18

Proctalgia Fugax, Levator Spasm, and Pelvic Pain: Evaluation and Differential Diagnosis

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Introduction

Pelvic pain is a common complaint with a wide range of causes involving multiple organ systems, making the diagnosis particularly complicated. The expansive differential can be divided into acute and chronic etiologies. The most common causes of acute pain include thrombosed external hemorrhoids, perianal abscesses, fistulas and anal fissures. Chronic pelvic pain also has a broad differential that includes several pelvic floor syndromes. The differential can be divided into organic or functional etiologies, the later of which relates to diagnoses that do not have a structural or anatomical cause.

Patients may present shortly after the acute pain starts or delay seeing a provider due to anxiety and embarrassment. Many may also ignore the pain altogether. Pelvic pain is especially distressing as it affects a sensitive area of the body that is difficult for patients to examine themselves. Nevertheless, with a systematic approach to the history and physical, the physician can identify most diagnoses and treat them successfully.

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General Approach to the Patient with Pelvic Pain

History

Many patients come with a predetermined diagnosis, such as hemorrhoids or anal fissure, which can be a distractor. It is essential to listen to key descriptions provided by the patient, focusing on pain characteristics such as character, duration, location, causative factors and associated signs and symptoms.

Evaluate the past medical, surgical and gynecological history. A family history may point towards a malignant etiology. A history of inflammatory bowel disease may raise suspicion of anal fistulas or fissure and a history of diabetes or HIV may point to an infectious cause. Sexual history may raise suspicions for sexually transmitted disease, anal dysplasia or cancer. A history of physical, emotional or sexual abuse may underlie certain chronic pelvic pain symptoms.

Physical Examination

While a complete examination is important, high-yield portions include the examination of the abdomen, inguinal region, perianal skin and soft tissue, buttocks and gluteal cleft, anal canal and rectum. The abdominal exam starts with inspection. Scars may reveal a history of

D. E. Beck et al. (eds.), *Fundamentals of Anorectal Surgery*,
https://doi.org/10.1007/978-3-319-65966-4_18

operations or trauma. Palpation helps appreciate distention and tympani in the setting of constipation. The point of maximum tenderness in the left lower quadrant can reveal diverticulitis. Metastases may present with hepatomegaly. The inguinal exam may identify lymphadenopathy, which may raise suspicions of an infectious and neoplastic process. Anal and low rectal cancers can present with palpable lymphadenopathy in metastatic disease [1].

The external and internal anal exams are crucial. This part of the evaluation requires extreme sensitivity on the part of the examiner, as it is both a physically and psychologically sensitive examination event for the examinee. Warn the patient before initiating any portion of the perianal and internal anorectal exam. By putting the patient at ease and earning trust, one can obtain helpful information in a more efficient manner and with minimal discomfort.

Starting with the external exam, visualization of the gluteal, intergluteal and perianal skin can reveal discoloration of the skin, thickened folds or scaling, masses, secondary openings of fistulae, swelling from abscesses, pilonidal pits, skin tags or thrombosis of external hemorrhoids. Gentle distraction of the buttocks may reveal anal fissures. Palpation of the perianal skin can identify abscesses or a mass concerning for a neoplastic process. Assess the size, firmness, fixation and tenderness of any lesions.

The digital rectal exam is best tolerated with adequate lubrication and in the case of pelvic pain, topical anesthetic should be considered to maximize patient tolerance. In the case of a visualized fissure on external exam, it is helpful to gently insert the examining finger to place initial pressure on the contralateral side. Palpate for masses and define the location in regards to anterior, posterior, right or left lateral quadrants. Define the distance of the lesion from the anal verge and anorectal ring. The firmness and fixation of the lesion are also discernable on digital exam and should be recorded.

Assess anal sphincter tone, which is usually hypertonic in most cases of anal fissures, but can also be found to be normal in some patients, which alters the treatment options for fissures. Starting at the coccyx, rotate the finger to each lateral quadrant, feeling for muscle spasm of the levator complex. This should also reproduce pain in the setting of levator syndrome.

Palpate for fluctuance with concomitant pain in the setting of perirectal abscesses. Posteriorly, fullness on palpation can identify presacral masses. Palpation of the coccyx can diagnose coccygodynia. For completion, rotate the finger anteriorly in men to identify point tenderness of the prostate in cases of prostatitis and for rectocele in women with a history of constipation.

If the patient is unable to tolerate the process, an exam under anesthesia should be performed. Repeated and aggressive pain-inducing examinations will not yield helpful information, especially in setting of investigating pelvic pain.

Anoscopy/Rigid Proctoscopy

Anoscopy and sigmoidoscopy allow for visualization of intraanal and rectal lesions. These findings can range from enlarged hemorrhoids to distinct neoplastic lesions to mucosal changes from sexually transmitted infections or inflammatory bowel disease.

Imaging/Testing

There are certain circumstances when diagnostic or confirmatory studies are required after a history and physical exam. This especially occurs when the history is concerning for an anorectal abscess but there are limited physical exam findings. A pelvis computed tomography (CT) may be helpful in this case and may have already been ordered by inexperienced practitioners. Diagnosis of pelvic floor disorders can be visualized with a defecogram or dynamic pelvic magnetic resonance imaging (MRI). Patients with a history concerning for outlet obstruction can be diagnosed with high-resolution anorectal manometry and a balloon expulsion study. A history of a retained foreign body should prompt an abdominal and pelvic radiograph in order to identify some objects, as well as evaluate for free air. Finally, the standard workup for an anal or rectal cancer includes staging with ultrasound or MRI

and CT.Colonoscopy should be added for routine screening or for suspicion of inflammatory or neoplastic processes.

Acute Pelvic Pain

Three of the most common causes of acute pelvic pain include thrombosed external hemorrhoids, anal fissures and anorectal abscesses (Table 18.1). Key components of the history and physical include asking when the pain first occurred and if an inciting event could be identified, such as a bowel movement or certain physical activities. If there is a history of bleeding or drainage, the amount and frequency should be quantified. Oftentimes, the history and physical are sufficient to confirming a diagnosis of most causes of acute pelvic pain.

Thrombosed External Hemorrhoid

Most patients will be able to pinpoint the exact time they developed a thrombosed external hemorrhoid (Fig. 18.1). It often occurs after straining, either with a bowel movement or with lifting heavy objects. It can also occur with diarrhea. The pain is sharp, constant and worse when they touch the area or sit. They may feel a "bulge" near the anus. The pain tends to increase over the first few days then gradually decrease after about one week as the thrombosis naturally resolves. They deny any fever [2, 3]. The exam will reveal a blue or purplish lesion that may be firm and tender or soft and nontender to palpation depending on when the patient presents.

Fig. 18.1 Thrombosed external hemorrhoid. With permission from [57] © 2014 Springer

Fig. 18.2 Circumferential thrombosis of internal and external hemorrhoids. With permission from [57] © 2014 Springer

Table 18.1 Three most common causes of acute pelvic pain

	Pain quality	Findings
Thrombosed external hemorrhoid	Sharp and constant	Enlarged purplish lesion on anal verge, firmness depending on chronicity
Fissure	Sharp, cutting and worse during bowel movements	Visible cut tear in anoderm
Abscess	Throbbing, worse with pressure, fevers/chills	Erythema, fluctuance, induration, active purulent drainage

If the hemorrhoid is symptomatic and firm, office excision and drainage of the clot can provide instant relief. Compared to a simple incision through the skin with enucleation of the clot, excision prevents recurrence at that particular hemorrhoid. When all external hemorrhoids are thrombosed (also known as hemorrhoidal crisis), an exam under anesthesia with excisional hemorrhoidectomy should be performed (see Chap. 17) (Fig. 18.2) [4].

Anal Fissure

Patients typically describe a sharp cutting pain that occurs during and right after passage of a bowel movement [5, 6]. The inciting event may be passage of a hard and constipated stool. The pain can last for minutes to hours after a bowel movement and the quality can change from sharp to a burning sensation. If the pain is very severe, patients may even describe being afraid of having a bowel movement. There may be spotting of blood on the toilet paper after wiping or drops of blood in the toilet water. Some may have tried warm soaks with some relief.

The exam can be especially challenging due to the acuity of pain. Distraction of the buttocks may reveal the fissure (Fig. 18.3). Typical locations include the posterior and anterior midline. Ectopic locations should raise suspicion of underlying infectious or inflammatory etiologies. Acute fissures often appear as a superficial tear of the anoderm. Chronic fissures may reveal exposed internal sphincter muscle fibers at the base with an associated sentinel pile and hypertrophic anal papilla.

If the patient tolerates the digital rectal exam, pain is reproduced with gentle palpation of the fissure and hypertonicity of the sphincter is often appreciated. Normal tone can also be present in some cases, which will alter treatment options for these patients.

Fig. 18.3 Anal fissure with skin tag. With permission from [57] © 2014 Springer

Treatment for typical fissures is initially medical and focuses on relieving the sphincter spasm. Bulking agents and sitz baths provide relaxation of the muscles but in many cases, the addition of a calcium channel blocker (nifedipine or diltiazem) or nitroglycerin-based creams may be required. This has high success but patients should be educated on potential side effects, including hypotension and headache.

The gold standard for treatment of chronic anal fissures is lateral internal sphincterotomy (LIS). The sphincterotomy is performed to the length of the fissure itself. This is performed on the lateral quadrant position and not through the fissure itself in order to prevent limit creation of a keyhole deformity.

Another surgical option is chemical sphincterotomy with botulinum A toxin (Botox). Botox functions by preventing acetylcholine release, which causes relaxation of the muscle. The effects last 3–6 months. There is risk of incontinence that is time-limited by the duration of the effects of the neurotoxin. A meta-analysis of randomized controlled trials was performed comparing Botox to LIS and showed higher recurrence rates with Botox and higher rate of minor anal incontinence for LIS [7]. Persistent fissures may be amenable to botulinum injections, contralateral internal sphincterotomy or anal advancement flap into the anal canal. Additional information on anal fissure is covered in Chap. 14.

Anorectal Abscess

Patients with acute abscesses will often present with the most discomfort that is described as worsening pressure and pain. The pain is often worse before and during a bowel movement with some improvement afterwards. It can also be worse with direct pressure, as some patients will be seen laying or sitting while shifting away from the affected side. Patients may describe associated fevers, chills or even difficulty urinating [8–11].

The exam may reveal erythema and induration of the perianal skin, with an area of fluctu-

ance or even a point of active drainage (Fig. 18.4). In these cases, an incision and drainage is indicated after injection of local anesthesia. This can be done with a catheter or an ellipse of skin should be removed to ensure adequate drainage while delaying skin closure (see Chap. 10). Packing is usually unnecessary unless there are concerns for hemostasis—any packing should be removed in a timely fashion (24 h or less). Blind and aggressive spreading within the abscess cavity in hopes to "break up loculations" has the risk of pudendal nerve injury and should not be performed.

When there is no identifiable abscess on external exam, the digital rectal exam can reveal an area of induration or fluctuance within the anal canal or rectum. It is common to have a deep postanal abscess with no external evidence. If the internal exam is also unrevealing but the suspicion remains high and a parasacrococcygeal approach does not drain a collection, an imaging study such as a pelvic CT can help localize the abscess. Larger abscesses, such as horseshoe abscesses, are best treated in the operating room. Overall, the location of the abscess will determine the approach of drainage.

Recurrent abscesses after incision and drainage usually indicate formation of a fistula-in-ano. Persistent drainage through the fistula may cause pruritus, which may in turn lead to chronic perianal pain.

Other Causes of Acute Pelvic Pain

While the previously mentioned etiologies are the most commonly encountered, the differential for anorectal and pelvic pain is broad and includes anything from idiopathic pruritus to inflammatory disease to neoplasm.

Pruritus Ani

Although it is a particularly sensitive organ, the perianal skin can have poor discrimination of sensation. Patients with pruritus ani will complain of burning or itching pain. They may endorse a feeling of being unable to get clean enough, prompting them to use medicated wipes or scrubbing of the area. Unfortunately, scratching the area starts a cycle of itching and augments irritation.

The exam will reveal skin changes such as excoriation, cracked skin, discoloration (white or red) thickened folds and moisture (Fig. 18.5). A digital rectal exam may reveal poor tone, which would allow for seepage. One should distinguish the presenting symptoms from cancers, anal fissures or infectious ulcers.

Treatment is nonsurgical and targets altering bowel habits. Patients should discontinue use of topical salves, soaps, hemorrhoid creams, and medicated wipes, as well as avoid excessive scrubbing and scratching. We recommend gentle

Fig. 18.4 Perianal abscess. With permission from [57] © 2014 Springer

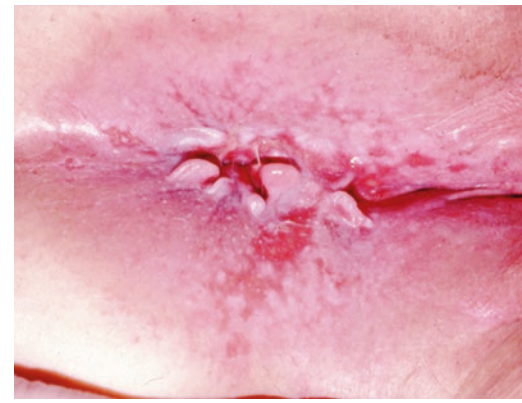

Fig. 18.5 Pruritus ani. With permission from [57] © 2014 Springer

blotting of the area after bowel movements with a cotton ball. Leaving a fluffed cotton ball at the anus helps absorb moisture and protect the surrounding skin. The use of desiccants like cornstarch can help in a similar way but can often act as abrasive if too much is used.

The quality of stools also needs to be addressed by normalizing or "drying" the stools with increasing dry soluble fiber intake and decreasing excess water intake to limit irritation. Fiber wafers are particularly useful in this situation.

Finally, for patients who have persistent symptoms despite adhering to these recommendations, consider a biopsy of the perianal skin to rule out other conditions such as lichen sclerosis et atrophicus, Bowen's disease, Paget's disease, and other conditions that may benefit from dermatologic referral or appropriate treatment based on biopsy results (see Chap. 13).

Hidradenitis Suppuritiva

Hidradenitis is a cutaneous condition believed to originate from a disorder of sebaceous apocrine gland metabolism and can often be misdiagnosed as perirectal abscesses. These patients will often have similar lesions in the axillae and groin. The exam may reveal multiple superficial abscesses in the perineal, axillary and inguinal regions. Careful examination will show sparing of the perianal verge skin from the disease process, as this area is devoid of hair and skin accessory glands.

Treatment depends on the extent of disease but always starts with antibiotics. Extensive lesions that persist despite medical treatment may require excision. As with typical perirectal abscesses, incision and drainage is indicated and extensive disease may require operative drainage (see Chap. 16).

Infectious

Infectious causes should always be considered in the differential of a patient presenting with anal or rectal pain in the setting of fever, perirectal drainage, or history of sexually transmitted disease (STD). One should inquire about HIV status, as proctitis from herpes simplex in an HIV positive patient can be particularly severe (see Chap. 27).

Gonorrhea

Many patients with gonorrhea will be asymptomatic. Those who have symptoms often endorse tenesmus and even severe anal pain. If the suspicion is high for gonorrhea, one should avoid the use of lubricants due to their antibacterial properties, which may provide a false negative culture. Swabs are placed on a Thayer-Martin agar, although newer tests include nucleic acid amplification tests (NAATs), which have higher sensitivity with similar specificity of cultures [12]. Treatment includes oral cephalosporins such as ceftriaxone 250 mg IV in a single dose. Concomitant treatment for chlamydia and evaluation for other STDs should be initiated.

Chlamydia

Chlamydia is the most prevalent infection among sexually active patients between ages 14 and 24. Although the infection starts in the anal canal, proctocolitis can develop with ulcer formation in the rectum. There are multiple variations of the species *C. trachomatis*. Serovars D through K are responsible for urethritis, pelvic inflammatory disease and proctitis. It is often asymptomatic but can present similarly to gonorrhea with proctitis and discharge. Anoscopy may show friable mucosa and discharge.

Gottesman et al point out that if a gram stain shows polymorphonuclear leukocytes in the absence of gonococci, the presumptive diagnosis is chlamydia [13]. Swabbing for NAAT testing is the test of choice. Treatment includes either azithromycin 1g orally once or doxycycline 100 mg orally twice daily for 7 days. These patients should also abstain from sex for 1 week after starting treatment to decreases transmission rates [14].

Lymphogranuloma venereum (LGV) is another subset caused by *C. trachomatis,* with the most common serovars being L1, L2 or L3. LGV presents with unilateral tender lymphadenopathy

in the inguinal and femoral region. Anoscopy may reveal ulcers and bloody mucous discharge from the rectum with fever and pain. The disease can lead to the formation of fistulae and strictures, if not treated. The diagnosis is made by clinical suspicion and confirmed with NAAT. Treatment involves doxycycline 100 mg orally BID for 21 days, or alternatively erythromycin base 500 mg orally 4 times daily.

Herpes Simplex/Zoster

The two types of herpes simplex virus are HSV 1 and HSV 2. Once infected, patients have a lifelong viral infection. HSV 2 is most commonly associated with genital herpes although HSV 1 can also present similarly. Patients have significant pain with associated perianal ulcers.The first outbreak may be with fever, lymphadenopathy on exam, and last longer than subsequent recurrences. The exam may reveal vesicles that rupture and form shallow ulcers that heal in about 3 weeks. The disease can extend into the anal canal and rectal mucosa, confirmed by visualizing ulcers or friable mucosa on anoscopy or proctoscopy. The pain can be quite severe, even causing difficulty with voiding.

Diagnostic tests include a viral culture and serologic testing. Because the virus persists in the ganglia of sensory nerves, there is no cure and recurrence is common. In some cases, associated radiculopathy in the lumbosacral distribution can cause bladder and sexual dysfunction, along with pain along the buttocks and thighs. For this reason, herpes zoster can be included in the differential for chronic pelvic pain as well. Treatment is acyclovir 400 mg orally 3 times daily for 10 days and should be supplemented with pain medication and warm soaks.

Syphilis (Treponema Pallidum)

Known as the "Great Impersonator", perianal syphilis can be misdiagnosed as anal fissures. The severe pain generally resolves on its own over 3–6 weeks, and is similar to that of a typical fissure [15]. Lesions are atypical in their location and may also be multiple. Early lesions are infectious but it may take a week for symptoms to present. Inguinal lymphadenopathy can often be appreciated.

Diagnosis is made by screening for rapid plasma reagin (RPR) followed by a treponemal test to confirm the diagnosis. Treatment is a single muscular injection of benzathine penicillin G, which generally cures the patient with early diagnosis.Doxycycline 100 mg orally twice daily for 2 weeks is acceptable for those with a penicillin allergy.

Chancroid (Haemophilus Ducreyi)

Overall, chancroid is less and less common in the US. The lesions start as a tender erythematous papule that becomes a pustule and then a painful genital or perianal ulcer. These may also have associated abscesses.

The diagnosis is confirmed with a gram stain and culture on chocolate agar. The gram stain may show gram-negative rods in small groups. This should be followed with a special culture media. The suspicion should be high if there are one or more painful genital ulcers with lymphadenopathy, with syphilis and HSV having been ruled out. The treatment is azithromycin 1 g orally once, ciprofloxacin 100 mg orally twice daily for 3 days or erythromycin base 500 mg orally twice daily for 3 days.

Granuloma Inguinale (Calymmatobacterium Granulomatis)

The exam for granuloma inguinale will reveal extensive ulcers of the genitalia and anus with granulation-like tissue and rolled edges. The lesions bleed easily and may have associated lymphadenopathy. Diagnosis requires visualizing the dark-staining Donovan bodies on biopsy. Treatment is doxycycline 100 mg orally twice daily for 3 weeks or until lesions are healed.

Overall, the sexually transmitted anorectal infections described can present with severe proctitis or ulcerations, as shown in Table 18.2. Once identified, antibiotics are the first line of treatment. Along with treating the acute infection, counsel patients on HIV as well as on the prevention of further infection and spread of disease.

Table 18.2 Sexually transmitted diseases associated with anorectal pain

Disease	Organism	Histology	Symptoms/signs	Treatment
Gonorrhea	Neisseria gonorrhea	Gram negative diplococcus in pairs/clusters on Thayer-Martin agar/NAATs	Pruritus, thick bloody mucopurulent discharge from anal crypts	Ceftriaxone 250 mg IM ×1
Chlamydia	Chlamydia trachomatis Serovars D-K	NAAT	Tenesmus, mild proctitis	Azithromycin 1 g PO ×1 or doxycycline 100 mg PO BID ×7 days
Lymphogranuloma venereum	Chlamydia trachomatis Serovars L1, L2, L3	NAAT	Small shallow ulcers with rapid healing, bloody mucoid discharge	Doxycycline 100 mg PO BID ×21 days
Herpes simplex	HSV 2, HSV 1 (less common)	–	Vesicles which form shallow ulcers, coalesce into groups with erythematous base	Acyclovir 400 mg PO TID ×10 days (less frequent dosing for recurrence)
Syphilis	Treponema pallidum	–	Chancre, clean based eccentric ulcer, rolled edges	Benzathine penicillin G 2.4 million units IM
Chancroid	Haemophilus ducreyi	Gram negative rods in small groups on chocolate agar	Indurated, tender papule, gray/yellow exudates at base	Azithromycin 1 g PO ×1, ceftriaxone 250 mg IM ×1, ciprofloxacin 100 mg PO BID ×3 days
Granuloma inguinale	Klebsiella (calymmatobacterium) granulomatis	Intracellular bacterium, dark staining Donovan bodies	Extensive and progressive ulcers with rolled edges, granulation-like tissue, bleed easily	Doxycycline 100 mg PO BID ×3 weeks or until all lesions healed

Perianal Crohn's Disease

Patients with Crohn's disease may present with pain from fistulae with associated proctitis. There may also be pain from sphincter spasm. Because the discomfort can limit an office exam, evaluation is typically best under anesthesia in order to identify and treat fistulae and undrained abscesses (Fig. 18.6). One should avoid fistulotomy in most cases and place draining setons instead. Fissures should be approached medically as with non-Crohn's patients, but sphincterotomy should be avoided due to poor healing (see Chap. 29).

Fig. 18.6 Perianal Crohn's disease. With permission from [57] © 2014 Springer

Proctitis/Pouchitis

Inflammation of the rectum typically presents as pain in the setting of an increased number of bowel movements per day, as well as associated mucoid

and bloody discharge. A thorough history is key as proctitis can arise from a history of radiation treatment, diversion, infections, chemical irritation and inflammatory bowel disease.

Pouchitis presents similarly and occurs in up to 50% of patients who undergo a restorative ileal pouch-anal anastomosis for conditions like ulcerative colitis. This condition develops due to bacterial overgrowth imbalance in the pouch. Similarly, there can be "cuffitis", or inflammation of any remnant anorectal mucosa [16].

The diagnosis is confirmed with proctoscopy, evidenced by inflammation of the rectal mucosa. Treatment depends on the cause of the inflammation. Proctitis treatment may involve the use of antibiotics as well as anti-inflammatory agents. Pouchitis treatment also involves antibiotics but recalcitrant cases may require steroids or even immunosuppressants. Extreme cases may require diversion or excision of the pouch.

Radiation

Radiation to the perineal and pelvic area can cause anodermal thinning, sphincter injury, stenosis and proctitis. These patients endorse a history of anal, distal rectal, vulvar or prostatic cancers. For this reason, the evaluation should involve a search for recurrent malignancy.

Symptomatic treatment includes the use of sucralfate suppositories, which requires a compounding pharmacy in most cases. If bleeding persists, a dilute 10% formalin solution can be instilled into the rectum [17]. More recalcitrant bleeding can be addressed with argon beam coagulation to achieve hemostasis by destroying damaged tissue.

Anal Stricture

Anal strictures can be iatrogenic or related to radiation or a history of malignancy. The patient will describe pain during defecation and a change in stool caliber. Difficulty introducing the finger or anoscope due to the stenosis confirms the diagnosis and a tearing of the anoderm can sometimes be appreciated.

Workup should include the ruling out of any associated or recurrent malignancy. Benign strictures can be addressed with dilation or anoplasty (see Chap. 14).

Anal/Rectal Cancer

The fear of malignancy is one of the major reasons patients seek evaluation for pelvic pain. Fortunately, most causes of pain are benign. Nevertheless, malignancy should always be considered in the differential, as anal and rectal cancers can present with pain. These patients may endorse unexpected weight loss or a family history of malignancy [18–20]. Low rectal cancers can present with a change in bowel habits, whether as thinner caliber stools or watery stools, as well as bleeding (see Chaps. 21, 22, and 23).

The abdominal exam may show distention in the case of near-obstructing tumors. There may be lymphadenopathy in low rectal or anal canal and margin squamous cell cancers. The digital rectal exam and rigid proctoscopy are essential in identifying the location of these tumors in reference to the anal verge, anorectal ring, prostate and vagina.

Anal cancers can present like fissures, with pain during and after bowel movements and spotting of blood. Despite appearing like typical fissures, anal cancer lesions will not heal despite appropriate medical treatment. A biopsy should be performed to confirm suspicions.

Conditions like Paget's (intraepithelial adenocarcinoma) or Bowen's (intraepithelial squamous cell carcinoma) disease will typically present with more chronic itching and irritation than pain. Like anal cancer, the pain can be associated with ulceration and sphincter spasm. Once confirmed histologically, treatment of these conditions is wide local excision.

Rectal Prolapse

The degree of pain with rectal prolapse often relates to the degree of prolapse. The pain is usually notable during and after defecation with associated prolapsing of tissue (Fig. 18.7).

Fig. 18.7 Rectal prolapse. With permission from [57] © 2014 Springer

Incarceration of a prolapsed rectum can present with acute pain.

The exam is confirmatory but may require a "toilet test" in order to demonstrate the prolapse, whether by having the patient valsalva on the toilet or by identifying the pathology on defecography. Defecography may also identify other prolapsing organs in the form of cystocele, uterine prolapse, and enterocele.

Chronic non-ischemic prolapse can often be reduced and definite surgical treatment can be planned electively. Acute incarceration may require more urgent surgical intervention and a resection proctosigmoidectomy should be performed in these cases. Additional information on rectal prolapse is presented in Chap. 8.

Retrorectal Tumors

In addition to being a rare diagnosis in general, retrorectal tumors will rarely present solely as chronic pelvic pain. Patients may have changes in stool caliber or constipation, as these extrarectal tumors cause external compression of the bowel lumen. The rectal exam may reveal a palpable mass posteriorly or laterally. Workup includes imaging with CT or MRI to characterize these lesions as cystic or solid and to identify their location and association with other pelvic structures (Fig. 18.8). A biopsy is not routinely required and essentially all lesions should be resected due to malignant potential (see Chap. 26).

Fig. 18.8 Retrorectal tumor. With permission from [57] © 2014 Springer

Prostatitis

Anterior rectal pain may originate from the prostate. Patients describe the pain as a dull ache. The causes of prostatitis can range from infectious to neurologic and these patients may endorse urinary or sexual dysfunction. The exam is often confirmatory with point tenderness upon palpation of the prostate with a lack of findings in the remainder of the exam [21]. A urological referral is indicated. Bacterial causes, including those of STDs, will require antibiotic treatment.

Gynecological Causes

Gynecological causes of pelvic pain vary widely but rarely present as anal pain specifically. Instead, patients describe a deep lower abdominal or pelvic pain with symptoms akin to constipation or outlet dysfunction. Causes can include rectocele, endometriosis, ectopic pregnancies or adnexal abnormalities and diseases. These should always be considered in the differential of pelvic pain in the female patient and may prompt a gynecological referral, given the broad range of possibilities beyond the scope of this chapter.

Neurogenic Pain

Disorders affecting the distal lumbar and sacral sensory nerves can cause pelvic pain. The etiologies of neurogenic pain are beyond the scope of this chapter but can include spinal anatomical abnormalities like ruptured discs, malignancies or bony abnormalities. The perianal and rectal exam may reveal weak sphincter tone and the neurological exam should also assess for lower extremity weakness and sensory changes. Workup involves imaging such as MRI and appropriate referral as indicated.

Chronic Pelvic Pain

Frequently recurring pain of the anal canal, rectum and pelvis is a complex issue that can stem from a variety of causes. This already sensitive issue is made more challenging as patients may see multiple specialists before the cause of their pain is correctly identified. Despite the considerable effect this pain may have on quality of life, only about 1/3 of patients will consult a physician. In addition, there remains a dearth of reliable research regarding these conditions overall [22]. Although 15% of women in the United States report chronic pelvic pain, the condition also affects men [23].

Similar to identifying the cause of acute pelvic pain, the history and physical exam are crucial to identifying the diagnosis. Chronic pain is distinguished from acute pain by the duration of symptoms, with patients experiencing pain for more than 6 months [24]. The pain can be constant or intermittent with variability of improvement with warm baths or bowel movements. The quality can be described in a number of ways, from sharp to dull and from burning to aching and cramping. A key aspect to elicit from a history of chronic pain is the tempo and variation during the course of a day.

Patients usually do not complain of fever, chills or bleeding. Other associated symptoms such as dyspareunia, vaginal bleeding, or dysuria should prompt appropriate subspecialty referrals. If there is a history of constipation, it may be specifically defined as difficulty with stool evacuation. Patients may also have a history of irritable bowel syndrome and anxiety or depression. Inquire about gynecological and obstetric history, as well as any history of psychosocial trauma or abuse. Obtain records of previous anorectal, urological and gynecological operations as well as reports from colonoscopies and pathology.

Key aspects of the physical exam for chronic pelvic pain should focus on the perineal exam and anorectal exam as described previously. A vaginal exam may reveal other pathologies such as discharge or cervical motion tenderness, which may suggest STDs. The pelvic floor musculature is another source of chronic pelvic pain that can be assessed with the digital rectal exam. Have the patient squeeze and valsalva. One will feel the finger pull up during a normal squeeze as the pelvic floor contracts and the descent of the perineum and the degree of prolapse can be appreciated on Valsalva maneuver. Evaluate spasm of the levator muscles or attempt to reproduce the pain described by patients by palpating these muscles. Finally, palpation of the coccyx may reproduce pain in coccygodynia.

Despite a comprehensive workup, one study found that an organic cause was found in only 15% of patients with chronic pelvic pain. These patients are placed under the subset of "functional" chronic anorectal and pelvic pain [25]. The goal of this section is to focus on the most common syndromes associated with chronic pelvic pain, including levator syndrome, proctalgia fugax, coccygodynia and pudendal neuropathy (Table 18.3). There is also a brief mention of urogynecological causes of chronic pain that one should be familiar with the workup.

Urogynecological Causes

The most common cause of pelvic pain in women is endometriosis. The degree and quality of symptoms depends on the location and depth of involvement of the extrauterine tissue deposits. When the gastrointestinal tract is

Table 18.3 Pelvic pain syndromes

	Pain duration	Exam
Proctalgia fugax	<20 min, usually seconds, can awaken from sleep	Episodic
Levator ani syndrome	>20 min	Tenderness with palpation of the puborectalis
Coccygodynia	Variable	Tenderness with palpation of coccyx
Pudendal neuralgia	Variable	Distribution along the pudendal nerve

involved, patients may describe cramping and changes in bowel habits, oftentimes coinciding with the menstrual cycle. While some patients are diagnosed by symptoms alone, many require a diagnostic laparoscopy for identification of the characteristic lesions. Treatment options range from hormone therapy to surgical excision of implants with concomitant hysterectomy and salpingo-oophorectomy in refractory cases. Other gynecological causes of pelvic pain include pelvic congestion, vulvodynia and vaginitis.

Urological causes of chronic pain include cystitis and urethral syndrome. These patients endorse may complain of dysuria along with changes in urgency and frequency. The workup should include urinalysis and may require cystoscopy. Both gynecological and urological causes go beyond the scope of this chapter and should prompt a referral to the appropriate specialist.

Pelvic Floor Pain Syndrome

Improper function of the pelvic floor musculature can cause significant chronic pelvic pain. The pathophysiology of these conditions is unclear but is likely related to spasm or tension of the striated muscles of the pelvic floor. Precipitating factors can vary from anxiety to childbirth to pelvic, anorectal and spinal operations [26, 27].

Levator Ani Syndrome

The levator ani syndrome is also referred to as puborectalis syndrome, chronic idiopathic perineal pain, pyriformis syndrome and chronic proctalgia. Patients describe a dull pressure or ache that distinctly worsens when sitting or lying down and improves with standing. This pain usually begins in the morning and worsens through the day, but rarely occurs at night. The pain can be associated with the rectum and extend to the sacrum, coccyx or even gluteal region and thighs. Although difficult to localize in some cases, some patients may complain of more pain on the left side of the rectum where the levator ani muscles insert into the pubic ramus [26, 28]. A bowel movement may provide relief, which distinguishes this diagnosis from other causes of pelvic pain.

There may also be overlap with other levator functional pathology. Specifically, some patients may also have obstructed defecation constipation due to non-relaxation of the puborectalis portion of the levator ani muscles. This may need cine video defecography to confirm the diagnosis. Treatment of the one set of symptoms may relieve the other set.

Patients may also have a history of anxiety or depression, recent stress or trauma or recent prolonged sitting. Although diagnosed more frequently in women and in those between 30 and 60 years of age, the actual prevalence is unclear [29].

In order to provide consistency in the diagnosis of this syndrome, the Rome III criteria were developed for levator ani syndrome and include the following: (1) chronic or recurrent episodes of rectal-area pain or aching, (2) lasting 20 min or longer, (3) occurring for at least 12 weeks in the past 12 months, and (4) in the absence of other causes [28]. This longer duration of pain distinguishes levator syndrome from proctalgia fugax.

During the exam, applying posterior traction to the levator muscle near its coccygeal attachment can often reproduce the pain. The experienced examiner may appreciate spasms of this muscle. Digital massage can improve symptoms in many

cases [30]. Even if these exam findings are not identified, the diagnosis of levator syndrome is still possible. Nevertheless, one should exclude other causes with additional tests such as colonoscopy, GI contrast studies, CT scan and in some cases diagnostic laparoscopy. The use of anorectal manometry has been reported but results are inconsistent [25].

Management is multifactorial and involves patient reassurance, pharmacological therapy and physical therapy. Reassurance is key as anxiety may augment symptoms for these patients. Reported use of anxiolytics in both oral and suppository form have been reported but side effects should be considered. Warm baths are thought to alleviate symptoms by relaxing the muscles and have no harmful side effects, although a review of the literature revealed a lack of scientific data to support its use [31].

Digital massage of the puborectalis sling on the affected side has been described. This is limited by the patient's discomfort. This is rarely the only therapy, as massage is often done in conjunction with sitz baths or a short course of oral benzodiazepines. Long-term effects are not clear [27, 28, 30].

Physical therapy of the pelvic floor is helpful in the treatment of these patients. This requires a dedicated physical therapist that is sensitive to the patient's needs and, often, initial resistance to this mode of therapy. Various techniques utilized by pelvic floor physical therapists include biofeedback, electrogalvanic stimulation and internal massage. The research behind these techniques show varied success and is based mainly on small studies with a wide range of follow-up.

Biofeedback was first described in 1991 and studies have been small with varied success rates [32–34]. By retraining the coordination and relaxation of the levator muscles, patients may be able to break the cycle of spasms. Case reports vary in success and none are controlled studies. [35].

Electrogalvanic stimulation (EGS), a technique first described in 1982, involves stimulation of the pelvic floor muscles with a transrectal probe [27, 36, 37]. The stimulation is administered for 20–30 min per session for 3 sessions a week, with the goal of fatiguing the muscles. Sohn et al. describe starting at a pulse frequency of 80 cycles per second with a gradual increase in voltage to the point of discomfort [38]. While there are reports of up to 70% of patients finding relief, the long-term response is less sustainable.

Botox injection has also provided a varied degree of relief in case reports [39]. Unfortunately, the literature also reports a variety of dosages and techniques for administering Botox [40, 41]. We percutaneously inject 100 units of Botox mixed with injectable saline at insertion points of the levator muscle and into the muscle belly.

Sacral nerve stimulation (SNS) has been reported in the treatment of functional anal pain but results are varied. Falletto et al. showed improvements in pain scores at a mean follow up of 15 months and recommended SNS as an option for pain refractory to biofeedback or medications [42]. However in one small study, Dudding et al. showed that SNS was not an effective treatment modality with a 5-year follow-up [43].

Other reported therapies include use of acupuncture, injection of local anesthetics or steroids into the arcus tendon of the levator muscle, or even surgical division of the puborectalis muscle. The later option resulted in a high incidence of incontinence to both stool and gas in case studies, making this therapy undesirable [44].

For all therapies described above, the literature remains highly variable in terms of the inclusion criteria, follow-up intervals, and sample size. Even among randomized studies, there remains variability regarding the number of treatments of one kind and consideration of the effect of previously attempted therapies. Finally, along with pelvic floor musculature dysfunction, there remains the variable of brain processing of pain that may be altered in these patients. For these reasons, patients with levator syndrome may have different therapies to choose from, but outcomes cannot be defined at this time.

Proctalgia Fugax

Patients with proctalgia fugax present with a distinct description of severe sharp pain that lasts for a few seconds to minutes, then resolves completely. The average duration is around 5 min and

patients are asymptomatic between episodes [45]. This severe pain can awaken them from sleep and is localized to the anus or rectum. Because symptoms are fleeting and generally infrequent, proctalgia fugax is difficult to evaluate. While there have been suggestions that stress, defecation, long periods of sitting or menstruation may trigger pain, there may be no obvious trigger identified [46].

The estimated prevalence is up to 18% of the general population but with less than a quarter of those patients reporting symptoms to a physician. The age range is wide but typically affects those around 50 years of age with higher prevalence in women [46, 47]. Spasm of the pelvic floor muscles as well as neuropathy has been implicated as causes. Anal manometry revealed increased resting anal pressures but no differences in squeeze pressure or sphincter relaxation of sphincter complex thickness in those with proctalgia fugax [47].

Although the history of this pain is distinct from other chronic pelvic pain syndromes with short episodes of severe pain, proctalgia fugax is a diagnosis of exclusion. The physical exam and workup can be extensive before this diagnosis is identified.

There are limited data regarding management, but include patient reassurance and treatment of muscle spasm. There are reports of use of topical antispasmodics or muscle relaxants (e.g. Beladona and opium suppositories) [48]. Biofeedback is a noninvasive method used to treat other causes of pain and may be helpful [45]. The use of Botox is more invasive and has been reported with injection of 50 units of the toxin into the anal sphincter with pain relief and no incontinence at 2 months [49]. Other reported modalities of treatment in small studies include the use of inhaled albuterol, intravenous Lidocaine and internal anal sphincterotomy.

Coccygodynia

Coccygodynia is described as pain localized at or around the coccyx. The pain is triggered with prolong sitting and patients often describe repetitive trauma or childbirth as the inciting event [50]. This diagnosis can be confirmed on exam by reproducing pain with palpation of the coccyx on digital exam and external palpation.Plain films of the pelvis are obtained to rule out fractures and more than 50% of patients show features of coccyx instability on imaging [51]. Relief of pain with local anesthetic injection into the coccyx also confirms the diagnosis. The cause of pain may also be related to associated levator spasm with traction on the coccyx.

Treatment begins by avoiding aggravating factors, with Sitz baths and sitting on a cushion in order to reduce pressure [52]. Other reported therapies include levator massage, physical therapy and injection with steroids into the joints and tissues around the coccyx. Coccygectomy is less commonly utilized as a first step in treatment but has been reported for refractory cases [53]. Outcomes have not been evaluated in controlled trials but one study showed that wound infection is the most common complication and outcomes are likely related to surgeon expertise [54].

Pudendal Neuralgia

The pudendal nerve, which runs through Alcock's canal, includes a mix of sensory and motor nerves originating from S2 to S4. Compression of the pudendal nerve at the ischium and obturator internus muscle causes burning, pinching or a twisting sensation that may be located in the perineum, vulvar or anorectal regions. The second common site of compression is at the ischial spine in the gluteal region. Sitting will often aggravate symptoms whereas standing will relieve them. The pain may be unilateral or bilateral. While entrapment is the most common etiology, other reported causes include herpetic and post radiation neuropathy [55].

As with the previous conditions, other causes of anal pain should be excluded. Confirmatory tests can include pudendal nerve terminal motor latency measurements, which will be prolonged. A CT scan may identify the point of nerve compression and a diagnostic CT-guided nerve block

by anesthesia can attempt to relieve the pain. This is usually done first with local anesthetic and if successful in relieving pain, additional injections are provided by peridural route. Long-term results are not available. A surgical decompression may also need to be considered [56].

Summary

Pelvic pain, whether acute or chronic, is one of the most common complaints bringing patients to the colorectal surgeon's office. Quality of life is significantly affected and made worse by a delay in diagnosis, prompted by patient embarrassment and referrals to several specialties. A thorough history and detailed anorectal exam can provide the diagnosis in most cases. The most common causes of acute pain are treatable and non-surgical treatments are effective in a majority of cases. Chronic causes of anorectal and pelvic pain are less well defined but are closely related to dysfunction of the pelvic floor musculature. While a multidisciplinary approach is helpful, it is important to be sensitive to the patient's embarrassment and frustration over this delicate problem. Diagnosing a patient with a pelvic pain syndrome requires a diligent and systematic approach, as well as a careful ear for revealing components of the patient's history.

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