Recent Advances in the Diagnosis and Treatment of Presacral Tumours

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Abstract

Presacral or retrorectal tumours (PST) s is rare lesions, which range from cysts (usually benign) to malignant masses, invading surrounding tissues in the pelvis. According to the tumour’s origin, characteristics and behavior, PSTs are classified as follows: congenital, neurogenic, osseous, miscellaneous, and inflammatory. They present variable signs and symptoms, a fact that may delay their diagnosis and/or cause an inappropriate treatment with bad prognosis. Nonetheless, modern imaging modalities, such as CT and MRI may discover the true nature of a lesion, as well as whether it infiltrates neighboring viscera. In that way, they substantially contribute to a definitive diagnosis and a correct preoperative planning. While neoadjuvant chemotherapy and postoperative radiotherapy may offer benefits in certain PSTs, surgical resection is the primary therapeutic management. The anterior, posterior and combined abdominosacral approach are applied, according to tumour location and its relation to adjacent structures. In general, clinicians should maintain high clinical suspicion of the disease to avoid delayed or false diagnosis. Multidisciplinary approach is crucial for a prompt and accurate treatment.

Keywords: Presacral; Retrorectal; Retroperitoneum; Rectum; Sacrum

Introduction

Presacral tumours (PST) s or retrorectal as they are also termed, represent heterogeneous and rare lesions, which may range from benign cysts to malignant masses that can infiltrate surrounding pelvic tissues and organs. Their incidence in the general population worldwide is unknown, because the majority of reports concerning these neoplasms come from tertiary medical centers [1]. Notably, the only case series on the disease not from a referral center was published by Uhlig and Johnson in 1975, demonstrating an incidence of two PSTs per year in the metropolitan population of Portland, USA [2]. Furthermore, Jao et al. [3] from Mayo Clinic concluded in 1985 that the disease is diagnosed in 1 patient for every 40000 hospital admissions.

PSTs are usually deficient of signs and symptoms, until they reach a considerable size, leading to a delayed diagnosis and thus the involvement of other sensitive structures and bad prognosis. However, modern imaging modalities, new surgical approaches and the progress in adjuvant therapy have contributed to a better management of PSTs, and decreased morbidity [4]. The following text attempts to summarize recent medical knowledge on these lesions through an in depth analysis of up-to-date medical literature.

Anatomy-Physiology

The presacral or retrorectal space represents the continuation of the retroperitoneum into the pelvis. This potential space is located between two anatomical structures, the presacral fascia of the sacrum (Waldeyer’s fascia) and the parietal peritoneum of the posterior abdominal wall. Its boundaries are formed anteriorly by the mesorectum, posteriorly by the anterior aspect of the sacrum, superiorly by the peritoneal reflection and inferiorly by the retrosacral fascia [1,5]. Laterally, the presacral area extends to the ureters, the internal iliac vessels, the lateral sacral artery, the sympathetic trunk, the hypogastric nerves, and the inferior hypogastric plexus at the lower levels. Embryologically, the presacral space is the area where fusion of the hindgut and the neuroectoderm of the spinal cord occur. In adults, this site contains retrorectal fat, loose connective tissue, lymph nodes, the median sacral vessels, the superior rectal vessels, as well as sympathetic and parasympathetic branches [5,6].

In case these neural and vascular structures are harmed or injured, the rectoanal physiology is seriously affected and may cause substantial musculoskeletal and/or neurologic morbidity.
# Table 1: Classification of presacral tumours [1,6,11,12].

<table>
<thead>
<tr>
<th>Type</th>
<th>Classification</th>
<th>Benign/Malignant</th>
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<tbody>
<tr>
<td>Cystic</td>
<td>Developmental Cysts</td>
<td>Benign</td>
</tr>
<tr>
<td></td>
<td>Epidermoid cyst</td>
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<td></td>
<td>Dermoid cyst</td>
<td>Benign</td>
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<tr>
<td></td>
<td>Enterogenous cyst</td>
<td>Most are benign</td>
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<tr>
<td></td>
<td>Cystic hamartomas</td>
<td>Benign</td>
</tr>
<tr>
<td></td>
<td>Anterior sacral meningocele</td>
<td>Benign</td>
</tr>
<tr>
<td></td>
<td>Teratoma</td>
<td>Benign (with malignant potential)</td>
</tr>
<tr>
<td>Solid</td>
<td>Chordoma</td>
<td>Malignant</td>
</tr>
<tr>
<td></td>
<td>Adrenal rest tumor</td>
<td>Benign</td>
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<tr>
<td>Neurogenic</td>
<td>Neurofibroma</td>
<td>Benign</td>
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<td></td>
<td>Neurilemoma (schwannoma)</td>
<td>Benign</td>
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<td></td>
<td>Ganglioneuroma</td>
<td>Benign</td>
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<tr>
<td></td>
<td>Neuroblastoma</td>
<td>Malignant</td>
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<td></td>
<td>Ganglioneuroblastoma</td>
<td>Malignant</td>
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<td></td>
<td>Ependymoma</td>
<td>Malignant</td>
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<td></td>
<td>Malignant peripheral nerve sheath tumours</td>
<td>Malignant schwannoma, Neurofibrosarcoma, Neurogenic sarcoma</td>
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<tr>
<td>Osseous</td>
<td>Giant-cell tumor</td>
<td>Benign</td>
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<td></td>
<td>Osteoblastoma</td>
<td>Benign</td>
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<tr>
<td></td>
<td>Aneurysmal bone cyst</td>
<td>Benign</td>
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<td></td>
<td>Osteogenic sarcoma</td>
<td>Malignant</td>
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<td></td>
<td>Ewing sarcoma</td>
<td>Malignant</td>
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<td></td>
<td>Myeloma</td>
<td>Malignant</td>
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<tr>
<td></td>
<td>Chondrosarcoma</td>
<td>Malignant</td>
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<tr>
<td>Miscellaneous</td>
<td>Lipoma</td>
<td>Benign</td>
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<tr>
<td></td>
<td>Fibroma</td>
<td>Benign</td>
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<td></td>
<td>Leiomyoma</td>
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<td></td>
<td>Hemangioma</td>
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<td></td>
<td>Endothelioma</td>
<td>Benign</td>
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<td></td>
<td>Desmoid</td>
<td>Locally aggressive</td>
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<tr>
<td></td>
<td>Liposarcoma</td>
<td>Malignant</td>
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<tr>
<td></td>
<td>Fibrosarcoma</td>
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<td></td>
<td>Malignant histiocytoma</td>
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<tr>
<td></td>
<td>Leiomyosarcoma</td>
<td>Malignant</td>
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<td></td>
<td>Hemangiopericytoma</td>
<td>Malignant</td>
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<td></td>
<td>Metastatic adenocarcinoma</td>
<td>Malignant</td>
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<tr>
<td>Inflammatory</td>
<td>Perineal or pelvirectal abscess</td>
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<td></td>
<td>Endometriosis</td>
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<td>Foreign body granuloma</td>
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<td></td>
<td>Infectious granulomas</td>
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<td></td>
<td>Diverticulitis</td>
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<tr>
<td></td>
<td>Crohn’s disease</td>
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<tr>
<td>Other</td>
<td>Pelvic ectopic kidney</td>
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<td></td>
<td>Hematoma</td>
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<td>Abscess</td>
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Notably, if the S3 nerve is injured bilaterally, the external sphincter malfunctions, not contracting when the rectum is dilated, leading to various degrees of incontinence. Interestingly, anorectal function is maintained if all unilateral nerve roots are sacrificed. Moreover, during sacrectomy, pelvic stability is maintained if greater than half of the body of S1 vertebra remains intact. However, if the site is radiated preoperatively, spinopelvic stability may be seriously compromised [1,7,8].

**Classification**

Various tumours, either congenital or acquired, may arise from and within the structures of the presacral space. A variety of classification systems attempting to categorize PSTs have been proposed by different authors, although none has been unanimously accepted [9]. The classification system first described by Uhlig and Johnson is the most frequently used one. According to the tumour’s origin this system includes the following categories: congenital, neurogenic, osseous, inflammatory and miscellaneous [2]. Another important aspect for tumour classification is its benign or malignant behavior. The latter is usually encountered in solid lesions. Lev-Chelouche et al. [10] have classified PSTs into congenital versus acquired and benign versus malignant. Metastatic and locally advanced colorectal and genitourinary neoplasms are usually not included in the category of presacral tumours (Table I).

### Clinical Manifestation

#### Congenital

Congenital tumours develop from embryonic tissue remnants and are usually benign. They may appear either as cystic or solid lesions. The former include developmental cysts and anterior meningoceles, while the latter include teratomas, sacrococcygeal chordomas, and adrenal rest tumours. Congenital tumours are the most common retrorectal lesions, 55% to 70% of all lesions in the presacral area, and the patients are usually females [11,12].

1. Cystic
   2. Developmental Cysts

**Epidermoid and dermoid cysts, enterogenous cysts (rectal duplication cysts) and cystic hamartomas (tailgut cysts)** fall under this category. They represent 60% of all congenital presacral lesions. Embryologically, they may develop from any of the three germ layers [11]. Epithelium always lines the developmental cysts (squamous for dermoid and epidermoid cysts, cuboidal, transitional or columnar epithelium for enterogenous cysts and cystic hamartomas). These lesions are usually multilocular and their walls are surrounded by fibres of disorganized smooth muscle [12]. Developmental cysts may be complicated with hemorrhage, infection or malignant degeneration. The treatment of choice is complete surgical excision of the cystic epithelial lining [13].

**Dermoid and epidermoid cysts**: Usually, benign they mainly affect middle-aged females. They arise when the ectodermal tube fails to close normally. Both types are constituted of stratified squamous epithelium. Moreover, dermoid cysts have skin appendages (sweat glands, sebaceous cysts, hair follicles), which is a useful feature for their differential diagnosis from epidermoid cysts. Dermoid, as well as epidermoid cysts, may communicate with the skin; if this is the case, a postanal dimple or sinus may also exist, representing a residual connection to the embryonic ectoderm [4,6]. Up to 30% of these cysts may be infected, impeding their differentiation from a complex perirectal abscess [11].

**Enterogenous cysts (Duplication)**: They develop from sequestration of the embryonic hindgut, which classifies them as endodermal. Columnar, transitional, cuboidal or squamous epithelium may line them. They often appear as multilobular, with a single dominant cyst and multiple satellite ones. Diagnostic criteria are continuity with the rectum, a well-defined muscular wall with a myenteric plexus and mucosal lining. Similarly with epidermoid and dermoid cysts, enterogenous cysts are more common in females and may become infected. Commonly, they are benign in nature, but malignant transformation should not be ruled out [1,6].

**Cystic hamartomas (Tailgut cysts)**: They arise from the hindgut, due to regression failure of a portion of the embryonic tail. Although multicystic and well-circumscribed in appearance, they do not have a capsule. Usually, a cystic hamartoma appears like a soft mass with thick walls filled with mucous. Histologically, they are similar to the intestinal tract, including squamous, columnar or transitional epithelium. These lesions mainly affect females and are benign in nature; however, malignant degeneration (most commonly adenocarcinoma) has been reported [14-16].

**Anterior sacral meningocele**: These rare lesions (5%) develop through a defect in the anterior sacrum that allows herniation of the dural sac [17]. This defect may appear in combination with presacral lipomas and/or cysts and may be associated with other congenital abnormalities, such as spina bifida, tethered spinal cord, urinary tract or anal malformations, and uterine or vaginal duplication [6]. In adults, the female-to-male ratio is 6:1, while it is 2:1 for children [18]. When the sac contains neural elements, it is then called myelomeningocele [19]. Anterior sacral meningocele is usually of sporadic occurrence, but it also may be part of the hereditary Currarino syndrome. This rare syndrome is caused by congenital caudal anomalies and its three main characteristics (Currarino triad), are a deformity of the sacral bone, malformations of the anus and rectum, and a presacral mass [20]. It may also be encountered in conditions with duralactasia such as Marfan’s syndrome and neurofibromatosis type 1, but this is a very rare case [21]. The “scimitar sign” is pathognomonic of the disease, and it describes a sacrum with a rounded concave border, although without destruction of the bone on plain radiograph. The continuity between the dural sac and subdural space means that the sac also contains cerebrospinal fluid. An anterior sacral meningocele may increase the pressure of the cerebrospinal fluid, giving rise to typical presenting symptoms, such as headache and defecation, associated with nausea and vomiting triggered by changes in body position. Secondary symptoms arise from compression of neighboring structures, such as constipation, urinary abnormalities and lower back pain. Aspiration or biopsy of the lesion should not be attempted, due to the risk of life-threatening meningitis, intracranial hypotension syndrome or even death.

**Teratomas**: Presacral teratomas are rare in adult life, although they represent the most common teratomas in the newborns. They arise from totipotential cells, and as a result these tumors contain tissue elements from all three germ layers (epithelium of the gastrointestinal tract, respiratory tract, and nervous system). They may be cystic, solid, or both, but most are cystic and benign [1,22]. Teratomas are categorized as mature, immature, or malignant (teratocarcinoma). The first contain recognizable epithelial or mesenchymal cells, while the second are comprised of endodermal, mesodermal or ectodermal elements. Germ cells are found in malignant teratomas, unlike the
teratomas with malignant transformation, which include malignant degenerated somatic cells [23]. Teratomas are more common in female patients, with the female to male ratio being 10:1. In pediatric patients, only 4% of the teratomas diagnosed at birth are malignant; those diagnosed later in childhood tend to be aggressive and are linked to poor prognosis. Generally, the risk of malignancy increases with age. Teratomas in children are believed to harbor some association with anomalies of the anorectum, urinary tract, and vertebrae. In the adult population, around 30% of teratomas have elements of malignancy at the time of resection, and there is a risk of 40% to 50% malignant transformation, which increases with incomplete resection. Teratomas often adhere to the coccyx; in that case, en bloc coccygectomy is the required surgical approach. Recurrence rate is 7.5% to 22%, and, when the coccyx is not respected, this rate may increase up to 37%.

**Solid**

**Sacrococcygeal chordomas:** They arise from the embryonic notochord and are the most common malignant PSTs (30 to 50% of all chordomas occur in this area). They are usually found on the midline cerebrospinal axis and are most commonly located at the sphenoclavial region and the sacrum [11,24]. Chordomas are more common in males and are rarely diagnosed before adulthood. They tend to grow slowly, but are locally invasive and give metastases (liver, bones and lungs) in about 20% of cases. Their macroscopic appearance is a lobulated, gelatinous mass, brownish-grey in color, commonly with a pseudocapsule. Sometimes they may appear cystic, due to intratumoral hemorrhage. Their characteristic histopathologic finding is the presence of vacuolated and rich in mucin and glycogen physaliphorous cells. Chordomas may cause either specific or nonspecific symptoms, when the tumour has reached a considerable size. By that time, resection on negative margins is much more difficult. Local recurrence is high and 10-year survival rates from 9% to 35% [11,25].

**Neurogenic Tumors**

These arise from pelvic nerve roots or peripheral nerves and are the second most common PSTs, accounting for about 10% of all [12]. The majority (around 85%) is benign including neurofibromas, neurilemmomas (schwannomas), and ganglioneuromas. Inversely, neuroblastomas, ganglioneuroblastomas, ependymomas, and malignant peripheral nerve sheath tumours (malignant schwannomas, neurofibrosarcomas and neurogenic sarcomas) fall under the category of malignant neurogenic lesions. Generally, they grow slowly and present non-specific symptoms. Therefore, they may be of large size at the time of diagnosis, leading to considerable blood loss intraoperatively and high complication rates. In case of symptoms, the route of the affected nerve determines the pain distribution and neurologic dysfunction [26-28].

**Osseous**

They may develop from bone, fibrous tissue, cartilage and marrow. They account for 10% of all PSTs and include benign and malignant lesions. Giant-cell tumour, osteoblastoma, and aneurismal bone cyst are benign, while osteogenic sarcoma, Ewing sarcoma, myeloma, and chondrosarcoma are malignant. They usually grow rapidly and are of significant size at the time of diagnosis; consequently, bone destruction or soft tissue calcification may usually be identified on plain radiograph. The lungs are their primary metastatic target and their overall prognosis is poor [6,12].

**Inflammatory and Miscellaneous**

Miscellaneous tumors represent 10% to 25% of all presacral ones. They include lipoma, fibroma, leiomyoma, hemangioma, endothelioma, desmoid (locally aggressive) (Figure 1), liposarcoma, fibrosarcoma, malignant histiocytoma, leiomysarcomas, hemangiopericytoma, metastatic adenocarcinoma, and inflammatory tumours. Although Uhlig and Johnson originally classified inflammatory tumors as a separate category of presacral ones, they should be considered secondary reactions to foreign substances (e.g., foreign body granulomas from barium leaks or suture material), or extensions of infectious processes from either the abdomen or perirectal space (e.g., pelvic sepsis, Crohn disease, and perforated diverticulitis).

**Clinical Manifestations**

**Symptoms**

PSTs, especially benign ones, are either asymptomatic for a long time, or they produce minimal or non-specific symptoms, such as constipation, paradoxal diarrhea, rectal tenesmus and sexual dysfunction. Most of the symptoms occur due to tumour compression on the rectum or other neighboring structures [11]. Pain occurs primarily in malignant tumours [29], in male patients older than 60 years [11] or in benign infected tumours [12]. The latter may also cause perianal discharge or drainage. Kye BH and Macafee DA report lower abdominal pain or discomfort as the most common symptom at first visit [30,31], whereas Menteg BB et al. [23] describe rectal pain and perirectal mass sensation as the main symptoms in their respective case series. When nerve roots or the sacral plexus are invaded, urinary incontinence or retention and bowel incontinence may occur [12]. Obstructive labor by the presence of a PST has been reported in women of reproductive age and this is an absolute indication for surgical resection, even if the tumour is benign or asymptomatic. Interestingly, some tumours produce more specific symptoms. A constant aching of mild intensity in the lower lumbar, pelvic and/or gluteal region is associated with chordomas, while an anterior sacral meningocele is linked to headache during defecation or intercourse, due to increased intracranial pressure.
Physical examination

PSTs are usually discovered randomly during routine physical examination (pelvic or rectal). The latter may determine the cranial extent of the lesion, as well as whether it is fixed or freely mobile, and its relationship to other pelvic structures. On palpation, PSTs usually feel soft and easy to compress. When tender, the tumor might be an infected developmental cyst or a primary perirectal abscess that extends supraregionally. A thorough neurologic and musculoskeletal examination must be performed in order to reveal any neurologic deficiency and document preoperative functional status [1,4,12]. Bimanual pelvic examination is very important in female patients to exclude conditions like ovarian or uterine adenocarcinomas, which are much more common conditions. In addition, complete colonoscopy should be performed in all patients, aiming to discover a synchronous colorectal adenocarcinoma.

Imaging

Plain radiographs

The role of plain radiographs has been limited by the use of Computed Tomography (CT) scans and Magnetic Resonance Imaging (MRI), as they are the imaging modalities of choice in diagnosing PSTs nowadays. Plain radiographs are usually normal and have little additional information to offer. However, they may reveal osseous destruction of the sacrum, calcifications, and small bone fragments or teeth in teratomas. The "scimitar sign", as described above, is pathognomonic for anterior sacral meningocele [4,6].

CT and MRI

The use of CT is to reveal any destruction of the bone cortex, the solid or cystic nature of the lesion and whether tumour is infiltrating any neighboring viscera. MRI, being more specific that CT may provide a detailed view of the anatomical correlations and the histology of the tumour [29], as well as neural tissue involvement [32]. Although both CT and MRI can demonstrate the cystic or solid nature of a lesion, it is difficult to determine its benign or malignant characteristics [30]. However, smooth-walled cystic lesions on MRI are normally benign, while solid or heterogeneous ones tend to be malignant [33]. Although CT and MRI are useful for preoperative planning, neither of them can provide a definitive diagnosis [34].

In general, anatomy and topography may be determined preoperatively, using the findings yielded by MRI and CT. The aim is to achieve optimal surgical planning and accurate-successful surgical resection [35,36]. While MRI appears to play a superior role in the diagnosis and preoperative planning of PSTs, both CT and MRI should be used in a complementary and not mutually exclusive manner [4,37].

Transrectal ultrasound

Another useful imaging tool in the diagnostic process is transrectal ultrasonography (TRUS). Most importantly, this modality presents a sensitivity of 100%, when combined with rigid proctoscopy. TRUS may aid to determine whether a lesion is cystic or solid, as well as whether rectal involvement is present. Normal TRUS findings eliminate the possibility of a PST, with the precondition that an experienced examiner is involved [6,29].

Other imaging techniques

Angiograms, venograms and fistulograms have occasionally been reported to play a supplementary role in the diagnosis and management of PSTs. In case there is tissue distortion due to mass effect by the neoplasm, an angiogram and/or venogram may be added to MRI (MR angiogram and venogram), in order to determine vascular anatomy and involvement. In patients that present with a chronically draining sinus, fistulograms may be useful to investigate the possibility of a developmental cyst [38]. In any case, imaging modalities aim to guide towards the correct surgical approach (anterior, posterior, or combined), as well as to determine the intraoperative extent of excision (local or en bloc resection).

Preoperative Biopsy

Preoperative biopsy in the management of PSTs appears to be rather "controversial", due to its potential complications and the accuracy of modern imaging techniques [39]. Highlighting the risk of infection of cystic lesions and needle-tract seeding with malignant cells (in case of malignant tumors), surgeons and radiologists have traditionally been discouraged from performing preoperative biopsy on lesions that are surgically excisable, as biopsy rarely affects the necessity of surgical intervention [10,11]. However, the scenery seems to change recently, with authors suggesting that biopsy may have a role for patients with malignant tumours that would benefit from adjuvant therapy. The suggested route of biopsy seems to be transperineal or presacral. The argument for this is that, in case of malignant lesions, the biopsy tract must be respected en bloc with the specimen to reduce the risk of recurrence in that tract [39,40]. Therefore, transanal, transvaginal, transperitoneal and transretroperitoneal routes should be excluded [1]. Yang BL et al. [33], in their exploration of the benefits of preoperative use of MRI, claim that the risk of a routine biopsy should be avoided and its indications limited to patients with metastatic disease or lymphoma. Rising to the defense of this argument, Macafee et al. [31] suggest that careful clinical evaluation and preoperative planning utilizing MRI combined with avoidance of routine preoperative biopsy, result in a good postoperative outcome.

A large retrospective study from Mayo Clinic Rochester, USA, reviewed all patients who underwent biopsy of PSTs, with the primary outcomes measured being the complications that were related to the biopsy, as well as the precision of preoperative imaging and biopsy, compared to the final pathology. It was concluded that preoperative biopsy is safe and concurrent with postoperative pathology in comparison with imaging. Consequently, percutaneous preoperative biopsy should be performed, so as to guide decisions, in view of the substantial differences in therapeutic approach for benign versus malignant solid PSTs and the current limitations of imaging [39].

Preoperative Therapy

Neoadjuvant chemotherapy may offer substantial benefits in certain PSTs, such as Ewing sarcomas and osteogenic sarcomas. Furthermore, the tyrosin kinase inhibitor Imatinib has been demonstrated to increase disease-free survival time, in cases of advanced chordomas. Similarly, the epidermal growth factor inhibitors Gefitinib and Cetuximab, have been shown to cause favorable results in patients with recurrent and metastatic chordomas [41,42].

Encouraging results have also been reported for chordomas, via transcatheter arterial embolization. This technique may be used in the immediate preoperative time, leading to decreased blood loss during the operation and facilitating total resection of the tumour [43,44].

Radiotherapy has an unclear role in the management of
PSTs. Generally, chordomas and other PSTs are considered to be radioresistant, although research on preoperative radiation on PSTs has been actively continued. Local control of these tumors may be achieved through recent modalities, including fractionated irradiation with charged particle carbon ion radiotherapy. Radiotherapy aims to aid surgical resection decreasing tumour size, treat tumour recurrence and clean infiltrated surgical margins [6,45].

**Surgical Approach**

Undoubtedly, surgical resection is the treatment of choice for all PSTs, even if they are asymptomatic. The reasons for that may be summarized as follows [1,4]:

1. Spontaneous infection is possible for cystic lesions, a condition that complicates resection. The possibility of recurrence is also increased and a second operation might be deemed necessary.
2. PSTs may hinder natural vaginal delivery in women of reproductive age.
3. Malignancy may lurk even in cystic lesions that appear benign.
4. There is a chance of malignant degeneration for teratomas.
5. PSTs may cause discomfort of varying degree and their excision substantially contributes to the improvement of the patient’s quality of life.

The surgical excision may be accomplice through three approaches: anterior, posterior and combined abdominosacral. The selection of a certain approach depends on tumour characteristics (nature, size and location). The potential infiltration of the sacrum, pelvic sidewall and adjacent structures should also be taken into consideration. Lesions that extend above S4 level are usually respected through the anterior approach, while lower lesions through the posterior one. In case the upper extent of the lesion is palpable on rectal examination, it is possible for the lesion to be resected transsacrally [46,47]. The combined approach is reserved for lesions that are larger or located in an intermediate position [48].

In any case, a multidisciplinary team including colorectal surgeons, neurosurgeons, orthopaedic and plastic surgeons should be activated, assess the various operative approaches and make the optimal decision [9]. Multidisciplinary collaboration raises the resection rate of the tumours, preventing unnecessary injuries and improves the prognosis and patient’s quality of life [48].

**Anterior approach (Transabdominal)**

This is selected when the tumour is large, located above the level of S3 or S4 and no nerves are involved (Figure 2). The patient is placed in lithotomy position. A median abdominal incision is performed, granting the surgeon a good view of the pelvic structures, iliac vessels and ureters. The retrorectal space is then dissected, the sigmoid colon is mobilized, the rectum is pulled to the front and excision of the tumour follows. It is highly important to protect and preserve the mesorectum as well as the rectal vessels. Since the middle sacral blood vessels and the presacral venous plexus are located in this region, they should be treated with care to avoid presacral hemorrhage. The ureter and main nervous branches in the retrorectal region should be kept safe from injury [9,47,48]. Usually, laparotomy is performed, but several recent papers have proposed laparoscopic surgery, which may yield equally satisfactory results, minimizing tissue trauma, especially when malignancy has been ruled out. Moreover, it is associated with low rates of postoperative complications, short hospital stay, no neurological dysfunction and satisfactory long-term results [49,50].

Generally, the anterior approach offers good visualization of the tumour’s location and extent, as well as its relationships with the surrounding structures, which is essential when performing maneuvers of total excision [51].

**Posterior approach**

This includes the transacral, transsacroccocygeal, transphincteric, transrectal and transanorectal techniques. Each of them has its own advantages and disadvantages and their use depends on the nature and peculiarities of the tumour, as well as the surgeon’s experience. The posterior approach is preferred for benign tumors that do not exceed 8 cm in diameter or for perineal fistulae located below the level of S4. Notably, it offers good access of the caudal section of the tumour [47]. The patient takes the Kraske position. An “S”-shaped or longitudinal incision is made at the level of the S3 vertebra, with precaution to avoid damaging the anal sphincter complex. Resection of the coccyx and distal sacrectomy may be performed, so as to attain better exposure of the presacral area. If sacrectomy is deemed necessary, at least one side of S2 must be maintained in order to prevent bowel and urinary disturbance [9,52].

When complete exposition of the tumor is achieved, digital rectal examination should be performed to attest the extension of the tumour [48]. The lesion is dissected from adjacent structures, including the rectal wall, which in most of the cases is not involved. When dealing with benign lesions, a fat plane is usually encountered between the lesion and the mesorectum, which makes the dissection easier [1]. In the case of very small, cystic lesions, the following maneuver may be helpful: the surgeon double-gloving their non dominant hand, inserts the index finger in the anal canal and the lower one-third of the rectum, and then applies pressure on the lesion, thus compelling it out towards the incision. In that way dissection of the lesion off the rectal wall is achieved and iatrogenic injury to the rectal wall is prevented. However, if the lesion has been infected, this dissection may be difficult, especially if the plane between the lesion and rectum is eradicated. In case secure separation of the lesion cannot be performed due to its adherence to the rectum, a portion of the rectal wall should be removed along with the lesion and the defect repaired in two layers. While routine coccygectomy used to be recommended, particularly in the case of teratomas, this seems to be no longer the
case, unless there is direct invasion of the coccyx by a malignant lesion or a lesion of uncertain malignant potential [12].

Importantly, the posterior approach is associated with certain disadvantages, including the deficient control over the pelvic vessels and the risk of injury to the lateral pelvic nerves. Careful selection of the cases may minimize these downsides [9].

**Combined abdominosacral approach (Abdominoperineal)**

A combination of the two aforementioned approaches is preferred, when the distal margin of the tumour is lower than S3 and the cephalic margin higher [33]. Furthermore, this approach is applied to cases where the neoplasms invade the rectum or adjacent structures. It is often the case that the presacral vessels are involved and should be ligated. It is also the choice in the case of infected cysts that involve the rectum or the presacral fascia. Then, normal planes are often unclear and many adhesions to the adjacent tissues complicate the operation [47].

This approach includes initially a lower midline laparotomy with the patient in the “sloppy lateral position”, permitting both abdominal and sacral access. The retrorectal space is investigated following the mobilization of the sigmoid colon, and the tumour is dissected from the mesorectum. Similarly, the tumour is separated posteriorly from the presacral fascia if possible. In cases en bloc resection is mandatory, due to tumour size or extensive infiltration, S3 should be preserved, or a colostomy should be created for malignant tumours. To minimize blood loss during the abdominal portion of the operation, careful ligation of middle and lateral sacral vessels should be performed, as well as internal vessels. However, the anterior division of the internal iliac artery must remain intact, to avoid perineal necrosis [4,6].

The next portion is initiated through a sacrococcygeal incision, with respect to the external sphincter. Rectal resection is completed, in case this was not performed during the abdominal portion. Moreover, the anal canal and the anus are removed in case this is mandatory. Suction drains are placed where appropriate and the perineal wound is sutured in layers [4].

Distinct benefits linked to the combined approach, may be the better visualization of structures through the anterior incision, as well as the improved exposure of the nerve roots via the posterior approach. For all three approaches intraoperative digital rectal examination is vital to avoid rectum injury, while dissociating the tumor. For trans-sacral and combined approaches, at least unilateral S3 and all of the S1-S2 nerve roots should be reserved and protected [47].

**Discussion**

PSTs are rare tumours with variable signs and symptoms. This fact may lead to delayed diagnosis, inappropriate treatment and thus bad prognosis. However, modern imaging modalities have substantially contributed to a prompt and accurate diagnosis, as well as better preoperative planning via a detailed description of tumour nature (solid or cystic) and extent. Moreover, preoperative biopsy may support the appropriate management of heterogeneously cystic and solid lesions [4].

Adjuvant chemotherapy and tyrosine kinase inhibitors and/or epidermal growth factor inhibitors may decrease PSTs, such as sarcomas and chordomas. Conversely, radiotherapy has an unclear role, due to the radioresistant nature of these tumours. However, high recurrence rates of malignant PSTs could be treated through modern radiotherapy, although the available data are still limited. Surgical management remains the primary and definite therapeutic approach for PSTs and numerous approaches have been described, aiming to a safer-more radical tumour resection. New instruments are available for the surgeons to use nowadays, while laparoscopic techniques are also performed [1,6].

Surgical resection of benign PSTs may offer 100% overall survival. Nonetheless, recurrence rates range according to the completeness of resection. Lev-Chelouche et al. [10] from Israel presented data on 21 benign PST cases, where no recurrence was discovered in 10 years of postoperative follow-up. Similarly, Glasgow et al. [29] from USA recorded no recurrence in such patients after a follow-up of 22 months. The same results were reported recently from Maddah et al. [50] from Iran for 29 benign cases and mean follow-up of 56 months.

Malignant PSTs offer variable rates of survival and recurrence, according to tumour biology, surgical resection and seedling of operative field with cancer cells. Moreover, adjuvant chemotherapy may contribute to better prognosis, while radiotherapy may decrease recurrence risk. Wang et al. [51] from Taiwan analyzed 22 cases of malignant PSTs, including chordomas and leiomyosarcomas and estimated a 5-year survival rate of 41%. Similarly, Lev-Chelouche et al. [10] investigated 21 cases of such lesions and presented 67% recurrence and 50% survival rate. Furthermore, Bergh et al. [52] studied 39 patients with chordoma and reported 44% recurrence and 84% 10-year survival rate. Interestingly, Kaiser et al. [53] concluded that local recurrence rate may increase from 28% to 64%, when sacrococcygeal chordomas were violated during the operation. Survival rates were extensively evaluated by McMaster et al. [54] from USA, through nine registries from 1973-1995 within the National Cancer Institute’s Surveillance, Epidemiology and End Result program (NSEER). 400 chordoma cases were reviewed and 5-year and 10-year survival rates were calculated at 74% and 32% respectively.

**Conclusion**

Active clinical and experimental research may aid in the development of new neoadjuvant chemotherapeutic agents, as well as new radiation modalities. Carbon ion radiotherapy may constitute a potential advancement in the field of PST therapy, as well as preoperative arterial embolization. Similarly, progress in laparoscopic and robotic surgery may open new horizons in PST surgery. Overall, presenting symptoms in PSTs are nonspecific, while the literature reveals a wide range of variability in physical findings. It is of great importance that clinicians maintain high clinical acumen and remain cognisant of the range of symptoms linked to PSTs, in order to avoid delayed or false diagnosis. It is noteworthy that accurate-prompt diagnosis and successful treatment of PSTs demand active involvement and cooperation of multiple medical specialties, with the surgeons in the leading role.

**References**

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