Anatomy and Pathology of the Ischiorectal Fossa

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After participating in this educational activity, the radiologist should be better able to identify the anatomic landmarks of the ischiorectal fossa on CT and MRI and to appreciate the systematic approach to the differential diagnosis of an ischiorectal fossa lesion.

Introduction

The ischiorectal fossa (IRF) contains only a few tissue components; therefore, most pathologic conditions that affect the IRF develop by direct extension from a bordering structure rather than intrinsic pathology within the fossa. The IRF primarily contains fat and neurovascular structures, whereas the borders contain genitourinary, enteric, and musculoskeletal structures. Furthermore, given the relative low incidence of tumors in this region, this anatomic space has received little attention in the radiologic literature.\(^1,2\) Traditionally, classification of IRF disease included infectious, inflammatory, congenital, and neoplastic conditions. A more practical approach, especially with regard to incidental findings, is to conceptualize a differential diagnosis on the basis of the origin, direction of extension, if any, into the IRF, and imaging characteristics of the abnormality. The objective of this article is to offer the radiologist a practical approach to IRF disease on the basis of identification of the specific direction of extension of the process versus identification of a lesion of the IRF proper.

Anatomy

Understanding the anatomy of the IRF, or as some authors refer to it, the ischioanal fossa, and the relationship of structures that border it are essential in formulating a differential diagnosis and providing the referring clinician with the necessary information to make an appropriate management decision. The IRF is a pyramidal space that is located within the perineum, below the pelvic diaphragm. It is composed of fat, neurovascular structures, lymphatics, and connective tissue (Figure 1).\(^1,2\) The IRF’s medial border is the levator ani and the external sphincter muscles.\(^2\) This medial border can be divided on the axial plane into the anterior urogenital space (urethra, vagina, or prostate) and posterior anorectal space. This division is based on the relationship to the transverse perineal muscle or an imaginary...
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The anterior border of the IRF is triangular, formed by the convergence of the medial urogenitalia and the lateral musculoskeletal structures; thus there is no distinct anterior border. The posterior border is at the base of this triangle and is distinct, comprising the sacrum, sacrotuberous ligament, and the lower aspect of the gluteus muscles. Communication of the right and left IRF occurs at the posterior border through the deep postanal space, which is located between the pelvic floor muscles superiorly and anococcygeal ligament inferiorly. Inferiorly, the IRF is bounded by the skin and subcutaneous fat of the perineum. Superiorly, it is bordered by the pelvic diaphragm, which contains the levator ani. Identification of extension of disease superior to the levator ani indicates extension beyond the confines of the pelvic diaphragm that often dictates an alternate surgical approach and can be an indicator of a complex clinical condition. Lastly, the IRF can be divided into the superior and inferior ischiorectal spaces by the septum of the IRF, which contains nerves and vessels that supply the anorectum and scrotal or labial branches of the internal pudendal vessels. In addition, it should be noted that some authors subdivide the IRF into the more superior IRF and a more inferior ischioanal fossa, using the anus as the boundary in the craniocaudal direction.

Imaging Techniques

The widespread use of CT and MRI has made imaging of the IRF more common. MRI is considered the imaging modality of choice for assessment of the IRF, given its superior tissue contrast and multiplanar capabilities.1 Diagnostic imaging, specifically MRI and CT, plays a critical role in identification, staging, and surgical planning of processes affecting the IRF. The coronal and axial planes are most useful in evaluating the IRF. The levator ani is best identified in the coronal plane, whereas the axial plane is more familiar to radiologists. When the two planes are used in conjunction, the IRF is identifiable and distinguishable from its boundary structures.2
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Ischiorectal Fossa Pathology

Most conditions affecting the IRF are due to secondary involvement from bordering pelvic or perirectal disease. These secondary conditions may indirectly affect the IRF because of mass effect, with a bulging appearance and preservation of the border structure, or they extend into and affect the IRF directly. The former usually is due to benign or infectious processes and the latter usually from malignancy, some infectious processes, or congenital or acquired malformations. The vast majority of lesions involving the IRF are benign and frequently cystic. Malignancies are less common, and they tend to be solid and show more aggressive imaging features. However, caution is recommended, as there is overlap of imaging characteristics of benign and malignant etiologies. Hemorrhage, frequently from trauma, surgical procedures, birth delivery, and bleeding disorders, can affect the IRF proper or secondarily affect it from extension from any of the borders (Figure 2).²

Ischiorectal Fossa Proper Lesions

Primary pathologic conditions originating in the IRF proper are rare. Lesions occurring primarily within the IRF include lipomatous tumors, aggressive angiomyxoma, vascular lesions, and neurogenic tumors. Simple lipomas in the IRF are characterized by homogeneous, circumscribed fat without enhancing solid components.² Mixed fat and nonfatty solid lesions should raise suspicion for a liposarcoma. An aggressive angiomyxoma is a rare soft tissue mass that occurs in young women, usually in the third to fifth decades of life.¹ When it occurs, it frequently originates within the IRF, commonly presents at a large size, and tends to smoothly displace rather than invade perineal structures.¹² Angiomyxoma has high signal on T2-weighted MR sequences because of its myxomatous content.¹ A “swirled” or “whorled” appearance on T2-weighted MR images and enhanced CT also has been described.³ The name “aggressive” refers to its high rate of recurrence after resection rather than its malignant potential.³

Hemangiomas are primary vascular lesions that appear bright on T2-weighted MR sequences with multiple slow flow vessels that may have phleboliths (best seen on CT), whereas hemangiopericytomas are rare vascular lesions that also may involve the IRF.² Low-flow (capillary, lymphatic, and venous) and high-flow (arteriovenous malformations and arteriovenous fistulas) vascular malformations also may present in the IRF.³ Some capillary, venous, and lymphatic malformations are associated with Klippel-Trénaunay syndrome, which also consists of soft tissue hypertrophy of the ipsilateral limb and a port-wine stain over the affected extremity.

Peripheral nerve sheath tumors are generally benign but occasionally malignant. The subtypes are neurofibroma and schwannoma. Both types are characteristically bright on T2-weighted MR images and may mimic water, but they will enhance after contrast medium administration. The “target sign” is a rim of higher signal surrounding a core of lower signal on T2-weighted MR images and is seen in some benign peripheral nerve sheath tumors. This sign cannot definitively distinguish neurofibromas from schwannomas, but the absence of this sign may suggest malignancy.⁴ Plexiform neurofibroma is a type of neurofibroma specific to neurofibromatosis type 1. This tumor is a network-like growth of tumor involving multiple fascicles of a nerve, resulting in a diffuse mass of thickened nerve fibers. In the IRF, it presents as a lobulated, T2-hyperintense mass arising from pudendal nerves in the pelvis, with characteristic

Figure 2. A 45-year-old man in the emergency department after a motorcycle accident. This coronal, reformatted, contrast enhanced CT scan demonstrates a small focus of contrast medium extravasation within the left IRF (solid white arrow), probably attributable to a venous bleed. The black arrow denotes the levator ani and the broken white arrow the septum of the IRF.

Figure 3. A 25-year-old man with neurofibromatosis type 1. This axial, T2-weighted, fat-suppressed MR image of the pelvis reveals multiple, lobulated, hyperintense masses (black arrow) with central hypointense foci or target sign (arrowhead) filling the IRF symmetrically, which is characteristic of a plexiform neurofibroma.
symmetric involvement of the IRF (Figure 3). Finally, hematogenous spread or metastasis to lymph nodes within the IRF is extremely uncommon; and lymphatic involvement, although rare, is most commonly due to anorectal or prostatic carcinoma.1,2

Primary lesions occurring within the IRF include lipomatous tumors, aggressive angiomyxoma, vascular lesions, and neurogenic tumors.

Ischiorectal Fossa Medial Lesions

The medial border of the IRF includes urogenital (anterior medial wall) and anorectal (posterior medial wall) spaces. Urogenital space lesions vary depending on the sex of the patient. The female differential diagnosis is broader and includes Gartner duct cysts, Bartholin gland cysts, Skene gland cysts, urethral diverticulum, trauma, and cervical and vaginal cancer. Although the male differential diagnosis is more limited to trauma or malignancy (prostate and penile cancer), both sexes may have urethral or bladder carcinoma. Gartner duct cysts are incomplete regression of the mesonephric (Wolffian) duct. These cysts occur within the anterolateral wall of the vagina, and when large, they can bulge into the IRF.2 These cysts are frequently associated with renal and ureteral abnormalities (e.g., congenital absence of the ipsilateral kidney and ureter, ipsilateral renal dysplasia, and crossed fused ectopia).2 Both CT and MRI show a well-circumscribed mass within the anterolateral wall of the vagina. On CT, Gartner duct cysts demonstrate water attenuation; MRI reveals marked hyperintense signal on T2-weighted sequences, unless complicated by proteinaceous or blood products. Bartholin cysts occur from ductal obstruction of Bartholin glands leading to cystic dilatation. These cysts arise from the posterolateral inferior wall of the vagina and are associated with the labia majora. Skene gland cysts usually occur at either the 3- or 9-o’clock position in the distal two thirds of the urethra and lack direct communication with the urethra lumen, whereas a urethral diverticulum is formed once the cyst ruptures into the urethra lumen, typically within its posterolateral wall.3 Cervical, vaginal, urethral, bladder, penile, and prostate cancer can present as a poorly circumscribed mass invading the IRF. Use of dynamic contrast enhanced or diffusion-weighted MRI can aid in depicting the extent of poorly circumscribed masses.

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If the lesion extends from the medial posterior wall, then anorectal space lesions are most likely. These include rectal duplication cysts, rectal cavernous hemangioma, rectal carcinoma, and inflammatory disease, including fistula in ano. Rectal duplication cysts arise from sequestration of the primitive hindgut during embryogenesis. Rectal duplication cysts are thin walled and may be multiloculated with or without communication with the associated rectal segment. Up to a third of all enteric duplication cysts may contain gastric mucosa, which can be complicated by perforation or bleeding. The presence of gastric mucosa can be identified with a technetium-99m sodium pertechnetate radionuclide scan. These duplication cysts are associated with the colon or rectum, and depending on the location and size of the lesion, can involve the IRF, typically causing mass effect. Anal duct or gland cysts are benign, mucus-secreting lesions that can have mass effect on the IRF.3 Rectal cavernous hemangiomas infrequently extend into the IRF. If they do, they exhibit a network of vascular lakes that appear as a serpiginous enhancing mass insinuating within the fatty components. A rectal cavernous hemangioma can present with pronounced rectal wall thickening and chronic rectal bleeding.1 Rectal and anal carcinoma can present as an irregular poorly circumscribed mass invading adjacent structures (Figure 4).1 High-resolution, T2-weighted MRI plays an important role in determining the extent of local disease.

Inflammatory processes involving the IRF include fistula in ano (FIA) (perianal fistula), which primarily is caused by intramuscular anal gland sepsis but can be secondary to trauma (e.g., birth delivery), inflammatory bowel disease, infection, tuberculosis, malignancy, or radiation therapy.5 FIA may present with or without associated abscess. Traditionally, preoperative planning for perianal fistulas was limited to digital rectal examination, proctosigmoidoscopy, probing with a blunt instrument, and fistulography in an attempt to delineate the course of the fistulous tract.6 However, MRI has become the imaging modality of choice to evaluate FIA.6 The St. James University Hospital classification is an MRI-based grading scale of perianal fistulas, which are divided into five grades based on the course of the fistulous tract and any associated abscesses. When evaluating FIA, the main landmarks are the anal sphincter complex, IRF, and levator ani; in addition, the radiologist should describe the fistulous tract on the basis of the anal clock.6

Figure 4. A 65-year-old woman with rectal carcinoma. This axial, contrast enhanced CT scan reveals a mass in the left IRF immediately adjacent to the medial border (arrow). Other contiguous CT images (not shown) demonstrate circumferential thickening of rectum contiguous with the left IRF mass.
This type of FIA has important surgical and clinical implications, because as a source of a pelvic infection, it can lead to sepsis.6

When evaluating a fistula in ano, the main landmarks are the anal sphincter complex, the IRF, and the levator ani.

Ischiorectal Fossa Lateral Lesions

The lateral boundary of the IRF is composed of musculoskeletal structures. Lesions involving this space include soft tissue and osseous tumors and complications of infection. Benign mesenchymal tumors and osseous tumors (e.g., osteochondroma), in addition to malignancies (chondrosarcomas, liposarcomas, osteosarcomas, Ewing sarcomas, ischiatic chordoma, and metastasis) can arise in the adjacent musculoskeletal and pelvic structures.1-3 Ischial osteomyelitis most commonly occurs secondary to direct inoculation from primary infection within the rectum, IRF, or pelvis and less commonly from hematogenous spread.

Ischiorectal Fossa Posterior Lesions

The posterior border contains the sacrum, anococcygeal ligament, and gluteus muscles, and thus it has a similar differential diagnosis as that of the lateral border.

Ischiorectal Fossa Superior Lesions

Most processes that originate superior to the pelvic diaphragm and extend into the IRF do so by descending along the posterior or posterolateral border of the IRF. Determining where the lesion is centered and whether the process is cystic versus noncystic are essential to formulating a sound differential diagnosis. When a mass is cystic, the differential diagnosis includes developmental cysts (tailgut, epidermoid, and dermoid cysts); anterior meningocele; and abscesses. Tailgut cysts are rare congenital lesions that frequently occur within the retrorectal space superior to the levator ani, but rarely they may descend along the posterolateral border into the IRF (Figure 7).1,7 They are thought to be caused by incomplete regression of the embryonic hindgut (or embryonic tailgut) and are mostly incidental lesions in middle-aged women.5 Imaging findings include a unilocular or multiloculated cystic mass adherent to the sacrum.2 On CT, a tailgut cyst appears as a circumscribed, water or soft tissue density, presacral mass without invasion of adjacent structures. On MRI, tailgut cysts typically have thin peripheral enhancement and demonstrate hypointense signal on T1-weighted and hyperintense signal on T2-weighted MR images. Epidermoid and dermoid cysts are benign epithelial cystic lesions that can be differentiated from each other on the basis of internal contents. Epidermoid cysts are usually unilocular with simple fluid content, whereas dermoid cysts may be multilocular, containing skin appendages such as hair follicles, sweat glands, and tooth buds with bright signal on T1-weighted MR images.5 Signal variability can occur in any of the developmental cysts when complicated by bleeding or infection, and solid components may arise in tailgut cysts from rare malignant transformation.5,7

It also is important to specify approximately how much of the external anal sphincter is involved, because at least 50% of the external anal sphincter must be spared by surgery to avoid stool incontinence. Grade 1 and 2 FIA involve only the internal anal sphincter and the intersphincteric space (grade 2 has an associated abscess), which is a potential space that is not usually seen unless involved by a pathologic process. Although grade 1 and 2 FIA do not extend into the IRF proper, a large associated abscess could result in mass effect protruding into the fossa.5 On the other hand, grade 3 and 4 FIA are both transspincteric (grade 4 has associated abscess) and extend through both the internal and external sphincters and directly into the IRF (Figures 5 and 6).6 Grade 5 FIA traverse the levator ani (levator plate) into the pelvis, with or without presence of secondary tracts or abscesses.

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rounded, and it is seen in approximately 50% of cases.\textsuperscript{5} Masses related to infection or inflammatory conditions have been discussed in the medial classification, but they may originate above the levator ani and present as an indistinct inflammatory process and/or a focal abscess.

**On MRI, tailgut cysts have thin peripheral enhancement and demonstrate hypointense signal on T1-weighted and hyperintense signal on T2-weighted images.**

Noncystic masses include perineal hernia, chordoma, teratoma, and masses related to infection or inflammatory conditions.\textsuperscript{1} A working knowledge of the patient’s pertinent clinical history, such as known inflammatory bowel disease, prior trauma, or instrumentation can help differentiate the origin of noncystic masses. A perineal (levator ani) hernia is one of the least common types of body wall hernias and is associated with increased intra-abdominal pressure (obesity, pregnancy, and chronic ascites); chronic infection; prior instrumentation; and, rarely, a congenital defect (Figure 8).\textsuperscript{1} Anatomically, perineal hernias can be classified as anterior or posterior depending on how they descend into the IRF on the axial plane in relation to the transverse perineal muscle.\textsuperscript{8} Chordoma is thought to represent a remnant of the primitive notochord that manifests radiographically as an expansile soft tissue mass originating within the sacrum. On CT, a chordoma appears as a heterogeneous, expansile, lytic lesion that may have intratumoral calcification.\textsuperscript{3} On MRI, the mass demonstrates predominately low signal on T1-weighted images, with intermixed foci of brighter signal because of hemorrhage or protein; and is hyperintense on T2-weighted sequences, with possibly some low intermixed signal because of hemosiderin.\textsuperscript{3} Various enhancement patterns have been

Anterior meningocele is a rare congenital defect associated with herniation of the dural sac through a defect in the anterior sacrum. An anterior meningocele can be sporadic or familial as part of the Currarino syndrome (sacral abnormality; presacral mass, including meningoceles or teratomas; and anorectal malformation or stenosis). Approximately 50% of patients have associated malformations that include spina bifida, spinal dysraphism, uterine abnormalities, and imperforate anus. The “scimitar” sign is considered a pathognomonic radiographic sign of an anterior meningocele when the sacrum is concave and

**Figure 7.** A 65-year-old woman with incidentally found tailgut cyst (arrows). Sagittal (A) and axial (B) T2-weighted MR images demonstrate a very bright signal, multiseptated mass that emanates from the pelvis through the superior border of the IRF and descends into the left posterior IRF (arrow in B). Differential diagnoses for a multiseptated, cystic mass centered at the presacral region with extension into IRF includes epidermal inclusion cyst, rectal duplication cyst, and anterior meningocele.

**Figure 8.** An 80-year-old woman with incidentally found posterior right perineal hernia. This axial, enhanced CT scan demonstrates a segment of small bowel herniating into the right IRF (arrow).
Sacral teratomas or sacrococcygeal teratomas contain elements of all three germ cell layers and are classified as mature, immature, or malignant depending on the degree of differentiation at pathologic analysis (Figure 9). Teratomas are typically well-defined, heterogeneous lesions that have both cystic and solid elements, with 50% containing fat or calcification. Identification of a teratoma in a neonate portends a better prognosis than in an adult. In both cases, complete surgical excision is required, as these lesions carry a significant malignant potential.

**Lesions Inferior to the Ischiorectal Fossa**

Structures inferior to the IRF include skin and subcutaneous fat. Processes that affect these structures include sequelae from surgery, infection, and neoplasm. Procedures can lead to migration of ectopic tissue that may result in epidermal inclusion cysts or perineal endometriomas. Infectious and inflammatory conditions include cellulitis, abscesses, fistulas, and Fournier gangrene (typically occurs in diabetic male patients with subcutaneous gas formation) (Figure 10). A variety of neoplasms originating from external genitalia (e.g., vulvar and penile), skin structures (usually squamous cell carcinoma), or sarcomas may arise from the inferior space.

**Conclusion**

Multiple pathologic processes involve the IRF, with the vast majority occurring secondary to extension of disease from bordering structures. Diagnosis and staging of these conditions is determined by whether the lesion originates from the IRF proper or is secondarily involved because of contiguous extension from a border structure. CT and MRI involve a crucial role in the evaluation, with MRI predominant because of its superior tissue contrast and multiplanar capabilities. Characterization of cystic versus solid lesions and extent of the lesion vis-à-vis the variety of structures in the IRF area are vital in providing the surgeon with the necessary information for the best surgical approach or management.

**References**

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1. All of the following are radiographic features of chordomas of the sacrum, except
   A. scimitar sign
   B. expansile soft tissue mass originating in the sacrum
   C. intratumoral calcifications
   D. lytic lesion
   E. heterogeneous lesion

2. A fistula in ano traverses both the internal and external anal sphincters with an associated abscess in the ischiorectal fossa. The grade of the fistula is
   A. 1
   B. 2
   C. 3
   D. 4
   E. 5

3. Which one of the following ischiorectal fossa proper masses presents with a core of low signal surrounded by a rim of high signal on T2-weighted MR images?
   A. Hemangioma
   B. Aggressive angiomyxoma
   C. Lipoma
   D. Benign schwannoma
   E. Liposarcoma

4. Figure 11 is a coronal, T2-weighted MR image of a 25-year-old, nondiabetic woman with Crohn disease who presented with anal pain. The most likely diagnosis is
   A. lipoma
   B. teratoma
   C. abscess
   D. vulvar carcinoma
   E. Fournier gangrene

5. Which one of the following imaging findings can aid the surgeon in counseling a patient regarding possible stool incontinence after surgery for a fistula in ano?
   A. Extension into the perineal skin
   B. Involvement of the ipsilateral ischiorectal fossa septum
   C. Presence of an associated ischiorectal fossa abscess
   D. Percentage involvement of the circumference of the external anal sphincter
   E. Ipsilateral involvement of the sacral plexus

6. The scimitar sign of the sacrum is a pathognomonic radiographic sign of
   A. sacral teratoma
   B. anterior meningocele
   C. tailgut cyst
   D. epidermoid cyst
   E. chordoma

7. A urethral diverticulum forms after rupture of which one of the following female cysts?
   A. Gartner duct cyst
   B. Skene gland cyst
   C. Bartholin gland cyst
   D. None of the above

8. Which one of the following lesions involving the ischiorectal fossa can be identified by a technetium-99m sodium pertechnetate scan?
   A. Plexiform neurofibroma
   B. Rectal cavernous hemangioma
   C. Tailgut cyst
   D. Rectal duplication cyst
   E. Teratoma

9. Which one of the following statements regarding an aggressive angiomyxoma in the ischiorectal fossa is true?
   A. It occurs equally in men and women.
   B. It often invades adjacent organs and structures.
   C. It is low signal on T2-weighted MR images.
   D. It has a target sign on T2-weighted MR images.
   E. It has a high rate of recurrence after resection.

10. Which one of the following is the imaging examination of choice for assessment of the ischiorectal fossa?
    A. CT
    B. Ultrasonography
    C. MRI
    D. Radiography
    E. Radionuclide imaging