All sacral and presacral tumors are rare. In one series patients with these tumors were estimated to account for approximately one in 40,000 hospital admissions. Tumors arising from the bone of the sacrum are by far the most frequent sacral tumors; chordomas are the most common and GCTs the second most common. Although sacrococcygeal teratoma is the most common sacral tumor in neonates, it is very rare in adults.

The author conducted an extensive analysis of the existing literature concerning tumors of the sacrum to characterize their clinical manifestations. It was found that an expansive space-occupying lesion of the sacrum usually leads to a specific pattern of clinical signs and symptoms throughout its natural history. This clinical pattern depends on the anatomical location of the lesion within the sacrum, its extension, and whether it compresses or invades neighboring structures. Because the histological type of the lesion plays only a minor role in this context, it is usually difficult or impossible to determine a sacral tumor type based on its clinical presentation alone. However, certain specific manifestations can be attributed to some of the tumor types, and a more general pattern of clinical presentation of an expansive sacral lesion can be elaborated. Local pain with or without pseudoradicular or radicular radiation is the most frequent initial symptom and is usually followed by the manifestation of a lumbosacral sensorimotor deficit; bladder/bowel and/or sexual dysfunction appear throughout the natural course of disease.

**KEY WORDS** • sacrum • tumor • lesion • neurological presentation

**REVIEW OF SACRAL ANATOMY**

**Osseous Structures of the Sacrum**

The sacrum is a complex bone, comprising five sacral vertebrae that have fused. In its center lies the longitudinal sacral canal, which opens caudally posteriorly into the sacral hiatus, an incomplete posterior closure of the S-5 lamina. The thick anterior or pelvic face of the sacrum is concave and contains four right- and left-sided anterior sacral foramina. The posterior face of the sacrum is convex, thin, and has four right- and left-sided posterior sacral foramina. The S1–4 spinous processes have merged to form a median sacral crest, caudal to which lies the sacral hiatus at S-5. The superior face of the sacrum consists of an upper endplate, bearing the L5–S1 disc, a wide ala to the right and left, and the horizontally positioned superior articular process.

**Joints of the Sacrum**

The L5–S1 joint articulates through a disc medially and two facet articulations on each side, allowing for a maximum of 10 to 15° of combined flexion–extension and 5° of rotation on either side. The sacroiliac joints and sacrococcygeal joints are amphiarthroses and have no measurable mobility.

**Ligaments of the Sacrum**

The sacrotuberous and -spinous ligaments connect the sacrum to the ischial tuberosity and ischial spine, respectively, and resist backward rotation of the lower sacrum.

**Abbreviations used in this paper:**

GCT = giant cell tumor; PNET = primitive neuroectodermal tumor.
The dorsal sacroiliac ligament spanning the sacroiliac joint and the interosseous sacroiliac ligaments within the sacroiliac joint restrain forward rotation of the upper sacrum. There is also a weak anterior sacroiliac ligament.\(^9\)

**Neurological Structures at the Sacrum**

The dural sac converges within the sacral canal in a craniocaudal direction and ends blindly at the level of S-2. The terminal filum, a prolongation of the pia, extends from the conus medullaris down to the end of the dural sac, where it fuses with the dura mater. It then continues further caudally and attaches to the peristemeum of the first coccygeal segment. The L-5 nerve roots exit the spinal canal through the L5–S1 foramen, passing through a small groove at the anterior surface of the sacral alae. The S1–4 nerve roots exit with their ventral rami through the anterior or sacral foramina, with their dorsal rami through the posterior sacral foramina. The S-5 roots exit at the caudal margin of the sacrum. The L-5 roots more laterally connect with the L-4 roots and then with the S1–3 and part of S-4 roots to form the lumbosacral plexus on each side.

The superior hypogastric plexus lies around the aortic bifurcation and anterior surface of the L-5 vertebra and the upper sacrum, and it connects to the inferior hypogastric plexus, which surrounds the different pelvic viscera. The caudal most portions of the paired sympathetic trunks lie on the pelvic surface of the sacrum, containing four or five ganglia each, and terminate by uniting in the midline to the ganglion impar anterior to the coccyx.\(^1\)\(^1\)\(^8\)\(^4\)\(^8\)\(^9\)\(^2\)

**Neighboring Structures at the Sacrum**

Anterior to the sacrum lies the presacral or retrorectal space, bounded by the rectum anteriorly, the sacrum posteriorly, the peritoneal reflection superiorly at the S2–3 junction, the levator ani and coccygeal muscle inferiorly, and the ureters and iliac vessels laterally.

The median sacral artery and vein from the aortic bifurcation, and to the iliac bifurcation respectively, overlie the sacrum in the midline down to the coccyx. Lateral sacral arteries arise from the internal iliac artery on each side and travel through the anterior sacral foramina, sometimes exiting through the posterior sacral foramina.

On the posterior surface of the sacrum, the erector spinae and multifidus muscles originate, and the pyriform muscle and gluteus maximus muscle originate at the lateral margin of the sacrum.\(^1\)\(^1\)\(^8\)\(^4\)\(^8\)\(^9\)\(^2\)

**CLINICAL MATERIAL AND METHODS**

The readily available English-language literature on sacral tumors was studied to determine the clinical manifestations of this lesion. Because the sacrum can harbor a variety of nonneoplastic tumors in the sense of space-occupying lesions, they were included in the analysis.

**RESULTS**

**General Clinical Manifestations**

The most common initial symptom of a sacral tumor is local pain due to its mass effect.\(^3\)\(^5\)\(^6\) As nerve roots become increasingly compressed or even infiltrated by tumor, radicular pain also develops, radiating uni- or bilaterally into buttocks, posterior thigh or leg, external genitalia, and perineum.\(^3\)\(^5\)\(^6\) This radiating sacral pain is often exacerbated by Valsalva maneuvers\(^6\)\(^9\) and predominates at night.\(^6\)\(^9\) The natural neurological history of an expanding sacral lesion will then be characterized by a single- but usually multiradicular sensory deficit and, at a later stage, motor deficit;\(^6\)\(^7\) eventually, bladder/bowel and/or sexual dysfunction is noted.\(^6\)\(^9\) These latter autonomic dysfunctions may evolve together or separately in any possible order and combination.\(^9\) Involvement of lumbosacral nerve roots in sacral lesions leads to certain specific deficits.\(^3\)\(^5\)\(^6\)\(^7\)\(^9\)\(^6\) A lesion involving the L-5 nerve root, commonly in its L5–S1 foraminal or extraforaminal course, may cause radicular pain and hypesthesias in the lateral thigh and calf as well as dorsum of the foot to the great toe. Motor weakness of the L-5 nerve root may result in weakened ankle dorsiflexion, great toe extension, knee flexion, and hip abduction. The straight-leg raise test is positive. A lesion involving the S-1 nerve root, in its canalicular, S1–2 foraminal or extraforaminal course, typically causes radicular pain and hypesthesias in the posterior thigh and calf as well as at the lateral and plantar face of the foot and the small toe. A motor deficit due to an S-1 lesion may result in weakened ankle plantar flexion, knee flexion, and hip extension. The straight-leg raise test is positive.

A canalicular, foraminal, or extraforaminal lesion of the S-2 nerve root can cause pain and hypesthesia in the posterior thigh or leg, the testicles or labia, and slight weakness of the ankle plantar flexion. Pain and hypesthesia in the outer perianal region and the penis or labia is typical of an S-3 nerve root lesion; inner perianal pain and hypesthesia are associated with an S-4 or S-5 root lesion. A unilateral lesion to the S-2 or S-3 nerve root usually leads to mild or moderate bladder, bowel, and/or sexual dysfunction, although in the case previously mentioned unilateral resection of the S2–4 roots did not result in bladder or bowel dysfunction.\(^9\) A bilateral lesion of the S-2 or S-3 roots always results in complete bladder, bowel, and sexual dysfunction.\(^9\)\(^2\) Unilateral or even bilateral lesions of the S-4 and/or S-5 roots do not result in autonomic dysfunction, even though anatomical work has shown some S-4 and S-5 root contribution to bladder and bowel function.\(^9\) In other words, and this has been shown experimentally by Gunterberg and colleagues\(^9\)\(^0\)\(^4\)\(^2\)\(^4\) bilateral preservation of the S-2 and S-3 roots is required for intact autonomic bladder and bowel function.

Several reflex changes can be observed in sacral tumors at neurological examination. An absent ankle jerk reflex is the most common, resulting from an afferent or efferent S-1 nerve root dysfunction.\(^5\)\(^6\)\(^7\) An absent plantar reflex stems from an afferent S-1 or efferent S-1 or S-2 root lesion.\(^1\) The absence of the bulbocavernosus reflex in men or the sphincter vaginae reflex in women usually reflects an afferent or efferent S-3 or S-4 root lesion.\(^1\) An absent anal reflex is attributed to an afferent or efferent S-4, S-5 or coccyx root lesion.\(^1\)

There are a number of nonneurological clinical manifestations of sacral tumors that arise when the lesions invade neighboring structures. Lateral extension of sacral...
Neurological manifestation of sacral tumors

tumors across the sacroiliac joints cause local pain at the joint, which is exacerbated when upright and walking. Anterior extension of the tumor into the presacral space can compress the rectum impeding bowel movements, bladder emptying, and uterine function, causing dystocia; however, no tumor has ever been observed to cross the presacral fascia and invade the rectum. Invasion of the origin of the gluteus maximus and pyriforme muscles leads to local pain and subsequently decreases hip extension and external rotation power.

Specific Manifestations of Various Sacral Tumors

Chordoma. Chordoma is the most common neoplasm of the sacrum. It is usually malignant, slow growing, locally aggressive tumor that originates from remnants of the notocord, and approximately 50% of all chordomas are located in the sacrum, 35% at the skull base, and 15% in the mobile spine. The mean age of patients at diagnosis is 40 to 70 years. Almost all patients suffer local pain, which is sharp or dull, continuous and frequently located in the rectum. Approximately 50% of patients experience additional sacral radicular pain or leg weakness and/or bladder and bowel dysfunction. A palpable rectal mass is found in the majority of the patients because the lesion commonly expands into the presacral space. The mean duration of symptoms prior to diagnosis is 2 years. The rectum is not involved anteriorly through the presacral fascia. Infiltration of the gluteal muscles has been observed.

Other Primary Malignant Bone Tumors. Ewing sarcoma, chondrosarcoma, osteosarcoma, myeloma, hemangiopericytoma, and lymphoma of the sacrum have been reported. Their clinical presentation is characterized by progressive relentless local pain with radicular lumbosacral radiation, progressive lumbosacral sensorimotor dysfunction, and sphincter dysfunction as well as direct compression of the rectum resulting in constipation. Fever and weight loss, typical symptoms of any malignant tumor, may be present.

GIANT CELL TUMORS. Giant cell tumors are the second most frequent tumor of the sacrum. Between 1 and 8% of GCTs are located in the sacrum, whereas the majority are found in long bones, typically in the distal femur, proximal tibia and distal radius. Giant cell tumors are commonly benign osteolytic, expansive bone tumors, however, local malignant transformation has been observed in up to 16%, and both benign and malignant pulmonary metastases have been found in 3 to 12% of cases. Most sacral GCTs are found eccentrically in the proximal sacrum, but can be located distally as well and are usually very large (diameter 5–11 cm) at diagnosis.

Patients with a sacral GCT are usually between the ages of 20 and 40 years when the diagnosis is established. Sacral GCTs manifest initially with local pain over the sacrum or lumbosacral junction that frequently radiate into the posterior aspect of one or both thighs. The mean duration of pain until diagnosis has been shown to range from 3 to 8 months. Weeks to months after onset of local pain, symptoms and signs of radicular compression appear, depending on the location and extent of the tumor. Because of their local invasiveness, sacral GCTs can cross the sacroiliac joint, causing local joint pain, or the L-5–S1 disc space, causing L-5 nerve root compression, and lead to direct compression of the rectum; most patients present with a palpable mass on rectal examination.

ANEURYSMAL BONE CYSTS. Aneurysmal bone cysts are hypertrophic lytic lesions with surrounding sclerosis and can be found in the sacrum. They usually reach a large size before causing neurological symptoms. Typical features include a 2-year history of local pain with different lumbosacral nerve root compression symptoms and an age between 10 and 20 years.

OTHER BENIGN PRIMARY INTRASACRAL TUMORS. Because of their extreme rarity, the clinical pattern of the following benign primary intrasacral sacral tumors has not been established: chondroblastoma, chondromyxoid fibroma, osteochondroma, osteoid osteoma, osteoblastoma, Paget disease, fibrous dysplasia, eosinophilic granuloma, and hemangioma.

SACRAL EPEMENDYOMA. Sacral ependymomas arise from ependymal cells of the terminal filum, expand the sacral canal, and are usually of the benign myxopapillary type. Local pain with or without radicular lumbosacral radiation is the common initial symptom, typically present in men 30 to 40 years of age. Sensorimotor symptoms and sphincter dysfunction usually follow, resulting in a cauda equina syndrome, and time from initial symptoms to diagnosis is typically 2 to 3 years.

SACRAL SCHWANNOMA. Sacral schwannomas or neurofibromas grow within the sacral canal and only rarely expand through the anterior sacral foramina into the presacral space. Local pain or radicular lumbosacral radiation is the common initial symptom, typically present in women 30 to 40 years of age. Sacral paresthesias and dysesthesias are very frequent, whereas motor symptoms and sphincter dysfunction are rare. Time from onset of initial symptoms to diagnosis is commonly 5 years.

SACRAL MENINGIOMA. Sacral meningiomas are even more rare than sacral ependymomas and schwannomas, arise within the sacral canal, and resemble sacral schwannomas in presentation.

SACROCOCCEYGEAL TERAOMA. Sacrococcygeal teratoma is the most common sacral tumor in neonates, although it is very rare in adults. These tumors are composed of multiple tissues foreign to the tissue in which they arise, and skin, teeth, central nervous system tissue, and respiratory and alimentary mucosa are commonly found in their tissue. They develop during intrauterine growth and prepartum diagnosis can often be made using ultrasonography. Sacrococcygeal teratomas can grow so large that their size causes dystocia. Postpartum, these tumors can usually easily be diagnosed because of a characteristic exophytic mass between the anus and the coccyx, covered by normal skin. Extension into the presacral space can lead to compression of the rectum or bladder. Motor deficits are not present at birth but may develop in childhood because of malignant invasion of the spinal canal. Onset of symptoms in adulthood is exceptional but has been reported.

PRESACRAL NEUROGENIC TUMORS. Neuroblastoma is an in-
completely differentiated tumor derived from embryonic neural crest tissue and clinically manifests in infancy.

Gangliouneuroma, originated from sympathetic ganglion cells, and neurofibroma, originating from sacral nerve roots, present clinically in young adults. These tumors typically cause constipation and urinary retention due to direct compression of the rectum and the bladder.

**Primitive Neuroectodermal Tumors.** Usually found intracranially, PNETs are malignant lesions of undifferentiated cells resembling germinall matrix cells of the embryonic neural tube. They can seed through the subarachnoid space into the spinal canal. One case of a sacral PNET has been reported, which was a primary PNET arising from the right S-1 nerve root, eroding the neural foramen and the sacral canal. Compression of the thecal sac resulted in a progressive S-1 sensorimotor deficit and then bladder and bowel dysfunction.

**Carcinoid Tumor.** Carcinoid tumor of the sacrum is extremely rare and limited to single case reports. It derives from endocrine gastrointestinal amine precursor uptake and decarboxylation cells, and is typically located in the appendix, ileum, lung, and rectum and can cause so-called “carcinoid syndrome” with flushing, wheezing, watery diarrhea, abdominal pain, heart failure and edema because of its release of serotonin and bradykinin.

A carcinoid tumor in the sacrum/presacral space can be a metastatic disease or associated with a rest of enteric tissue. Bladder and/or bowel dysfunction has been described as clinical presentation, but not the carcinoid syndrome.

**Amyloid of the Sacrum.** Amyloid tumors usually occur in the context of multiple myeloma or plasma cell dyscrasia. One case of a 10-cm-diameter primary sacral amyloid has been described.

**Sacral Metastases.** The clinical presentation of sacral bone metastases cannot be unified and follows the unspecific clinical pattern as mentioned previously. The history of a known primary and general symptoms such as weight loss and fever raise the suspicion of a secondary sacral lesion in this context.

**Posterior Sacral Meningocele, Myelomeningocele, and Lipomyelomeningocele.** Spinal dysraphism is the most common congenital anomaly in neurosurgery. If a myelomeningocele is present, asymmetrical sensory or motor deficits, some form of neurogenic bladder dysfunction, and often hydrocephalus in the context of a Chiari Type II malformation, are found immediately after birth. In a lipomyelomeningocele a fatty mass is attached to the conus medullaris or terminale filum. It can manifest at birth as a neurological deficit similar to myelomeningocele; it can also manifest in childhood or young adulthood as progressive lower-extremity sensorimotor and sphincter dysfunction due to growth of the lipoma and progressive mechanical compression, or due to progressive tethering of the spinal cord during axial growth of the spine. Lumbosacral cutaneous abnormalities such as a subcutaneous lipoma, hemangioma, hairy patch, dimples, sinus tract, or skin tag are typical, as are orthopedic foot deformities and scoliosis.

**Anterior Sacral Meningocele.** An anterior sacral meningocele is a cerebrospinal fluid-filled protrusion of the meninges through a defect in the anterior sacrum. It can cause chronic constipation, urinary obstruction or frequency, and dysostotic or dysmenorrhea due to direct mechanical compression of the rectum, bladder, and uterus, respectively. Neurological symptoms include bladder and anal sphincter dysfunction and leg pain and paresthesias caused by compression of the lumbosacral plexus, respectively. Shift of cerebrospinal fluid from the anterior sacral meningocele into the spinal dural sac during bowel movements has been associated with headache.

**Benign Sacral Meningeal Cysts.** Benign sacral meningeal cysts are frequent coincidental findings in the radiological examination of the sacrum, and their pathogenesis is poorly understood. A familial tendency has been described. In the most common classification they are divided into extradural meningeal cysts without nerve root fibers, extradural meningeal cysts with nerve root fibers, and intradural meningel cysts.

Benign sacral meningeal cysts may cause symptoms typically in the third or fourth decade and manifest as of local pain due to osseous erosion as well as sacral radicular pain and paresthesias, especially in the perineal region. Sphincter dysfunction and sensory loss or motor weakness are very uncommon.

**CONCLUSIONS**

The most common pattern of neurological manifestation during the natural course of a sacral tumor is initial local pain with or without pseudoradicular or radicular lumbosacral irradiation; this is followed by lumbosacral sensorimotor deficit and finally by bladder and/or bowel and/or sexual dysfunction.

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Neurological manifestation of sacral tumors

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