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REVIEW ARTICLE

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# An Uncommon Cause of Chest Pain from Tarlov Cysts

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## ABSTRACT

Tarlov cysts are often regarded as an incidental finding found on imaging as most patients are asymptomatic. However, Tarlov cysts can be symptomatic or increase in size over time to cause symptoms. We present a case of an uncommon cause of chest pain in a patient with Marfan syndrome who underwent extensive investigations before being diagnosed with radicular chest pain from Tarlov cysts at the thoracic level. Decompression of Tarlov cysts resulted in resolution of his chronic devastating chest pain.

## ARTICLE HISTORY

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## KEYWORDS

Tarlov cysts, Marfan syndrome, Chest pain.

## Introduction

Tarlov cysts are defined as cysts formed within the nerve-root sheath at the dorsal root ganglion, which are filled with cerebrospinal fluid [1]. The cysts are most frequently found in the spinal canal of the sacral spine, although they could be located anywhere within the spine [2]. Tarlov cysts are usually found incidentally on MRI and patients are often asymptomatic [3]. However, in some patients, Tarlov cysts can cause radicular pain or neurological deficits from local pressure effects and nerve root compression.

In our case, we describe a patient with Marfan syndrome who suffered from chronic chest pain due to symptomatic Tarlov cysts causing radicular chest pain. A 34 year-old teacher presented with 3-year history of chronic chest pain. He was diagnosed with Marfan syndrome at the age of 12. This was confirmed by genetic testing, in addition to the presence of marfanoid features including arachnodactyly and lens subluxation. He did not suffer from any other comorbidity and regularly had transthoracic echocardiogram for aneurysm surveillance over the years. He smoked 10 cigarettes a day for the last 15 years and drank 2 standard drinks of alcohol daily but had never used illicit drugs. He was first referred to a cardiologist for investigation of chest pain. Cardiac examination and ECGs were persistently normal. He could not tolerate treadmill test as chest pain occurred with acceleration for the heart rates above 100bpm. However, there was no associated hypotension or shortness of breath. His transthoracic echocardiogram showed normal size and function of the left and right ventricles, there were no wall motion abnormalities or valvular abnormalities, mean pulmonary artery pressure was normal. He had repeated serial cardiac troponin, NT-proBNP and D-dimer which were all negative. CTPA ruled out pulmonary embolism and aneurysm. CT coronary angiogram did not show any obstructive coronary disease. His chest pain was assessed as non-cardiac chest pain. He was treated with Paracetamol and NSAIDs with no relief. His chest pain became progressively worse in intensity, occurring both at rest and on exertion. There were no associated orthopnoea or paroxysmal nocturnal dyspnoea. A cardiac MRI with contrast was performed which ruled out pericardial or myocardial disease. He was referred to a Respiratory physician for further investigation of chest pain. Pulmonary function tests showed mild obstructive airway disease with normal DLCO and KCO. V/Q scan did not show any ventilation perfusion mismatch. Following that, he was referred to a rheumatologist to investigate for rheumatological cause of chest pain. Repeated blood tests including FBC, EUC, LFT, CRP, ESR were normal, autoimmune screen including ANA, DNA, C3, C4, RF, Anti CCP and ENA were all negative. He was prescribed Gabapentin in addition to NSAIDs with no improvement in his chest pain.

At the patient's request, he was re-referred to the cardiologist. Coronary angiogram was performed which did not show any obstructive coronary artery disease. He was prescribed Isosorbide Mononitrate and Beta Blocker for presumed microvascular angina but again, there was no response to treatment. His chest pain continued to increase in severity and persisted at all times except during sleep. Given all the negative findings despite extensive investigations, patient was referred for psychiatric assessment. He was diagnosed with functional chest pain causing significant anxiety, he was started on anxiolytics with no improvement in chest pain.

Subsequently, his GP referred him to a neurologist to investigate for post herpetic neuralgia as potential cause of chest pain. Neurological examination was unremarkable. Based on VZV serology and PCR, recent herpetic neuralgia was ruled out. Other investigations to look for TB, sarcoidosis and syphilis

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including interferon gamma assay, serum calcium, ACE and RPR were also negative. He then had an MRI of cervical and thoracic spine, which revealed perineural cysts involving T6, T7 and T8, confirmed by radiologist to be Tarlov cysts with fluid collection in the dorsal root ganglia causing compression of the nerve roots and neuronal cell bodies.

The patient was diagnosed by the neurologist with radicular chest pain, corresponding to the location of Tarlov cysts at the level of T6 to T8. He was referred to a neurosurgeon for decompression of Tarlov cysts as a last resort. He underwent fluoroscopy guided decompression whereby fluid was aspirated from the cysts and filled with fibrin sealant to prevent refilling. He had immediate and complete relief from his chronic devastating chest pain following decompression of the cysts.

## Discussion

Tarlov cyts, also known as perineural cysts were first described by American neurosurgeon Isadore Tarlov in 1938 as an incidental finding during autopsy [4]. In a meta-analysis of 132666 subjects with Tarlov cysts conducted in 2021, it showed global prevalence of 4.2%, with female predominance (5.8%) vs male (3%) [5]. Tarlov cysts have also been associated with connective tissue disorders. A case series by Tracz et al. in 2023 identified 220 patients with Tarlov cysts. 21% of patients had underlying connective tissue disorders, most commonly Ehlers-Danlos syndrome (6.4%) and Marfan syndrome (3.2%) [6]. Our literature review showed more cases of symptomatic Tarlov cysts in patients with Marfan's syndrome [7-11]. The identification of nerve fibres within the wall of the cyst is pathognomonic for Tarlov cysts [4]. A few theories existed for the pathogenesis of Tarlov cysts. Tarlov proposed that trauma caused subarachnoid haemorrhage which impeded venous drainage in the perineurium and epineurium, leading to rupture and cyst formation [12].

According to Fortuna et al., perineural cysts were congenital in origin, caused by arachnoid proliferations within the root sleeve [13]. The hydrostatic forces exerted on a one way valve mechanism permit CSF inflow into the cysts and produce pulsative waves, which causes the cysts to expand in size [14]. Although symptomatic Tarlov cysts are rare with reported incidence of approximately 1%, [15] some cysts can enlarge over time and cause compression to the motor axons in the ventral roots [16]. They can also erode surrounding bony tissues, causing irritation of periosteal pain fibres [17]. Most commonly reported in the sacral spine, Tarlov cysts in the lumbosacral region can cause lower back pain, sciatica from lumbosacral nerve root involvement, leg weakness and paraesthesia, cauda equina syndrome, neurogenic claudication, and bladder dysfunction. Rarely, Tarlov cysts occurring in the cervical and thoracic region can cause radicular pain in the neck, chest, arms and hands, as well as upper limb weakness and paraesthesia. In a clinical report by Hulens et al., [18] using electromyography to assess symptomatic Tarlov cysts, it was documented that even smaller cysts of less than 1 cm can cause nerve damage. Due to limited knowledge about Tarlov cysts, they are often regarded as incidental finding that lacks clinical significance. Moreover, some radiologists omit commenting on Tarlov cysts in their report, which makes it more difficult to diagnose. MRI is the imaging modality of choice to diagnose Tarlov cysts as it provides significantly greater soft tissue contrast [19].


Case courtesy of Domenico Nicoletti, Radiopaedia.org, rID: 71492
This is a Sagittal T2 view of an MRI showing a large Tarlov cyst occupying the spinal canal from L5 to S3-S4 measuring 8 x 3cm in diameter.
Please note this is not the MRI image of our patient.

Management of Tarlov cysts ranges from conservative management, minimally invasive interventions to surgery. Conservative management include analgesia and physical therapy. Image-guided aspiration of symptomatic Tarlov cysts and injection of fibrin sealant into the cysts using the two-needle technique has been proven to be an effective, safe and less-invasive alternative to the open surgical treatment of these cysts [20]. In our case, this was the method of treatment. There is no consensus on the optimal surgical method but surgical treatment strategies conceptually involve either diversion of CSF flow or a direct microsurgical approach [17]. They carry significantly higher complication rates, longer hospital stays, and recovery compared to percutaneous aspiration-fibrin sealant interventions [17].

In summary, we presented a case of Tarlov cysts causing radicular chest pain in a gentleman with Marfan Syndrome. This patient was seen by multiple specialists and underwent extensive investigations over the span of 3 years before the diagnosis was eventually made and decompression of the cysts resulted in resolution of his chest pain. We hope this case raises clinician awareness of symptomatic Tarlov cysts and the treatment available. Although most Tarlov cysts are asymptomatic, there are increasing evidence that Tarlov cysts can potentially cause clinically debilitating symptoms. It is also worth noting that asymptomatic Tarlov cysts can increase in size over time and become symptomatic. They should be considered in the differential diagnosis of patients presenting with radicular chest pain, when the common causes of chest pain have been excluded.

## References


1. Goyal RN, Russell NA, Benoit BG, Belanger JM. Intraspinal cysts: a classification and literature review. Spine (Phila Pa 1976). 1987; 12: 209-213.
2. Guo D, Shu K, Chen R, Ke C, Zhu Y, et al. Microsurgical treatment of symptomatic sacral perineurial cysts. Neurosurgery. 2007; 60: 1059-1065.
3. Lucantoni C, Than KD, Wang AC, Valdivia-Valdivia JM, Maher CO, et al. Tarlov cysts: a controversial lesion of the sacral spine. Neurosurg Focus. 2011; 31: E14.
4. Tarlov I. Perineurial cysts of the spinal nerve roots. Archives of Neurology & Psychiatry. 1938; 40: 1067-1074.
5. Klepinowski T, Orbik W, Sagan L. Global incidence of spinal perineural Tarlov's cysts and their morphological characteristics: a meta-analysis of 13,266 subjects. Surg Radiol Anat. 2021; 43: 855-863.
6. Tracz J, Judy BF, Jiang KJ, Caraway CA, Yang W, et al. Interventional approaches to symptomatic Tarlov cysts: a 15-year institutional experience. J Neurointerv Surg. 2023.
7. Robinson L, Dominguez R, Cabrera J, Yeakley JW, Fenstermacher MJ, et al. Multiple meningeal cysts in Marfan syndrome. AJNR Am J Neuroradiol. 1989; 10: 1275-1276.
8. Wang B, Moon SJ, Olivero WC, Wang H. Pelvic pain from a giant presacral Tarlov cyst successfully obliterated using aneurysm clips in a patient with Marfan syndrome. J Neurosurg Spine. 2014; 21: 833-836.
9. Paterakis K, Brotis A, Bakopoulou M, Rountas C, Dardiotis E, et al. A Giant Tarlov Cyst Presenting with Hydronephrosis in a Patient with Marfan Syndrome: A Case Report and Review of the Literature. World Neurosurg. 2019; 126: 581-617.
10. Paisan GM, Crandall KM, Chen S, Burks SS, Sands LR, et al. Closure of a giant anterior sacral meningocele with an omental flap in a patient with Marfan syndrome: case report. J Neurosurg Spine. 2018; 29: 182-186.
11. Arnold PM, Teuber J. Marfan syndrome and symptomatic sacral cyst: report of two cases. J Spinal Cord Med. 2013; 36: 499-503.
12. Tarlov IM. Spinal perineurial and meningeal cysts. J Neurol Neurosurg Psychiatry. 1970; 33: 833-843.
13. 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: 259-268.
14. Mummaneni PV, Pitts LH, McCormack BM, Corroo JM, Weinstein PR. Microsurgical treatment of symptomatic sacral Tarlov cysts. Neurosurgery. 2000; 47: 74-78.
15. Langdown AJ, Grundy JR, Birch NC. The clinical relevance of Tarlov cysts. J Spinal Disord Tech. 2005; 18: 29-33.
16. Hulens M, Bruyninckx F, Dankaerts W, Vansant G, De Mulder PA. Electromyographic Abnormalities Associated with Symptomatic Sacral Tarlov Cysts. Pain Pract. 2016; 16: 81-88.
17. Murphy K, Nasralla M, Pron G, Almohaimede K, Schievink W. Management of Tarlov cysts: an uncommon but potentially serious spinal column disease-review of the literature and experience with over 1000 referrals. Neuroradiology. 2024; 66: 1-30.
18. Hulens M, Rasschaert R, Bruyninckx F, Dankaerts W, Stalmans I, et al. Symptomatic Tarlov cysts are often overlooked: ten reasons why-a narrative review. Eur Spine J. 2019; 28: 2237-2248.
19. Shoyab M. Tarlov cysts in back pain patients: prevalence, measurement method and reporting points. Br J Radiol. 2021; 94.
20. Murphy KP, Ryan S. Shrinking of a Tarlov cyst. BMJ Case Rep. 2019; 12.