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A case report of a large gluteal schwannoma with pelvic extension

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ABSTRACT

Benign schwannomas are slow-growing, painless tumours originating from Schwann cells, which form the sheaths of peripheral nerves. These tumours are relatively rare, with an incidence of 1-3 cases per 100,000 individuals annually. While they are most commonly found in the head, neck, and spine, schwannomas can also occur in the extremities, particularly in the upper limbs. Tumours in the pelvis and gluteal region are less frequent but are clinically significant due to their potential to compress adjacent structures.

We present a case of a 74-year-old male patient who underwent surgery for a large schwannoma in the gluteal region, extending into the pelvis at the level of the piriformis muscle and sciatic foramen. Initially misdiagnosed and treated as sciatica, this case highlights the importance of considering schwannomas in the differential diagnosis of patients presenting with neurological symptoms in unusual locations. Sciatic symptoms that do not respond to conservative treatment should be further investigated, and a thorough palpation of tender and painful points should always be performed to aid in the potential diagnosis of a local soft tissue tumour.

INTRODUCTION

Benign schwannomas are slow-growing, painless tumors that originate from Schwann cells, which form the sheaths of peripheral nerves. These tumors are most commonly observed in females between their 20s and 40s. While the typical size does not exceed 6 cm in diameter, cases with larger tumors, reaching up to almost 30 cm, have also been documented (1). Schwannomas frequently occur in the region of the head, neck and spine, while the localization of these tumors in the extremities is somewhat less common (2). Schwannomas are more

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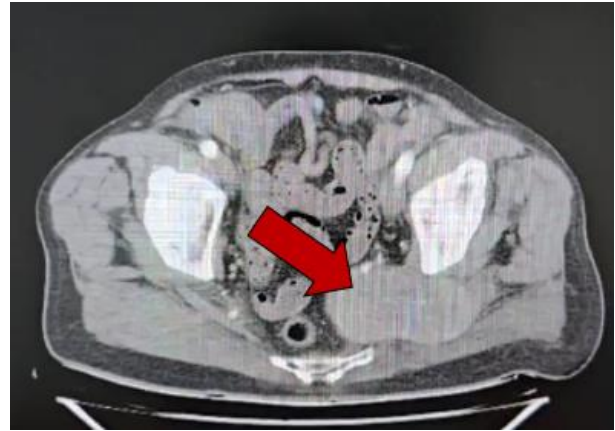
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frequently found in the superficial layers of the body along peripheral nerve locations. However, they can also develop in deeper, less visible regions like the retroperitoneum or mediastinum. In such hidden locations, diagnosing schwannomas is often postponed due to the lack of noticeable clinical symptoms until the tumor grows significantly larger, leading to compression of intra-abdominal and intra-pelvic structures (1, 3, 4). The majority of schwannomas are asymptomatic; however, they may lead to functional impairments and pain, depending on the specific nerve involved (5). In locations where schwannomas can grow asymptotically for years, clinical symptoms typically appear when exceptionally large tumors begin to compress intra-abdominal or pelvic organs (1).

We present a 74-year-old male patient with a massive gluteal schwannoma extending into the pelvis in the projection of the piriformis muscle.

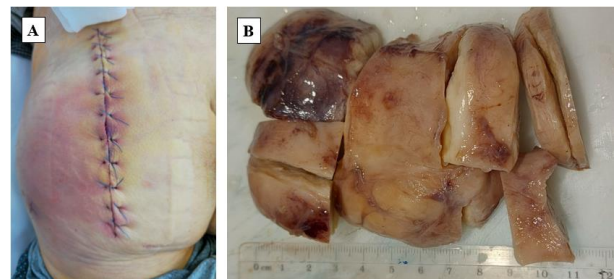
CASE REPORT

The case presents a 74-year-old patient who sought medical attention due to pain in the gluteal region and difficulty sitting, accompanied by numbness in the left leg. The patient had previously undergone several months of treatment for lumbosciatic symptoms, which did not respond well to the prescribed therapy. Due to the emergence of symptoms, including difficulty and pain during sitting and since deep palpation of the gluteal region revealed a painful mass, while deep abdominal palpation was painless and unremarkable, a computed tomography (CT) scan of the abdomen and pelvis was performed. The scan revealed an extensive well defined tumor measuring approximately 15 x 8 x 5 cm in the right gluteal region, with extension into the pelvis, predominantly in the region of the left piriformis muscle and sciatic foramen (picture 1). The tumour appeared hyper-vascular. The radiological diagnosis was a mass of neurogenic origin, most likely the sciatic nerve and suggested neurofibroma or schwannoma, while our initial clinical diagnosis of a soft tissue sarcoma involving the left gluteal region and pelvis was considered. Due to the clear visualization on the CT scan and the patient's pronounced claustrophobia, it was decided not to proceed with further radiological evaluation, specifically not to perform an magnetic resonance imaging (MRI).



Picture 1. A CT scan of the pelvis, with an arrow marking a large, well-defined tumor, which is partly located in the pelvis and partly in the gluteal region, in the projection of the piriformis muscle and the sciatic foramen.

A two-stage surgery was planned, with the first stage involving the resection of the tumor from the gluteal region. The patient was operated on by a team consisting of a general surgeon, orthopedic surgeon, and neurosurgeon. The first stage of the surgery involved a linear incision in the gluteal region (Picture 2A), in the projection of the sciatic nerve. Through careful dissection, which included both blunt and sharp techniques, the tumor was removed in one large piece (Picture 2B), and there was no need for a second surgical stage. It was impossible to trace the originating nerve. The patient recovered well, without neurological deficits, and three months post-surgery, there were no signs of tumor recurrence on the follow-up CT scan. The patient remained symptom-free, with a normal neurological examination. Histopathology confirmed the diagnosis of a schwannoma, and further clinical follow-up was recommended.



Picture 2. (A) The image shows a linear incision in the left gluteal region in the projection of the sciatic nerve, one day after surgery. **(B)** The image shows the tumor removed in its entirety, and then cut by the pathologist for further histopathological examination.

DISCUSSION

Schwannomas are relatively uncommon benign tumors, with an estimated incidence of 1-3 cases per 100,000 individuals annually. These tumors typically arise from Schwann cells in peripheral nerves and are most commonly found in the head, neck, and spine. In terms of extremity schwannomas, these tumors are more commonly observed in the upper limbs, though they can also be found in the lower extremities. These schwannomas generally present as slow-growing, well-circumscribed, and mobile masses, often palpable under the skin. Schwannomas located in the pelvis and gluteal region are much less frequent but are clinically significant due to their potential to cause compression of adjacent structures (6, 7). Although these tumors are more commonly observed in female patients at a younger age, typically between the 2nd and 4th decades of life, we present a rare case of schwannoma in a 74-year-old male patient, with a specific localization in the gluteal region, pelvis, and in the projection of the piriformis muscle and sciatic foramen. The tumor is of considerable size, indicating that it has been asymptomatic for a relatively long period.

While the exact pathogenesis of schwannomas remains unclear, their development may be associated with specific gene mutations. Schwannomas of peripheral nerves are typically well-defined, encapsulated masses that are usually round and connected to the nerve. These tumors originate from Schwann cells and are often located eccentrically, affecting one or two fascicles while leaving the other neural fascicles of the nerve intact and displaced. Schwannomas form within the endoneurium and are encased by the perineurium and fibrous epineurium, which collectively surround and encapsulate the tumor (8). In the case of retroperitoneal, pelvic, sacral, and gluteal schwannomas, the average growth rate of the tumor is about 2 mm per year, e.g. schwannomas have a slow growth rate. Additionally, these schwannomas display strong concealment abilities due to the large pelvic space, and thus are associated with a range of clinical symptoms, such as compression of pelvic organs or nerve tissue, bone destruction, sciatica, lower back pain, difficulty with urination and defecation due to bladder and rectal compression, and weakness in the lower limbs (8, 9). In our patient, the size of the tumor suggests that it was likely

asymptomatic for several years. Symptoms then developed, which were initially interpreted as sciatica. However, as the patient did not respond to standard therapy after several months, a follow-up examination was performed, during which a painful palpable mass was found in the gluteal region. This led to further radiological evaluation.

Computed tomography (CT) and magnetic resonance imaging (MRI) are commonly employed imaging modalities for assessing soft tissue tumors. However, CT's diagnostic utility is limited by its relatively low resolution and suboptimal soft tissue contrast, often failing to clearly depict the stromal heterogeneity characteristic of schwannomas. Enhanced resolution can often be achieved with the use of intravenous contrast agents, such as iopamino. MRI, when available, is considered the preferred imaging technique (10). In our case, CT imaging clearly revealed a large, well-defined, and demarcated tumor in the left gluteal region, extending into the pelvis at the level of the sciatic foramen. The tumor's clear delineation, without surrounding soft tissue reaction, raised suspicion of a schwannoma, although our initial diagnostic assumption was a sarcoma. Due to the clear visualization on CT and the patient's claustrophobia, we opted not to perform an MRI. The subsequent course of action was surgical treatment. Initially misdiagnosed and treated as sciatica, this case highlights the importance of considering schwannomas in the differential diagnosis of patients presenting with neurological symptoms in unusual locations. Sciatic symptoms that do not respond to conservative treatment should be further investigated, and a thorough palpation of tender and painful points should always be performed to aid in the potential diagnosis of a local soft tissue tumor, subsequently followed up with the necessary radiological diagnostics.

Complete surgical removal is the treatment of choice for large schwannomas. Some authors recommend wide local excision for retroperitoneal and pelvic schwannomas, believing that malignancy can never be entirely ruled out. However, tumor recurrence or malignant transformation is extremely rare in benign schwannomas (1, 11). Successful surgical resection of retroperitoneal or pelvic schwannomas requires thorough and meticulous preoperative planning. The patient should be well-informed about the potential risks of residual

functional impairment, which may arise from nerve damage or muscle fibrosis. Challenges in tumor excision often stem from the tumor's proximity to surrounding neurovascular structures and its blood supply. Ideally, the surgical team should include different specialists such as neurosurgeon, general surgeon, vascular surgeon, and urologist. The anesthesiologist should also have experience managing procedures with a high risk of significant blood loss. Surgery in multiple stages is often used for similar tumors (1). In our case, the clinical diagnosis initially suggested a sarcoma, but radiological evaluation, specifically the CT scan, led to the conclusion that the tumor was most likely a schwannoma. As a result, a team consisting of a neurosurgeon, general surgeon, and orthopedic surgeon was assembled, along with an experienced anesthesiology team, and a two-stage surgical approach was planned. However, through a combination of sharp and blunt dissection, the tumor was completely removed via a posterior approach in a single stage. The nerve origin was not identified, even though preoperative suspicion pointed to the sciatic nerve. The patient's preoperative symptoms completely resolved, and the neurological signs, primarily the tingling in the left leg, were most likely a result of compression on the sciatic nerve.

Although large schwannomas of the same location have been described in the literature, in our case, the tumor was one of the largest schwannomas of this location reported so far.

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