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NEUROLOGIC AND MYOFASCIAL DISORDERS MIMICKING COCCYGODYNIA

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INTRODUCTION

The «idiopathic» nature of coccygodynia in up to one-third of affected patients should prompt to search for still overlooked explanations, including occult neurologic disorders, either in spinal cord, cauda-equina, sacral roots, or small branches of the coccygeal plexus (1).

NEUROLOGIC DISORDERS

Demyelinating disease

Pelvic pain as the presenting symptom of demyelinating disease is rare. However, several cases of pain around the coccyx heralding neuromyelitis optica (Devic's disease) have been reported (2). Hypoesthesia or anesthesia in the perineum is not a feature of coccygodynia, and their description should suggest other diagnoses, including this unusual aetiology. Devic's disease is a group demyelinating syndromes, now considered distinct from multiple sclerosis, which primarily affects the spinal cord and optic nerves, and can be diagnosed on enhancing spinal cord lesions on MRI but also on some serological features. Indeed, the identification of specific antibodies directed to aquaporin 4 (AQP4-IgG) led to the distinction from multiple sclerosis, although up to 25% of the clinically diagnosed NMO patients are seronegative for AQP4-IgG which can conversely also be found in other conditions (including systemic lupus erythematosus). A subgroup of patients with Devic's syndrome negative forAQP4-IgG might be identified by antibodies directed to myelin oligodendrocyte glycoprotein (MOG-IgG) (3).

Dysesthetic paroxysmal pain of the pelvic level has also been reported as an early sign of mutliple sclerosis (4). Although this possibility has been only exceptionally described, it might not be so infrequent. In one the authors' experience, two unpublished cases of young women with unexplained transient (several weeks or months) coccygodynia developed multiple sclerosis one and four year after the first onset of their coccygeal pain.

Schwannoma

Some atypical and slowly worsening coccygodynia can reveal small schwannoma of the distal cauda equina (5). As those schwannomas develop in sacral nerve roots, coccygodynia is often associated with visceral pain, like intractable vulvodynia in women (6). Laparoscopic en bloc resection of the sacral schwannomas can be performed, with sparing of the sacral nerve roots and without reported recurrence or worsening of symptoms. Compared with classic neurosurgical approaches, laparoscopic exposure of the rectum, ureters, and sacral nerve roots might render the procedure safer and easier, with less risk of postoperative functional morbidity (6).

Other neurogenic tumours

Extra-adrenal paragangliomas of the spine usually present as intradural tumours in the cauda equina, but they may also present as atypical coccygodynia when they affect the sacral canal (7). The majority of those spinal paragangliomas are benign, slowly growing tumours with low proliferative activity, but local recurrence has been reported. As postoperative radiation therapy for patients with incomplete excision may not prevent recurrence, gross tumour removal should be the goal of surgery (7).

Epidural meningioma of the sacral canal presenting as coccygeal and left-sided S2-4 dermatomal pain has also been reported (8). Sensory loss and enhancing mass lesion in the sacral canal scalloping the bone between S-2 and S-4 help not to misdiagnosis patients as common coccygodynia, which is mandatory since those tumours can behave aggressively, with recurrence and event metastases despite surgery (8).

Ependymoma of the filum terminale (9) or giant tumour of cauda equina (10) can also be revealed by coccygodynia (9). The filum terminale is considered a fibrous structure that extends from conus medullaris of the spinal cord to coccyx, but dissection of stillborn cadavers or adult patients undergoing sectioning of the filum terminale in an untethering surgical procedure, showed that in roughly a third of cases there were gross anatomical connections between the filum terminale and nerve roots that contain nerve fibers (11). Similar deductions were made when studying the embryogenesis of filum terminale in foetal specimens (12). However tethered cord has not yet been described as a source of coccygodynia.

Tarlov cysts and arachnoiditis

Severe chronic coccygodynia can be related to arachnoid cyst of cauda equina, as demonstrated by the observation that resection of the cyst abolished coccygodynia completely and permanently (13-14).

However, this diagnosis remains difficult, since most arachnoid cysts are asymptomatic, and imaging cannot certify that the cyst is indeed responsible for the coccygodynia. The association of incomplete cauda equina syndrome is highly suggestive of a relationship. Other clinical features (paroxysmal pain exacerbated in standing position, relieved in dorsal position and revived by ramming the sacrum) are less reliable (15). Intradural injections of corticosteroids provided lasting pain relief in some patients (15), but are no longer advised nowadays. Various surgical strategies, including posterior decompression, cyst wall resection, CT-guided needle aspiration with intra-lesional fibrin injection, and shunting have been advocated (16-Lucantoni). However, in operative patients, the rates of short-term and long-term improvement in clinical symptoms are not clear. It has also been stressed that, although neurological deficit frequently improves, pain is less likely to do so (16).

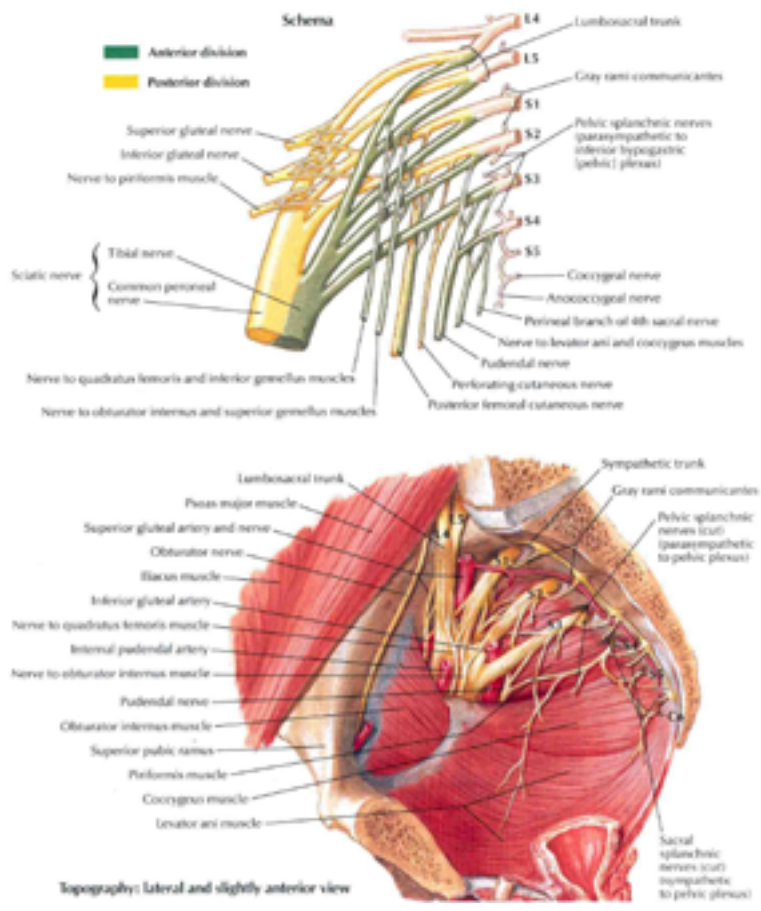
The decision to operate or not depends on the persistence and intensity of pain, and on whether signs of neurological deficit are present (15). When several sacral nerve root cysts are observed on MRI/myelography the diagnosis should not be ruled out because of the discrepancy between the localized unilateral perineal pain and the multiple cysts observed on both sides. Indeed, only one nerve can be constricted or adherent to a cyst wall, as observed during surgery in some reports where only one root was constricted by hyperplasia of the dura mater, and adhered to the cyst wall, whereas other nerves were not (17).

Arachnoiditis can also reveal by atypical coccygodynia (18), but other features are present in most patients, like atypical sciatica.

Neuropathic change in the Trolard nerve, and distal entrapment neuropathies of the sacral plexus.

The coccygeal plexus is formed within ischiococcygeus from the ventral rami of S4, S5, and Co1 with a contribution (gray rami communicantes) from the sacral sympathetic trunk. It gives rise to anococcygeal nerves which pierce ischiococcygeus and the sacrospinous ligament to supply the subcutaneous tissue on the dorsal aspect of the coccyx. Some branches from the plexus pass medially anterior to the coccyx. The coccycgeal plexus is formed within ischiococcygeus rather than on its pelvic surface and appears to supply skin in the anococcygeal region. It probably also contributes to the innervation of ischiococcygeus, the sacrospinous ligament, coccygeal ligaments, and periosteum (1). In rats there is evidence for nerve fibers in the discs and predominant innervation by CGRP neurons. The neurons innervating the discs mostly derived from S1 to S3 dorsal root ganglion, especially S2 and S3 (19). Thus, coccygeal plexus deserves to be considered as a potential pain generator that may be implicated in some patients with coccydynia (1).

The posterior nerve of the second sacral root mixes with bundles from the third and fourth sacra roots to create a posterior sacral plexus between the sacrum and the overlying spinal muscles. A terminal nerve exiting from this plexus has been called posterior gluteal nerve by Trolard. It runs downwards the lateral border of the sacrum towards the coccyx in a canal located just behind the sacro-tuberal and sacro-epinous ligament and is usually surrounded by a large amount of fat tissue. This Tolard nerve is roughly 10 cm long. It gives some nerve endings to the sacro-coccygeal joint and ends below the skin in the paramedian line of the sacrum, where it divides in two branches: one is ascending towards the second caudal vertebra, but the second goes down and ends in the subcutaneaous fat at the posterior aspect of the coccyx (20). Entrapment neuropathy of this nerve has not yet been specifically described, but suffering of the posterior branch of the S2 to S4 posterior nerve when they cross through the superficial sacroiliac ligaments has been postulated (21) and occurrence of abnormal muscles in the pelvic wall, like persistent sacrococcygeus ventralis muscle, could also theoretically induce chronic striction of S4 and S5 roots (22).



More probably, some lasting allodynia of the coccyx area following falls on the buttocks or similar injuries, not explained otherwise, could be ascribed to neuropathic changes of Trolard nerves. Such cases could be the best indication, if any, for sacral rhizotomy, since this intervention appeared beneficial in previous studies only in patients with pain distribution limited to the S-4 S-5 area (23).

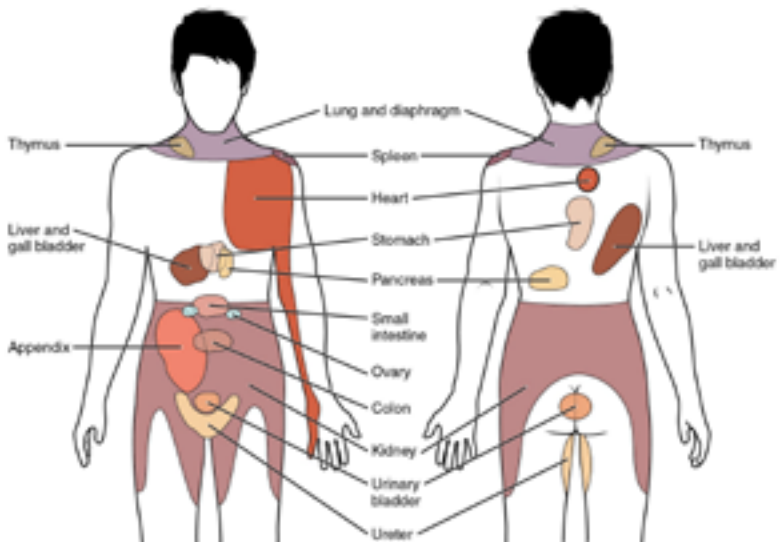
Neuro-vascular entrapment at the infracardinal level of the sacral plexus has also been reported recently as an explanation for some chronic coccygodynia worsened by sitting but not accompanied by neurologic dysfunction (24). This unusual cause of 'coccygodynia' should be suspected in patients with atypical presentation such as a mix of gluteal and coccygeal pain (24). They can be often significantly relieved by laparoscopic decompression and separation of the offending vessel from the nerves (24). It remains rare situations however, since most sacral nerve root lesions do not cause acute pain, and are accompanied by sacral sensory loss and urinary, anorectal or sexual disorders (25).

Small nerve branches have been observed adjacent to the ventral aspect of the intercornual ligament and, in one case, traversing the ligament. Ipsilateral sacral and coccygeal cornua are therefore normally bridged by an intercornual ligament that is probably innervated (26). Small neuromas in this area have not yet been described, but could be an explanation of unexplained coccygodynia occurring without injury, with clinical features suggestive of a neuroma (electric pain). This is even more probable that glomic tumours have been described as a source of coccygodynia (27). Indeed, although most glomus bodies are normal vestigial arteriovenous anastomosis found in 35% of humans, which can mimic glomic tumours (28-29), development of a glomus tumour can also occur in the coccygeal region. It can be the cause of coccygodynia, and long lasting coccygodynia can be relieved after removal of the glomus tumour without coccygectomy (27).

REFERRED PAIN TO THE COCCYGEAL AREA

From disc herniations

Some disc lesions can induce pain projected to the coccyx region (18, 30). Although the L5-S1 levels are mostly observed in our experience, even L1-L2 disc can be the source of the pain, as described in elite tennis player after backhand strokes. In this case, MRI examination showed an injured L1-L2 disc with fluid inside the disc with a signal similar to blood, and palpation over the spinal processes of L1-L2, reproduced the intense pain projected to the coccyx region (31).



From urologic conditions

Although poorly studied, and rarely observed, referred pain in the coccyx area is regularly ascribed to a bladder origin in many textbooks on projection of visceral pain, as illustrated below.

MYOFASCIAL SYNDROMES OF PERINEAL MUSCLES

The most common functional disorders of the pelvic floor muscles, accompanied by perineal pain are levator ani syndrome, proctalgia fugax, and myofascial syndrome with coccygodynia (32). Those syndromes are in fact considered as differential diagnoses of coccygodynia rather than as a source of coccygeal pain (33), although chronic contraction of muscles inserted on the coccyx bone might theoretically also enhance pain arising from the coccyx for other reasons.

Levator ani syndrome is episodic perineal (mostly rectal) pain caused by spasm of the levator ani muscle. Patients complain of dull ache above the anus or higher in the rectum and a feeling of constant rectal pressure or burning. However, the pain may also be felt in the low pelvis or coccygeal region. It may be relieved by walking or pelvic tightening exercises similar to Kegel exercises. The hallmark diagnostic indicator of myofascial pelvic pain is myofascial trigger points in the pelvic floor musculature that refer pain to adjacent sites, including sometimes the coccyx area (34). An interdisciplinary team is essential for identifying and successfully treating those conditions.

CONCLUSION

Coccygodynia as first manifestation of multiple slerosis and Devic's syndrome are certainly very rare event, and neurogenic tumours of the sacral area remain exceptional findings. On the opposite, many patients with so-called 'primitive' coccygodynia might suffer from cryptic suffering of small branches of S3 to S5 roots, either in the spinal canal (Tarlov cyst) or downwards. Entrapment of Trolard nerve, or of the sacral plexus, is indeed probably frequently unrecognized, as well as neuromas in the small nerve branches traversing the ventral aspect of the intercornual ligament.

BIBLIOGRAPHY

1. Woon JT, Stringer MD. Redefining the coccygeal plexus. Clin Anat 2014;27:254-60.
2. Loschner AL, Snyder JE. Pelvic pain as an unusual first presentation of a demyelinating disease. J Gen Intern Med 2008;23:1917-20.
3. Van Pelt ED, Wong YY, Ketelslegers IA, Hamann D, Hintzen RQ. Neuromyelitis optica spectrum disorders: comparison of clinical and magnetic resonance imaging characteristics of AQP4-IgG versus MOG-IgG seropositive cases in the Netherlands. Eur J Neurol 2015 doi: 10.1111/ene.12898.
4. Solaro C, Uccelli MM, Mancardi GL. A patient with multiple sclerosis and Down's syndrome with a rare paroxysmal symptom at onset. Eur J Neurol 1999;6:505-7.
5. Kinnett JG, Root L. An obscure cause of coccygodynia. Case report. J Bone Joint Surg Am 1979;61:299.
6. Possover M, Kostov P. Laparoscopic management of sacral nerve root schwannoma with intractable vulvococcygodynia: report of three cases and review of literature. J Minim Invasive Gynecol 2013;20:394-7.
7. Falavigna A, Righesso O, Volquind D, Salgado KB, Teles AR. Intraosseous sacral paraganglioma with extradural extension: case report. Acta Neurochir (Wien) 2010;152:475-80.
8. Rutherford SA, Linton KM, Durnian JM, Cowie RA. Epidural meningioma of the sacral canal. Case report. J Neurosurg Spine 2006;4:71-4.
9. Charpentier J, Messimy R, da Lage C. Coccygodynia revealing an ependymoma of the filum terminale. Complete removal without sequelae. Rev Neurol 1968;118:160-2.
10. Chateau R, de Rougemont J, Bonneville B, Barge M, Groslambert R, Perret J. Coccygodynia revealing a giant tumour of the cauda equine. J Med Lyon 1967;48:573-8.
11. Gaddam SS, Santhi V, Babu S, Chacko G, Baddukonda RA, Rajishekhar V. Gross and microscopic study of the filum terminale: does the filum contain functional neural elements? J Neurosurg Pediatr 2012;9:86-92.
12. Jang HS, Cho KH, Chang H, Jin ZH, Rodriguez-Vazquez JF, Murakami G. The Filum Terminale Revisited: A Histological Study in Human Fetuses. Pediatr Neurosurg 2015 Nov 24 [Epub ahead of print]
13. Kepski A, Rudnicki S. Arachnoid cyst of the cauda equina: a contribution to the etiology of coccygodynia. Neurol Neurochir Pol 1978;12:109-12.
14. Ziegler DK, Batnitzky S. Coccygodynia caused by perineural cyst. Neurology 1984;34:829-30.
15. Dehaine V, Weschker B, Ziza JM, de Gennes C, Robain G, Fohanno D, Metzger J, Godeau P. Coccygodynia disclosing Tarlov's cysts. Rev Med Interne 1990;11:280-4.
16. Lucantoni C, Than KD, Wang AC, Valdivia-Valdivia JM, Maher CO, La Marca F, Park P. Tarlov cysts: a controversial lesion of the sacral spine. Neurosurg Focus 2011;31:E14.
17. Iwasaki Y, Yamauchi R, Akino M, Isu T, Abe H, Tashiro K, Kenmotsu O. Sacral nerve root cysts manifesting as localized unilateral perineal pain--case report. Neurol Med Chir (Tokyo) 1992;32:96-9.
18. De Andrés J, Chaves S. Coccygodynia: a proposal for an algorithm for treatment. J Pain 2003;4:257-66.
19. Orita S, Ohtori S, Taniquchi A, Yamashita M, Yamauchi K, Inoue G, et al. Direct evidence for sensory innervation of the dorsal portion of the Co5/6 coccygeal intervertebral disc in rats. Spine (Phila Pa 1976) 2010;35:1346-52.
20. Robert R, Salaud C, Hamel O, Hamel A, Philipeau JM. Anatomie des douleurs de l'articulation sacro-iliaque. Anatomy of the sacro-iliac joint pain. Rev Rhum 2009 76:727-33.
21. Le Goff B, Berthelot JM, Maugars Y. Ultrasound assessment of the posterior sacroiliac ligaments. Clin Exp Rheumatol. 2011;29:1014-7. Epub 2011 Dec 22.
22. 22-Nair V, Nair RV, Mookamabika RV, Mohandas Rao KG, Krishnaraja Somayaij S. Persistent sacrococcygeus ventralis muscle in an adult human pelvic wall: a variation for surgeons to note. J Chin Med Assoc 2011;74:567-9.
23. Albrektsson B. Sacral rhizotomy in cases of ano-coccygeal pain. A follow-up of 24 cases. Acta Orthop Scand 1981;52:187-90.
24. Possover M, Forman A. Pelvic Neuralgias by Neuro-Vascular Entrapment: Anatomical Findings in a Series of 97 Consecutive Patients Treated by Laparoscopic Nerve Decompression. Pain Physician 2015;18:E1139-43.
25. Labat JJ, Robert E, Delavierre D, Sibert L, Rigaud J. Symptomatic approach to chronic neuropathic somatic pelvic and perineal pain. Prog Urol 2010;20:973-81.
26. Woon JT, Stringer MD. The anatomy of the sacrococcygeal cornual region and its clinical relevance. Anat Sci Int 2014;89:207-14.

27. Kim HS, Yang SH, Park HJ, Park HB, Cho HS. Glomus tumour as a cause of coccydynia. Skeletal Radiol 2013;42:1471-3.
28. Santos LD, Chow C, Kennerson AR. Glomus coccygeum may mimic glomus tumour. Pathology 2002;34:339-43.
29. Maggiani F, Kashima T, Ostlere SJ, Athanasou NA. Immunophenotypic analysis of glomus coccygeum associated with coccygodynia. Skeletal Radiol 2011;40:1455-9.
30. Nicola N, Thal HU. Coccygeal pain associated with intervertebral disk displacement. Nervenarzt. 1975 Apr;46(4):212-3.
31. Baranto A, Hellström M, Swärd L. Acute injury of an intervertebral disc in an elite tennis player: a case report. Spine (Phila Pa 1976) 2010;35:E223-7.
32. Andromanakos NP, Kouraklis G, Alkiviadis K. Chronic perineal pain: current pathophysiological aspects, diagnostic approaches and treatment. Eur J Gastroenterol Hepatol 2011;23:2-7.
33. Chiarioni G, Asteria C, Whitehead WE. Chronic proctalgia and chronic pelvic pain syndromes: new etiologic insights and treatment options. World J Gastroenterol 2011;17:4447-55.
34. Pastore EA, Katzman WB. Recognizing myofascial pelvic pain in the female patient with chronic pelvic pain. J Obstet Gynecol Neonatal Nurs 2012;41:680-91.