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# Unusual Giant Pre-Sacral Multicystic Schwannoma: A Case Report and Differential Diagnosis

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## Abstract

**Background:** Schwannomas are generally benign neoplasms arising from the nerve sheath. Presacral schwannomas are very rare entities and difficult to diagnose, representing less than 15% of all retrorectal space tumors. Benign schwannoma sometimes displays degenerative changes, such as cyst formation, calcification, hemorrhage, and hyalinization. Usually these degenerations are partially seen in the tumors. **Objective:** To point out that presacral schwannoma can display markedly multilocular cystic degeneration. **Case Report**: We present this unique case of a 24-year-old man diagnosed with an unusually large pure multilocular cystic schwannoma, which is revealed by digestive, urinary, and nonspecific neurological symptoms. The patient was successfully treated with radical surgery via an anterior approach leading to the recovery of symptoms. **Discussion and Conclusion:** This tumor was unusual in its totally multicystic appearance and its resemblance to a wide spectrum of lesions that can occur in the pre-sacral space, such as hydatid and developmental cysts. Preoperative diagnosis is essential to prevent major neurological deficits during surgical intervention.

## Keywords

Multicystic Schwannoma, Pre-Sacral Space, Differential Diagnosis

## 1. Introduction

Schwannomas are benign pseudo-encapsulated tumors that arise from the neop-

lastic transformation of Schwann cells of the neural sheath. Pre-sacral (PS) schwannomas are found in less than 5% of all spinal schwannomas whoever the exact incidence still unknown since the majority remain asymptomatic [1] [2].

Some degenerative changes like calcification, fibrosis and hemorrhage are frequently seen in schwannomas, but cystic change is rare [3]-[5]. A purely multilocular cystic schwannoma that looked like a "Wasp Nest" is exceptional and never seen before.

The PS space is a clinically important space that concerns several disciplines and is affected by a wide variety of benign and malignant conditions which can be a challenge in their management [6]-[10]. The main objective of this article is to present this unique case of giant PS multicystic schwannoma and to formulate appropriate differential diagnoses list by considering the specific clinical and imaging features of a given case, which is a mandatory step in pre-surgical planning.

## 2. Case Report

A 24-year-old young man from a rural area, suffering from proctalgia for 2 years with progressive worsening. The clinical examination found minimal rectal bleeding, dysuria, and unsystematized paresthesia of the right lower limb evolving for 1 year.

Pelvic CT scan showing a PS cystic lesion with seated fluid density with smooth septa and calcifications. The mass extends into the endo-sacral canal and erodes the anterior surface of the sacrum with an invasion of the left sacral holes S3/S4 and S4/S5 (**Figure 1**).

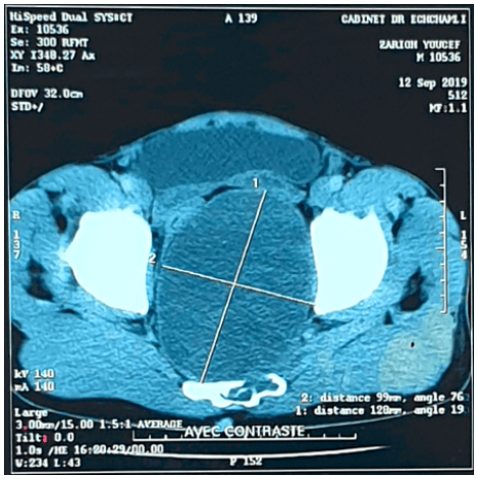


**Figure 1.** Pelvic CT scan showing presacral cystic lesion with seated fluid density with smooth septa and calcifications. The mass extends into the endo-sacral canal and erodes the anterior face of the sacrum with the invasion of the left sacral holes S3/S4 and S4/S5.

Magnetic resonance imaging (MRI) shows a giant multicystic lesion developed in the PS space measuring 96 × 126 × 128 mm. The lesion had a low signal in the T1-weighted sequence and it had a clear multilocular fluid content with a

high signal in T2-weighted with an attenuated high signal in FAT saturation sequences, delimited by thin septa and well circumscribed by a wall in low signal T2 . Calcification-related signal voids were identified in all MRI sequences. No solid component is identified and no contrast enhancement has been demonstrated. A massive portion of the mass falls into the pre-sacral space pushing the rectum and bladder forward. A comprehensive workup for any hydatid cyst was negative. Cystic schwannoma, Developmental cysts and other diagnosis had been evoked. Therefore a surgical indication undertaked for resection, exploration and histological evidence (**Figures 2(a)-(c)**).

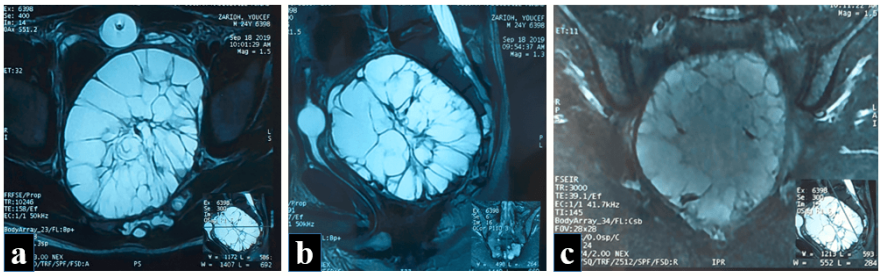


**Figure 2.** MR images demonstrating a giant multicystic lesion developed in the pre-sacral space. The lesion had a clear multilocular fluid content with a high signal in T2-weighted with an attenuated high signal in FAT saturation sequences, delimited by thin septa and well circumscribed by a wall in low signal T2. Calcification-related signal voids were identified in all MRI sequences. No solid component is identified and no contrast enhancement has been demonstrated. A massive portion of the mass falls into the pre-sacral space pushing the rectum and bladder forward. (a) Axial T2-weighted image. (b) Sagittal T2-weighted image. (c) Coronal fat saturation-weighted image.

The surgery was performed through a collaboration between the neurosurgeon and the visceral team via an anterior approach. A median abdominal incision was made and exploration discovers a pelvic process with a sacred implantation base, pushing the bladder forward and the left ureter and coming into contact with the external and internal iliac artery and vein with contact with the rectum. The opening of the recto-vesical fornix allowing a release of lateral parietal adhesions of the mass. Then the liberation of the mass of vascular attachments and left ureter and dissection of the mass of the anterior face of the sacrum was carried out. After the ligature of the ilio-ombilical artery, total removal of a solid mass carrying mesorectum and small cysts in contact with the roots was accomplished. Hemostasis was made by surgicel and surgiflo. The mass was rubbery, with the consistency of fibrin glue (**Figure 3**).

Histopathological and immunohistochemical examination showed Antoni B area tissue with nuclear palisading in a neurofibromatosis background, supporting the diagnosis of cystic schwannoma (**Figures 4(a)-(c)**).

The patient enjoyed a great recovery. The postoperative course was uneventful and was discharged home 5 days after surgery without any complications. At 1-month follow-up, the patient reported that preoperative paresthesia, dysuria, constipation, and rectorrhagia were markedly improved. The 1 year postopera-

tive MRI showing no evidence of disease recurrence or other localization on the cerebrospinal axis (Figures 5(a)-(c)).

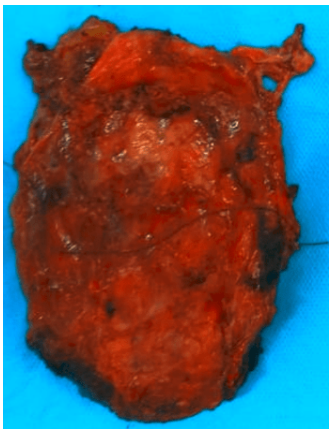

Figure 3. Pathological specimen of giant multicystic schwannoma.

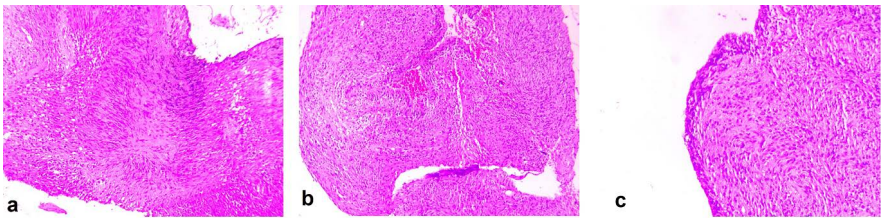


Figure 4. Histological image of the schwannoma. (a) Palisade arrangement of tumor cells forming Verocay bodies. (b) Alternation between Antoni A high cellular zone and Antoni B low cellular zone. (c) Ependymal cells in the surface.

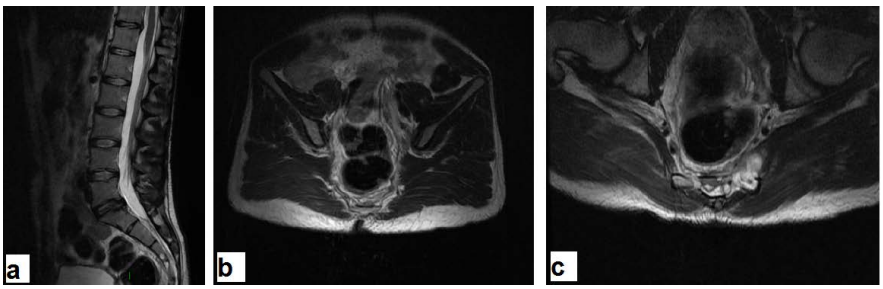


Figure 5. The 1 year postoperative cerebrospinal MRI that show no evidence of residual tumor, no recurrence and no other localization. (a) Axial T2-weighted image. (b) Coronal T2-weighted image. (c) Axial T2-weighted image.

## 3. Discussion

PS schwannomas are rare and slow growing tumors, which arises most commonly in the sacral nerves [7]. The majority of PS schwannomas are solid tumors. Predominantly cystic lesions are rarer. A purely multilocular cyst is exceptional and has never been published before in the literature.

Because of their slow-growing nature, symptoms do not manifest until the tumor grows to be quite large. Even when the tumors have grown large, they remain a diagnostic dilemma because of a wide variety of non-specific symptoms

and findings on imaging [8]-[11]. Presenting symptoms are most commonly progressive lower back pain, numbness or paresthesia, neurological deficits, and failure of conservative therapy such as physical rehabilitation [8].

However, acute lower back pain may also be caused by nerve root torsion or hemorrhage within the tumor itself [12] [13].

Non-specific abdominal pain or discomfort, constipation, and rectorrhagia, or urological symptoms like urinary incontinence and hematuria have also been reported [14]. Most of them are related to mass effect and compression or involvement of surrounding organs.

MRI is the most useful imaging modality for the diagnosis of these tumors. Schwannomas have a low-to-intermediate signal intensity on T1-weighted images and a heterogeneous appearance with focal areas of hyper- and hypo-intensity corresponding to cyst formation on T2-weighted images. In contrast, rim enhancement of an intradural extramedullary mass is characteristic of a schwannoma [8] [15] [16]. Well-developed or "ancient" schwannomas often show cystic changes secondary to stromal and vasculature degeneration, demonstrate as non-enhancing T2-hyperintense fluid pockets. Cystic schwannomas often contain mixoïd stroma with a mucoid matrix surrounded by collagenous fibrous tissue filled with mucopolysaccharide acid, the reason why some tumor has a cystic part that appears in high signal T2-weighted sequences and T1 weighted with or without late gadolinium enhancement [8] [11] [12] [14] [16]. CT is also helpful which allows a better analysis of the involvement of the sacral bone and can show a non-specific, well-circumscribed lesion with a low or heterogeneous signal, sometimes with central necrosis [8] [16].

The differential diagnosis for a large cyst in this location with multilocular appearance and sacral bone involvement is extensive, including a wide variety of conditions that occur in the retrorectal space including hydatid cyst, developmental cysts, cystic sacrococcygeal teratoma, anal duct or gland cyst, necrotic rectal leiomyosarcoma, extraperitoneal adenomucinosis, cystic lymphangioma [8] [17]-[19].

### 3.1. Hydatid Cysts

Hydatid cysts are caused by Echinococcus granulosus. Skeletal hydatidosis only occurs in 0.5% - 2% of cases. The localization in the PS space is very rare [20]-[23], the most frequent clinical manifestations are an abdominal mass, pain and, less frequently, compression of organs such as the bladder or the rectum [22]. Hydatid disease can affect any part of the human body and often has non-specific imaging results and serological tests are not always positive, according to the literature [24]. CT scan and MRI make it possible to know the extent of the disease, but also to control the regression after treatment [25]. Both techniques show the multilocular and cystic nature of tumors [22]. The rim sign is more evident on T2-weighted MRI images and represents the pericyst and parasitic membranes. It is described as a low signal intensity edge that suggests hydatid disease rather than other non-parasitic processes [25] [26].

### 3.2. Developmental Cysts

Developmental cysts are the most common congenital entity found in the retrorectal space in adults. They are classified into epidermoid and dermoid cysts, enteric cysts and neurenteric cysts according to their origin and histopathological features. Although developmental cysts are often asymptomatic, patients may present with symptoms resulting from a local mass effect on the rectum with a retrorectal mass palpable at digital rectal examination and from local compression on the lower urinary tract. Obstructed labor and sciatic pain have been reported. A well-defined, unilocular or multilocular thin-walled cystic lesion is the primary imaging feature. Rarely, a sacral bone defect and calcifications are associated with dermoid, tailgut, and neurenteric cysts [27].

### 3.3. Cystic Sacrococcygeal Teratomas

Sacrococcygeal teratomas are germ cell tumors containing elements derived from the three germ layers they are benign and are the most common PS lesions found in the pediatric age group. The prevalence of malignancy increases with age; however, they are rarely discovered in adult life. On CT or MRI imaging, sacrococcygeal teratomas appear as heterogeneous and well-defined lesions with solid cystic and mixed components. Rarely, sacrococcygeal teratomas are completely cystic and these are more likely to be benign. Sacrococcygeal teratomas contain fat or calcifications in 50% of cases. Some patients (20%) present with sacrococcygeal bone abnormalities [27] [28].

### 3.4. Anal Duct or Gland Cyst

Anal duct or gland cysts are mucus-secreting cysts lined with a combination of stratified squamous epithelium, columnar epithelium, and transitional epithelium. They often communicate with an anal canal or crypt. CT and MR imaging shows a uni- or multilocular cystic lesion near the anal sphincter in the retrorectal space. Anal duct or gland cysts may involve the coccyx and the ischio-anal space [29].

### 3.5. Rectal Leiomyosarcoma

Rectal leiomyosarcomas are rare malignant smooth muscle tumors, accounting for less than 0.1% of all malignant rectal tumors and are more common in men. They can involve the rectal lining, leading to superficial rectal ulceration and bleeding. Imaging shows a well-circumscribed, heterogeneously enhanced multilobular tumor with cystic necrotic components with the possibility of sacral invasion [30].

### 3.6. Subperitoneal Pelvic Adenomucinosis

Subperitoneal pelvic adenomucinosis is rare. Cysts are lined with glandular epithelium and filled with thick gelatinous material. Computed tomography and MRI show well demarcated, homogeneous and compartmentalized cystic le-

sions. Rarely, wall nodules and calcifications or bone involvement are observed [31]-[33].

### 3.7. Cystic Lymphangioma

Cystic lymphangioma is a rare congenital benign tumor and is more commonly seen in the pediatric age group and in cervical locations. Occurrence in a subperitoneal pelvic location is rare. Cysts are lined with endothelium and filled with a clear fluid. Cystic lymphangioma appears as a well-defined multicystic lesion with thin septa. The most common complications are intracystic infection and bleeding [34].

## 4. Conclusion

In conclusion, the diagnosis of multicystic schwannoma in the presacral space is extremely rare and can mimic other entities on clinical and radiographic imaging findings. Care should be taken to include this type of tumor in the differential diagnosis of a retrorectal cystic lesion, as treatment of a schwannoma may be significantly different from another such as a hydatid or developmental cysts.

## Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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## Conflicts of Interest

There are no conflicts of interest.

## Author Contributions

Khoulali Mohamed contributed to patient management, data interpretation, and writing the manuscript. Mehfoud Ibrahim and Mejdoubi Anasse contributed to acquisition, analysis and interpretation of data. Oulali Nourddine, Moufid Fayçal, Bouziane Mohammed and Housni Brahim designed the project and supervised the accuracy and integrity of all stages of manuscript preparation. All authors were involved in the management of the patient from admission to follow-up, worked on the draft, and revised it together.

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