International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2023: 1.843

Giant Perineural Cysts Simulating Pelvic Pathologies

Dr. Gayatri Ulhas Harshe[1], Dr. Mitusha Verma[2], Dr. Deepak Patkar[3], Dr. Rushabh Narsihbhai Patel[4]

[1]Consultant, Radiology Department, NMSSH, Mumbai

[2]Senior Consultant, Radiology Department NMSSH, Mumbai

[3]Head of the Department, Radiology Department NMSSH, Mumbai

[4]Fellow in Body Imaging, Radiology Department, NMSSH, Mumbai

Abstract: ***Background:*** ***Giant perineural cyst are mostly symptomatic and these mimic pathologic entities. Tarlov cyst (TC) or perineural cysts are the cerebral spinal fluid (CSF) filled sacks usually located at the junction of the dorsal ganglion and posterior nerve root sheaths of spinal nerves. Two patients with lower abdominal pain. An ultrasonographic evaluation in first patient reveals hydrosalpinx. And in second patient pelvic collection. MRI showed multiple, bilateral and almost symmetric tubular presacral space and pelvic extension.*** ***Conclusion:*** ***MRI is a very useful for evaluation of perineural cyst. It familiarizes the clinicians with unusual presentation of perineural cysts, which helps unnecessary investigations and early detection.***

Keywords: Perineural cyst, Tarlov cyst, Giant, nerve root cyst, hydrosalpinx, pelvic collection

Abbreviations and Acronyms: MRI (Magnetic resonance imaging) , TCs (Tarlov cysts), CSF(cerebrospinal fluid)

1. Introduction

Tarlov cysts (TCs) are perineural cysts that are filled with cerebrospinal fluid (CSF) and have mainly a sacral origin, therefore also called sacral perineural cysts [1,2]. They develop from the dorsal ganglion or the spinal posterior nerve root and arise between the perineurium and endoneurium [1]. The lining of these lesions contain nerve fibers [3,4] however exact etiology has not been established.

Several hypotheses have been suggested such as inflammation, trauma, congenital origin, degenerative processes and Park et al even suggested familial inheritance as a possible cause[3]. American neurosurgeon, Isadore M. Tarlov first described five cases of such a cyst in 1938 that he had encountered while performing autopsies and published an article on the same [5]. Tarlov hypothesized that hemorrhage into the subarachnoid space caused accumulations of red blood cells, plugging of veins of the perineurium and epineurium which finally leads to rupture and cyst formation. Out of a total seven patients Tarlov's 1970 article, four had a history of trauma [6]. Such a hypothesis was also supported by Schreiber and Haddad [7]. Also, because many patients did not have history of trauma, Fortuna et al. hypothesized that the origin of perineural cysts was congenital caused by congenital proliferations within the root sleeve [8].

Endopelvic extension of TCs is uncommon [9] and can be misdiagnosed as adnexal mass on gynecological ultrasound imaging and can lead to unnecessary laparotomy [2].

Plain x- rays involving perineural cysts may appear normal however sometimes they show bony erosion of the spinal canal or the anterior or posterior neural foramina [10]. CT scans can delineate as isodense CSF filled cystic masses at the foramina may or may not be accompanied with bone re-modelling, which extends into the pelvis [11].

MRI gives much better soft tissue contrast and is currently the investigation of choice in the diagnosis and further management of perineural cysts. These cysts are CSF signal intensity cysts and follows CSF signal on all pulse sequences on MRI imaging. These cysts are hypointense on T1 images and hyperintense on T2 images, without any diffusion restriction and show thin peripheral wall enhancement [12]. Post contrast myelography shows filling of the meningocele sac one hour after injection of contrast agent and is highly suggestive of a perineural cyst. This is replaced by MR myelography sequence. Electro myelography can be used in assessing patients with neurologic symptoms [13].

2. Case Report

Table 1: Clinico-radiological features of patients

	Patient 1	Patient 2
Sex	Female	Female
Age	46 years	48 years
Clinical symptoms	Chronic pelvic pain and spotting after coitus. Both symptoms were present since past 6 months.	Patient was asymptomatic.
Ultrasound diagnosis	There are well-defined, thin walled, anechoic, cystic lesions with internal septations seen in bilateral adnexa. No evidence of any debris or	There is evidence of anechoic, thin walled, tubular structures/ lesions, with few incomplete septae which are seen in both adnexa, right larger than left. However, its relation with ovaries could not be demonstrated.

	solid component within. Remote possibility of hydrosalpinx.	Sonography differentials given were: 1. Hydrosalpinx 2. Collections in the pelvic cavity.
MRI imaging findings	• Presence of unusually large lobulated, thin walled predominantly T2 hyperintense cysts extending along the sacral nerve roots, bilaterally into the pelvis. • These are hypointense on T1 weighted images. • No evidence of diffusion restriction. • No significant post contrast enhancement. • MR delineates the ovaries separately.	• Presence of large lobulated, thin walled predominantly lobulated T2 hyperintense cysts extending along the S4 sacral nerve roots, bilaterally into the pelvis. • The cysts are hypointense on T1 weighted images without any significant post contrast enhancement. • No evidence of diffusion restriction. • In addition, in this patient MR sagittal T2 weighted images through whole spine were obtained, which show few small perineural cysts in upper and lower dorsal nerve roots. • MR delineates the ovaries separately.
Largest cyst size (transverse diameter in cm)	Left sided - 2.7cm Right sided- 2.3 cm	Left sided- 1.2cm Right sided- 2.1cm
Treatment given	Symptomatic treatment only	As the patient was asymptomatic no treatment was given. Patient was only scheduled for routine follow ups.

Case I

In our first case, a 46-year-old female, had chief complaints of chronic pelvic pain and spotting after coitus in the last 6 months. She had history of fever, neurological symptoms or per vaginal discharge.

Cervical erosion and other cervical pathologies were ruled out by clinical and PV examination, following which a transabdominal and transvaginal ultrasound was performed.

The ultrasonography (Case 1, Image I) revealed a multicystic and septate lesion on the left side was noted. Differentials of ovarian mass and hydrosalpinx were made. Since the lesion seemed septate and complex tumour markers, namely CA-125 and CEA (carcinoembryonic antigen) were also evaluated. However, the results for the same came back negative.

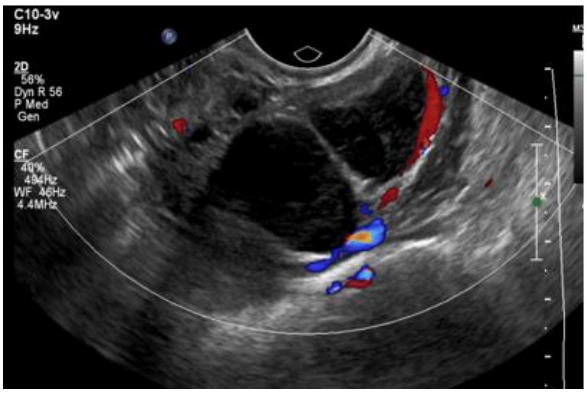
Furthermore, an MRI of the pelvis with contrast (Case 1, Image II) was done as the next diagnostic procedure. On the MRI it was found that there was a presence of large lobulated, thin walled predominantly lobulated T2 hyperintense cysts extending along the S4 sacral nerve roots, bilaterally into the pelvis. The cysts are hypointense on T1 weighted images without any significant post contrast enhancement.

No evidence of diffusion restriction.

In addition, in this patient MR sagittal T2 weighted images through whole spine were obtained, which show few small perineural cysts in upper and lower dorsal nerve roots.

The patient was referred to the gynaecology department where she was explained the various conservative and surgical approaches for managing her symptoms.

She chose to go ahead with medical management with NSAIDS.



Case I:

Image I: Trans Vaginal Ultrasound: - it reveals lobulated anechoic cystic lesions with internal septations seen in bilateral adnexa. Ovaries are seen adjacent to this lesion. No vascularity or any solid component seen.

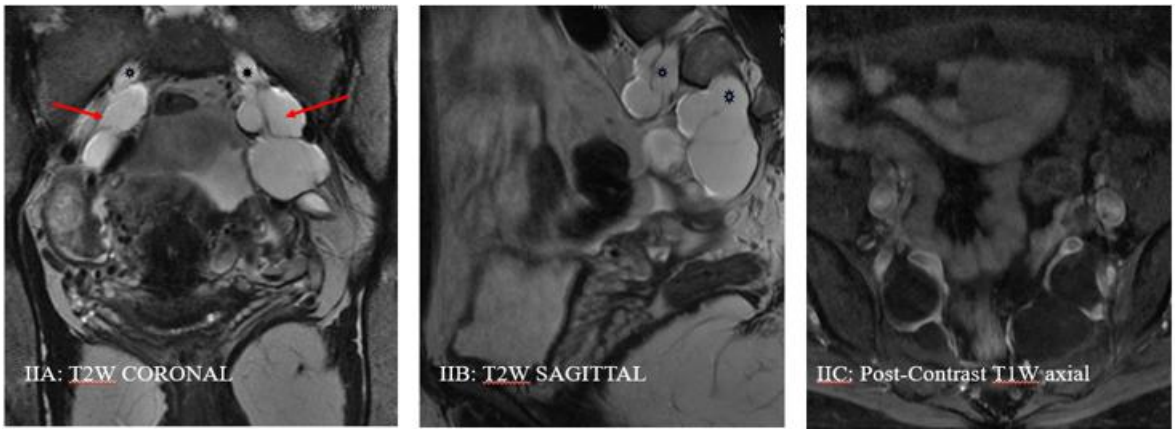


Image II: MRI Pelvis P + C (IIA, IIB, IIC): Large elongated, thin walled peripherally enhancing, predominantly T2 hyperintense and T1 hyperintense cysts, extending along the sacral nerve roots through sacral foramina bilaterally into the pelvis. No Solid component or partial septae

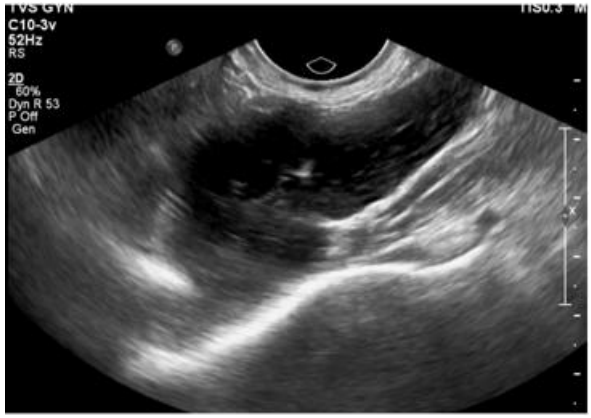
Case II

The second case, a 48-year-old female had come to the hospital for a routine health checkup.

As part of gynecological aspect of checkup an ultrasound was performed during which it was revealed to show presence of elongated anechoic cystic lesions with multiple internal septations in both adnexa, right larger than left. Ovaries were seen separately. No vascularity or any solid component seen within the lesions on either side.

As there was a positive finding on ultrasound an MRI of the pelvis along with contrast was done. It revealed two large elongated, thin walled peripherally enhancing, predominantly T2 hyperintense and T1 hypointense cystic lesions, extending along the sacral nerve roots through sacral foramina bilaterally into the pelvis, right more than left. No solid component or partial septae.

Since the patient had no symptoms, she did not opt for any form of treatment and agreed for routine follow ups to assess the cyst size and visit again if new complaints or symptoms appear.



Case 2:

Image I: Trans Vaginal Ultrasound:
It reveals elongated anechoic cystic area with partial internal septations seen in both adnexa, right more than left. Ovaries are seen separately. No Vascularity or any solid component seen within the lesions

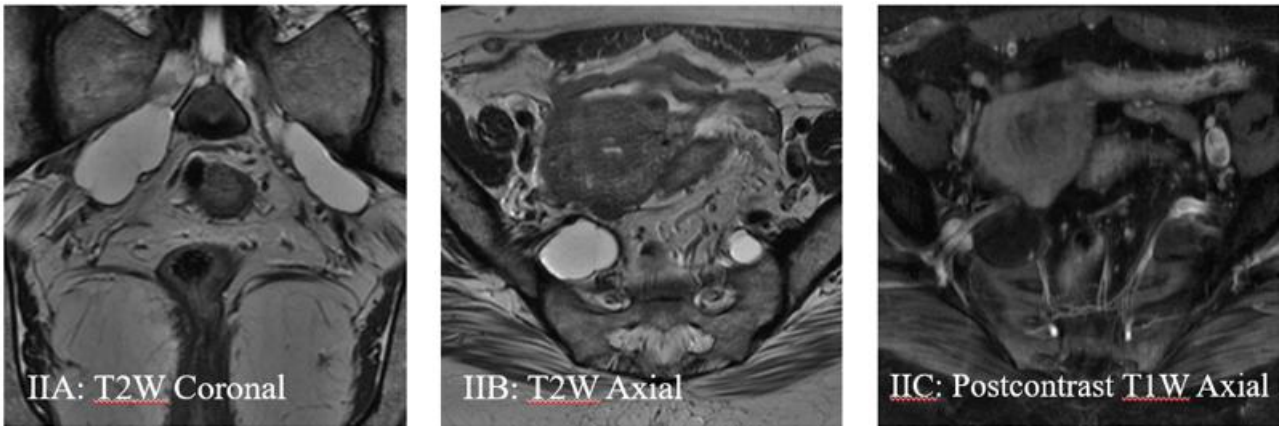


Image II: MRI Pelvis P + C (IIA, IIB, IIC): Large elongated, thin walled peripherally enhancing, predominantly T2 hyperintense and T1 hyperintense cystic lesions, extending along the sacral nerve roots through sacral foramina bilaterally into the pelvis, right more than left. No Solid component or partial septae

3. Discussion

Epidemiology:

Tarlov Cysts (TC's) are more common with women than in men with a 2:1 ratio [15,16] and are usually found in individuals with an average age of 40.4 +/- 14.3 years [15,17]. They are symptomatic in about 1% of the cases [2].

Pathophysiology:

The ball valve mechanism is where the TC's have micro-connections with the subarachnoid space and as CSF continuously fills the cavity of these cysts, it causes them to increase in size and compress the surrounding nerve fibres and resultant neurological symptoms set in [18].

4. Signs and Symptoms

The most common symptoms associated with such cysts are lower back, pelvic, perineal, sacral or lumbar pain or even present with sciatica [2]. Nonspecific symptoms include abdominal pain and headache [19,20,21]. 10% cases also show bladder, bowel or sexual dysfunction[17] and very rarely lead to cauda equina syndrome [22].

Most TCs are small, multiple and are asymptomatic and maybe detected incidentally on pelvic sonography or pelvic MRI. Large TCs are defined as those which are 1.5 cm or more in diameter. These large TCs are rare and can lead to widening of neural foramina and bony changes.[2]

Sonographically, the cysts have internal echoes[23,24] and appear slightly beaded and elongated posteriorly. These characteristics that help differentiate TCs from adnexal cysts are their more posterior location, bilateral presentation and lack of movement with respiration as they are connected to the extra dural space of the spinal canal and not the pelvic floor[25]. Similar features can be seen with abscesses, hematomas and lymph nodes[19,24].

The diagnosis maybe confirmed using CT scan, MRI or myelography, however MRI is the modality of choice as it gives good tissue resolution, multiplanar reconstruction and

absence of bony interphase[20,21]. As the cysts are CSF filled, they show a low signal on T1 weighted and high signal on T2 weighted MRI[26]. An enhancement of the cyst in post contrast images could suggest a giant cystic sacral schwannoma or neurofibroma that can rarely mimic a TC[27]. The presence of internal thread-like structures/ septations or debris on transvaginal sonography and corresponding heterogenous T2 weighted MRI signal may represent the blood or non-blood related cells and support the traumatic cause of TCs[24].

It should be emphasized that dedicated sacral MRI gives superior information compared to lumbosacral MRI. Also, 3D T2 acquisition which allows multiplanar reformation can give maximum information regarding the cysts and their relationship which is useful is surgical treatment is planned.

It is believed that myelography can be used to distinguish TCs from dural ectasias and arachnoid cysts[17].

There has been no single best modality of treatment of TCs yet[18]. Asymptomatic TCs usually are not treated[28].

TCs that are primarily causing symptoms in the patient after ruling out all other causes should be treated[18].

The primary step in managing such patients is with medical therapy in the form of pain-relieving medications like non-steroidal anti-inflammatory or neuropathic pain medicines[29]. Oral steroids[18] or injection of steroids into the epidural region has also helped manage pain due to TCs[30].

Another conservative approach is pelvic physiotherapy which can help manage pain[29].

Surgical treatment is provided for large cysts (1.5cm or more) or symptomatic cysts. Tarlov had suggested complete cyst excision along with its corresponding nerve root and ganglion to treat symptomatic cysts[31].

Interventional options include CT guided interventions like cyst aspiration can be a temporary form of management as CSF can re-accumulate. Another option being CT guided injection of fibrin-glue promotes cyst contracture[18].

Surgery is undertaken for those patients where interventional procedures did not improve quality of life.

Cyst to subarachnoid space, cyst to peritoneal and lumbo-peritoneal shunts, decompressive laminectomy, resecting the cyst neck or cyst wall, using a bipolar cautery to reduce cyst size and curbing the fistula inlet are the various invasive options available[19] with due risks of transitory aseptic meningitis, fat embolisms, CSF leakage, positional headaches and nerve root affection causing sensory and motor deficits in the patient[29].

5. Conclusion

The reported case is a very uncommon presentation of TCs owing to 3 features: bilateral, almost symmetric large TCs and extending from L5 to S3 roots, with all these extending into the pelvis. Further-more very limited or no bone erosion was observed and few clinical symptoms were present. This imaging presentation mimicked adnexal mass on pelvic ultrasound examination. TCs must be considered in the differential diagnosis of such lesions, as well as giant sacral cystic schwannoma which must be included in the differential diagnosis of giant presacral TCs.

References

[1] Kim S, Lee Hj, Park JH, Kim T and Nam K (2020) Tarlov Cysts Misdiagnosed as Adnexal Masses in Pelvic Sonography: A Literature Review. Front. Med. 7:577301. doi: 10.3389/fmed.2020.577301
[2] Boukobza M, Roussel A, Fernandez-Rodriguez P, Laissy JP. Giant multiple and bilateral presacral Tarlov cysts mimicking adnexal mass – imaging features. Int Med Case Rep J. (2018) 11:181–4. doi: 10.2147/IMCRJ.S147791
[3] Park HJ, Kim IS, Lee SW, Son BC. Two cases of symptomatic perineural cysts (Tarlov cysts) in one family – a case report. J Korean Neurosurg Soc 2008; 44: 174–178.
[4] Hirst JE, Torode H, Sears W, Cousins MJ. Beware the Tarlov cyst. J Minim Invasive Gynecol 2009; 16: 78–80.
[5] Tarlov I: Perineurial cysts of the spinal nerve roots. Arch Neurol Psychiatry 1938;40(6):1067–1074.
[6] Tarlov IM. Spinal perineurial and meningeal cysts. J Neural Neurosurg Psychiatry 1970;33:833-43.
[7] Schreiber F, Haddad B. Lumbar and sacral cysts causing pain. J Neurosurg 1951;8:504-9.
[8] Fortuna A, La Torre E, Ciappetta P. Arachnoid diverticula: A unitary approach to spinal cysts communicating with the subarachnoid space. Acta Neurochir (Wien) 1977;39:259-68.
[9] Maleci A, Bianco F, Onnis G, Di Lorenzo N. Iatrogenic spinopelvic cerebro-spinal fluid fistula. Case report. J Neurosurg Sci. (1995) 39:261–3.
[10] Taveras JM, Wood EH. Diagnostic neuroradiology. 2nd ed. Vol 2. Williams and Wilkins: Baltimore; 1976. p.1139-45.
[11] Tabas JH, Deeb ZL. Diagnosis of sacral perineural cysts by computed tomography. J Comput Tomogr 1986;10:255-9.
[12] Rodziewicz GS, Kaufman B, Spetzler RF. Diagnosis of sacral perineural cysts by nuclear magnetic resonance. Surg Neurol 1984;22:50-2.
[13] Nishiura I, Koyama T, Handa J. Intrasacral perineurial cyst. Surg Neurol 1985; 23:265-9.
[14] Sun JJ, Wang ZY, Teo M, et al. Comparative outcomes of the two types of sacral extradural spinal meningeal cysts using different operation methods: a prospective clinical study. PLoS One. 2013;8(12): e83964.
[15] Kuhn FP, Hammoud S, Lefèvre-Colau MM, Poiraudeau S, Feydy A. Prevalence of simple and complex sacral perineural Tarlov cysts in a French cohort of adults and children. J Neuroradiol. 2017;44(1):38–43.
[16] Murphy K, Oaklander AL, Elias G, Kathuria S, Long DM. Treatment of 213 patients with symptomatic Tarlov cysts by CT-guided percutaneous injection of fibrin sealant. AJNR Am J Neuroradiol. 2016;37(2):373–379.

[17] Lucantoni C, Than KD, Wang AC, Valdivia-Valdivia JM, Maher CO, La Marca F, et al. Tarlov cysts: a controversial lesion of the sacral spine. Neurosurg Focus. (2011) 31: E14. doi: 10.3171/2011.9.FOCUS 11221.
[18] Ishii K, Yuzurihara M, Asamoto S, Doi H, Kubota M. A huge presacral Tarlov cyst. Case report. J Neurosurg Spine. 2007 Aug;7(2):259-63. doi: 10.3171/SPI-07/08/259. PMID: 17688070
[19] Slipman CW, Bhat AL, Bhagia SM, Isaac Z, Gilchrist RV, Lenrow DA. Abdominal pain secondary to a sacral perineural cyst. Spine J 2003; 3: 317–320.
[20] Caspar W, Papavero L, Nabhan A, Loew C, Ahlhelm F. Micro- surgical excision of symptomatic sacral perineural cysts: a study of 15 cases. Surg Neurol 2003; 59: 101–106.
[21] Baker JF, Fitzgerald CW, O'Neill SC, McCormack D. Cauda equine syndrome secondary to bilateral sacral Tarlov cysts. Spine J. 2014;14(6):1065–1066.
[22] Forsnes EV, Segna RA. Dural ectasia presenting as an adnexal mass. Obstet Gynecol 1996; 88: 712-713.
[23] McClure MJ, Atri M, Haider MA, Murphy J. Perineural cysts presenting as complex cystic adnexal masses on transvaginal sonography. AJR Am J Roentgenol 2001; 177: 1313-1318.
[24] H'ng MW, Wanigasiri UI, Ong CL. Perineural (Tarlov) cysts mimicking adnexal masses: a report of three cases. *Ultrasound Obstet Gynecol*. 2009;34(2):230–233.
[25] Rodziewicz GS, Kaufman B, Spetzler RF. Diagnosis of sacral perineural cysts by nuclear magnetic resonance. Surg Neurol 1984; 22:50-2.
[26] Attiah MA, Syre PP, Pierce J, Belyaeva E, Welch WC. Giant cystic sacral schwannoma mimicking Tarlov cyst: a case report. *Eur Spine J*. 2016;25(Suppl 1):84–88.
[27] Acosta FL Jr, Quinones-Hinojosa A, Schmidt MH, Weinstein PR. Diagnosis and management of sacral Tarlov cysts. Case report and review of the literature. Neurosurg Focus. (2003) 15: E15. doi: 10.3171/foc.2003.15.2.15
[28] Elkins N, Hunt J, Scott KM. Neurogenic pelvic pain. Phys Med Rehabil Clin N Am. (2017) 28:551–69. doi: 10.1016/j.pmr.2017.03.007.
[29] Mitra R, Kirpalani D, Wedemeyer M. Conservative management of perineural cysts. Spine. (2008) 33:E565–8. doi: 10.1097/BRS.0b013e31817e2cc9
[30] Elsawaf A, Awad TE, Fesal SS. Surgical excision of symptomatic sacral perineurial Tarlov cyst: case series and review of the literature. Eur Spine J. (2016) 25:3385–92. doi: 10.1007/s00586-016-4584-3