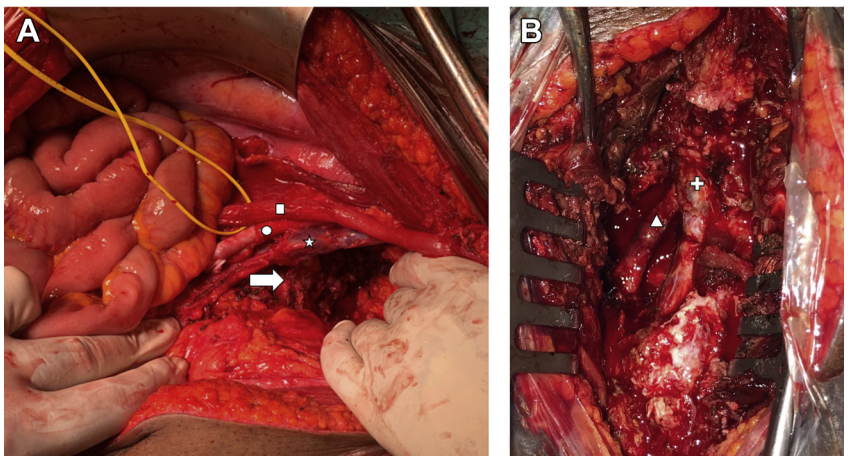

Figure 2. Intraoperative pictures. (**A**) Anterior portion of tumor has been removed in first stage. □, Left ureter; ○, left common iliac artery; ☆ left common iliac vein; → anterior wall of sacrum. (**B**) Posterior portion of tumor has been removed in second stage. △, Left S1 spinal nerve has been preserved; +, thecal sac.

DISCUSSION

Schwannomas are benign tumors originating from the sheaths of peripheral nerves or spinal nerve roots, however, they may rarely show malignant features.[11] Malign schwannomas are generally associated with von Recklinghausen disease.[12] Schwannomas are usually detected in the head and neck. Schwannomas located in the sacral region are very rare, and account for only 1% of cases.[8] These are slow-growing tumors. Symptoms can be detected when the spinal canal is completely filled by the mass or when the tumor grows into the retroperitoneal area by eroding the

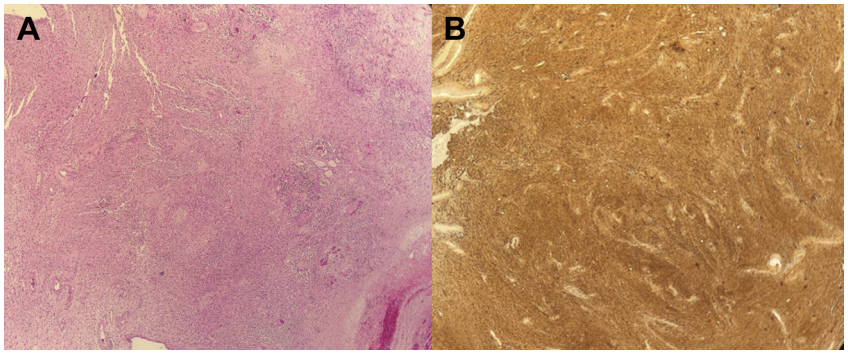

Figure 3. (**A**) Spindle cell proliferation with hypercellular and hypocellular areas and hyalinized vessels (hematoxylin and eosin, x 100), (**B**) immunohistochemical results for S-100 is diffuse positive in tumor cells (S-100, x 100).

sacrum, thus resulting in compression of bladder, colon, ureter, or neurovascular structures in the pelvis. Most of the patients' neurologic deficits are not present at the time of diagnosis, even though the tumor arises from the peripheral nerve sheath.[13] These can continue growing for a long time without symptoms. As a result, the tumors are huge when discovered.

Schwannomas can appear with various symptoms depending on the structure they compress. Bladder compression presents as urinary retention, colon compression as constipation, ureter compression as hydronephrosis, and neurovascular compression as swelling and pain in the lower extremity.[9] The current case report showed that the patient exhibited severe and painful symptoms and presented with a giant pelvic mass with features of urinary retention, constipation, bilateral hydronephrosis, and swelling and pain in the left lower extremity.

Klimo et al.[10] have developed a classification that assists the surgeon to understand the nature of tumor and also aids to decide the proper surgical approach. Ragurajaprakash et al.[9] reported a type II giant sacral schwannoma case, reviewed the literature, and stated that there were only 17 type II giant sacral schwannomas in literature. Also, only 2 cases of giant sacral schwannoma causing bilateral hydronephrosis have been reported in the literature.[7,9] Incomplete resection was performed in 1 of them.[9] Therefore our patient is the second case of completely resected giant sacral schwannoma with bilateral hydronephrosis in the literature.

Many schwannoma series[14-17] showed female dominancy and most often in their second to fifth decades of life. Our patient was 46 years old, which is consistent with previous studies.

Schwannomas have no specific radiologic features. However, MRI may show calcification, isointense signal to the adjacent skeletal muscle in T1-weighted images (T1WI), hyperintense signal in T2-weighted images (T2WI), heterogenous contrast enhancement due to hemorrhagic changes, and cystic appearance.[14,18] Also, bone erosion can be seen on computed tomography images. There is a wide differential diagnosis for sacral masses. Chordoma, giant cell tumor, neurofibroma, paraganglioma, and sarcoma should be kept in mind for differential diagnosis.[19,20] Imaging methods show many characteristics of tumors, but the exact diagnosis is made by pathologic examination. There was heterogenous contrast enhancement owing to hemorrhage on T1WI of our patient. Also, the tumor was isointense on T1WI, and hyperintense on T2WI compared with adjacent skeletal muscle, as mentioned in previous studies. However, in our study, the MRI findings of sacral schwannoma was a well-circumscribed lesion with complex signal intensities on T2WI. This heterogeneous appearance may be due to remote hemorrhage and degeneration, necrosis, and liquefaction of tumor tissues. Therefore a well-circumscribed lesion with a heterogenous signal intensity on T2WI may be more helpful for preoperative diagnosis.[14]

Sacrum has a complex anatomy, which makes it difficult to excise schwannomas when giant and locally extending, even though they are benign.[6,21] When deciding on the surgical approach, the location, size, and proximity of the mass to the surrounding tissues should be considered. Anterior, posterior, or combined anterior/posterior approaches should be preferred. In some cases, the tumor can be totally removed in a single session with a single approach, but in most cases, the combined approach should be preferred because it may be necessary to remove more bone in one approach to increase the surgical exposure, which may result in postoperative pain and iatrogenic spinal instability. In addition, it is more likely to damage neighboring normal anatomic structures in a single approach. According to a review conducted by Khan et al.,[6] total excision rate is higher (71%) in combined approach and anterior or posterior approaches.

Regarding extent of resection, in some reports, it has been stated that total excision caused neurologic deficit[22,23]; in other reports, they concluded that partial excision might cause recurrence.[15,23] In some series, subtotal resection was preferred for reducing morbidity.[15,22,24] However, recurrence is a major problem in these series. The total removal is more likely prone to neurologic deficit, and partial removal is more likely to cause recurrence, or even rarely malignant transformation.[25] Pongsthorn et al.[15] reported a recurrence of 16% in their series of subtotal removal of sacral schwannomas. Also, when tumor recurs, the probability of morbidity will be higher in the second surgery because of the adhesion of surrounding structures, such as the bladder, bowel, or neurovascular structures. Therefore total excision should be the aim. In our patient, we performed the anterior approach first to protect abdominal and pelvic structures, and then the posterior approach to preserve the sacral nerves and sacrum. There was no need for instrumentation or brace because of the protection of spinal integrity with relatively limited bone removal in the posterior approach and the absence of tumor invasion in the sacroiliac, lumbosacral joints. However, the patient encountered neurologic deficit, albeit transient. Previous studies described that intraoperative neuromonitoring decreases the complication rate due to neural tissue damage.[6,24] We did not use it in our case;

however, it could prevent the development of neurologic deficit.

CONCLUSIONS

Giant sacral schwannomas with bilateral hydronephrosis are rare tumors. Their surgical treatment is challenging due to their huge size and complex anatomy of sacrum. Giant sacral schwannomas should be operated with a multidisciplinary approach by a neurosurgeon and general surgeon to achieve the best outcome and total removal. The ideal and curative treatment of sacral schwannomas is the complete removal of the tumor without complications and spinal instability. Adjuvant treatment is not recommended. Further studies are needed to reveal recurrence rates.

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