ORIGINAL ARTICLE

Vestigial retrorectal cystic tumors in adults: A review of 30 cases

Tumeurs kystiques vestigiales rétrorectales de l’adulte : une série de 30 cas

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Summary
Objectives. — Retrorectal tumors are uncommon. This is a report on a series of vestigial retrorectal cystic tumors in adults that were surgically removed at two medical centers in France. We also assessed the significance of imaging and, in particular, magnetic resonance imaging (MRI) in the management of these lesions.

Methods. — The medical files of adult patients operated on for vestigial retrorectal cystic tumors over the past 15 years were retrospectively studied and, in particular, the radiological studies, the treatment and the histopathology.

Results. — Thirty patients underwent surgery for vestigial retrorectal cystic tumor. A preoperative diagnosis was possible in almost all cases. The surgical procedure was justified by preoperative imaging and included the transanal approach (three cases), posterior approach (23 cases), anterior approach (two cases) and combined posterior and anterior approach (two cases). Imaging can identify a multilobular lesion that may require the use of a coccygeal approach to achieve complete resection. During a mean follow-up of 3.2 years (range 0.5–15 years), two patients had a local recurrence (successfully surgically removed), and a further two patients were lost to follow-up. Only one lesion was malignant.

Conclusions. — Preoperative imaging, and especially magnetic resonance imaging, enables both a specific diagnosis and the selection of the optimal surgical procedure for the treatment of vestigial retrorectal cystic tumors in adults.

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Résumé

Objectifs. — Présenter une série homogène de 30 tumeurs vestigiales chez l’adulte opérées dans deux centres et l’intérêt de l’imagerie par résonance magnétique (IRM) au plan diagnostique.

Patients et méthodes. — Les dossiers des 30 patients, opérés sur les 15 dernières années d’une tumeur vestigiale rétroméssinaire, ont été réévalués de façon rétrospective avec relecture des lames et étude comparative avec l’imagerie IRM.

Résultats. — Le choix de la voie d’abord a bénéficié de l’imagerie préopératoire et en particulier de l’IRM : voie endoanale (trois cas), voie rétroméssinaire (23 cas), voie abdominale (deux cas), voie double abdominale et périméssinaire (deux cas). L’IRM permet de suspecter la malignité et de détecter le caractère multiloculaire de certaines lésions conduisant à pratiquer parfois une résection du coccyx pour une exérèse complète. La série ne comporte aucun décès, le taux de morbidité était de 20 % et sur un suivi moyen de 3,2 ans (extrême 0,5–15 ans), deux patients ont présenté une récidive locale réséquée une nouvelle fois avec succès, deux patients ont été perdus de vue et une seule tumeur s’est révélée maligne.

Conclusions. — L’étude rétrospective de 30 tumeurs vestigiales insistant sur l’apport de l’IRM permet d’affiner la démarche diagnostique pour une pathologie rare mais qu’il faut aborder judicieusement.

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Introduction

Retrorectal tumors in adults are uncommon (Jao et al. [1] reported an incidence of 1/40,000), and two-thirds of such cases are congenital. Among these congenital retrorectal tumors, 60% are vestigial cysts. We report here on a series of 30 patients with retrorectal vestigial tumors treated between 1989 and 2005 at two French medical centers, including the contribution of magnetic resonance imaging (MRI) in the diagnosis and treatment of these lesions.

Methods

We, retrospectively, studied the radiological studies from the medical files of 30 patients treated within the last 15 years for vestigial retrorectal cyst at two surgical centers (Besançon and Lyon, France). The radiological studies included conventional biplane radiography, endorectal ultrasonography (EUS), computed tomography (CT) and magnetic resonance imaging (MRI). The long period of time covered by the study meant that it was not possible to compare the different radiological methods.

Treatment was always surgical, and the objective was to remove the entire lesion as revealed by the radiological study (particularly in the case of multilobular lesions) to avoid any recurrence of disease and malignant degeneration, but with the least amount of consequent impairment. The surgery used the transanal approach, posterior approach, abdominal laparotomy or a combination of abdominal and posterior approaches.

The colon was mechanically prepared the day before, and the rectum cleansed with an antiseptic solution. For the posterior approach, the patient was placed in a prone position; the incision was paramedian with dissection of the tip of the coccyx and opening of the median raphe to allow a clear view of the posterior wall of the rectum. On occasions, the insertions of the gluteus major muscle on the sacrum were severed and the coccyx resected.

The histopathological results were reexamined and, retrospectively, compared with MRI scans specifically for the present study. For the histopathological analyses, the classification based on that of Malafosse et al. [2] was used; this divided vestigial cysts into two groups depending of their origin, with epidermoid and tailgut cysts, and rectal duplication in one group, and teratomas in the other.

Results

Patients

There were 23 female patients and six male patients, with a mean age of 43 years (range 16–77 years) and a past medical history including:

- unknown neonatal–perineal surgery in one case;
- massive proctorrhagia requiring a blood transfusion in one case;
- surgical treatment of anorectal suppuration in one case;
- surgery for an anterior sacral meningocele in one case.

Only one patient had a medical history of transanal surgical excision of a retrorectal tumor, although the histological findings were not clear.

The lesions were revealed by anoperineal pain in 10 cases, tenesmus in two cases, chronic proctorrhagia in two cases, a perianal tumor in two cases and anoperineal abscess in two cases. In 12 cases, the lesion was revealed by chance during a complete physical examination, including one that was diagnosed during a follow-up of rectal cancer and another following pregnancy.

In each case, the lesion was palpable on rectal examination, allowing its volume, extent and consistency to be estimated. Local examination revealed a postanal dimple in two cases, and there was anal and vulval contact in one case.

Radiology

Conventional radiography (X-rays) was performed on 27 patients and was normal in 23 cases; in the remainder, there was erosion of the sacrum (two patients) and tumor...
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was made possible by EUS and CT. For the seven cases not scanned by MRI, the diagnosis MRI scan also revealed an associated vestigial lesion (one and the rarity of the lesion. With acute suppuration, the protocol because of the retrospective nature of the study only six cases before being abandoned for MRI instead. and a multilocular lesion in the other four. CT was used in their liquid contents; EUS revealed a unilocular lesion in four and a multilocular lesion in the other four. CT was used in only six cases before being abandoned for MRI instead.

MRI was performed in 23 cases without a standardized protocol because of the retrospective nature of the study and the rarity of the lesion. With acute suppuration, the MRI scan also revealed an associated vestigial lesion (one case). For the seven cases not scanned by MRI, the diagnosis was made possible by EUS and CT.

**Table 1** Histopathological study of retrorectal vestigial tumors in adults (30 cases).

<table>
<thead>
<tr>
<th></th>
<th>Number</th>
<th>Mean size and range (cm)</th>
<th>Unilocular</th>
<th>Multilocular</th>
<th>Acute infection</th>
<th>Malignant degeneration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epidermoid cyst</td>
<td>5</td>
<td>5.2 (3–7)</td>
<td>5</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Tailgut cyst</td>
<td>16</td>
<td>4.6 (1–10)</td>
<td>7</td>
<td>9</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Rectal duplication</td>
<td>4</td>
<td>6.5 (2–13)</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Teratoma</td>
<td>4</td>
<td>11.5 (6–15)</td>
<td>1</td>
<td>3</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Unclassifiable</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>30</td>
<td>16</td>
<td>14</td>
<td>3</td>
<td>1</td>
<td></td>
</tr>
</tbody>
</table>

calcifications (two patients). In one case, there was an associated anterior sacral meningocele. EUS was performed on 16 patients, allowing exploration of tumor volume and multicystic nature, and its relationship to the rectal wall and sphincter. In a preliminary study of the initial eight patients, the examination was performed with an Olympus™ GF-UM (7.5–12 MHz) echoendoscope, and proved the reliability of that technique for small lesions, permitting the diagnosis of vestigial cystic tumors in seven cases. In these seven, the tumors were 2 to 7 cm in diameter, and hypoechoic due to their liquid contents; EUS revealed a unilocular lesion in four and a multilocular lesion in the other four. CT was used in only six cases before being abandoned for MRI instead.

MRI was performed in 23 cases without a standardized protocol because of the retrospective nature of the study and the rarity of the lesion. With acute suppuration, the MRI scan also revealed an associated vestigial lesion (one case). For the seven cases not scanned by MRI, the diagnosis was made possible by EUS and CT.

**Treatment**

The objective of treatment was to remove the whole of the lesion after careful study of the EUS scans and, in particular, the MRI scans (especially useful for multilobular lesions) to avoid recurrence of the disease and malignant degeneration. The operations used a transanal approach in three cases (unilocular lesions with diameters < 2 cm), rectotomy in two cases and an intersphincteric procedure in one case due to the possibility of an abscess in the intersphincteric zone. The posterior approach (retroanorectal) was the most common technique for resection and was used in 23 cases, with resection of the coccyx in 15 of them. In two patients, surgical exploration uncovered a transsphincteric fistula running towards a postanal dimple: this was excised after seton drainage for two months in one, and the track sutured in the other. The abdominal approach was used in two patients with large lesions (6 cm and 15 cm) that were situated higher up the rectum. One of these lesions was cancerous, and required additional resection of the rectum and a low colorectal anastomosis. The combined posterior and abdominal approach was used in two patients: one was a multicystic teratoma following a previously incomplete anterior resection, and needed a posterior approach with resection of the coccyx; the other was a teratoma with a diameter of 15 cm.

**Histopathology**

According to the modified classification of Malafosse et al. [2], vestigial tumors can be divided into two groups:

- vestigial (or developmental) cysts, derived from embryological tissue usually present in this site, including:
  - epidermoid cysts (five patients), which are usually unilocular with a keratinized squamous epithelium, without skin appendages, and ‘pasty’ contents,
  - enteroid cysts, including tailgut cysts (16 cases) and rectal duplications (four cases). The tailgut cysts in half of our cases were multilocular, with ciliated or mucinous columnar epithelium or, sometimes, transitional or squamous epithelium, with disorganized smooth muscle bundles. The contents were varied — fluid, mucinous or pasty, or all three. The rectal duplications were usually unilocular and in close contact with the rectal wall, and composed of intestinal mucosa, submucosa and well-formed muscularis propria, comprising two layers separated by a neural plexus with ganglionic cells. In one patient, the mucosa was partially destroyed and, in another case, the lesion was infected;
- vestigial cysts, or teratomas (four patients), are derived from embryological tissue foreign to the site and are unusual because they are usually a childhood lesion. Composed of various derivatives of the three embryonal layers — ectodermic, mesodermic and endodermic tissues — in one case, the teratoma had undergone malignant transformation into an epidermoid carcinoma.

One lesion was unclassifiable because its wall was destroyed by infection. The histopathological results are summarized in Table 1.

**Mortality and morbidity**

There were no deaths in this series. Six patients (20%) had postoperative complications, including: three cases of posterior wound infection, but with good recovery; one case of anastomotic leakage after rectal resection that required three further operations; one case of anoperineal infection with rectoperineal leakage, which was successfully dealt with by a sliding advancement rectal flap; and one case of iatrogenic burns on the thighs caused by the antiseptic.
Table 2  Surgical approaches to treat recurrences of vestigial retrorectal tumors in adults (30 cases).

Table 3  Retrorectal tumors in adults, with incidences of malignancy, reported in the literature.

Local recurrence and survival

Twenty-six patients had a mean follow-up time of 3.2 years (range 0.5—15 years), two patients were followed for less than six months and two patients were lost to follow-up. The follow-up included a clinical examination and EUS or MRI one year after surgery. A recurrence or residual lesion was observed twice (Table 2). In one instance, the initial lesion was removed by the transanal approach because of its small size and unilobular aspect. Four months later, the patient was operated on again, using the posterior approach with resection of the coccyx to remove the same lesion (tailgut cyst). There was no recurrence after 14 years of follow-up. In the other patient, the initial lesion, which appeared to be unilobular, was removed by the posterior approach without coccygeal resection, but was probably incomplete as a second operation was necessary a year later, using the same procedure, but with resection of the coccyx. There was no recurrence after five years of follow-up. The patient, who had a sacrococcygeal teratoma with degeneration into epidemoid carcinoma, received adjuvant radiochemotherapy and had no recurrence after eight years of follow-up.

Discussion

Vestigial retrorectal cystic tumors in adults are unusual, so only a few large series are described in the literature [1, 3—9] (Table 3). Indeed, a recent review of retrorectal tumors by Hobson et al. [10] reported that, except in specialized centers, a general surgeon is likely to diagnose only one retrorectal tumor in the whole of his professional life. Other series of retrorectal tumors have been published, but are limited [11—16]. Vestigial tumors are generally found in women and are often revealed by chance during a routine physical examination. Its symptoms are limited to pain, bleeding or a rectal syndrome. In one case-report [17], a huge gluteal mass made the diagnosis possible.

When the first symptom of the lesion is suppuration and a postanal dimple is found (characteristic of a tailgut cyst; see Fig. 2D), this may be mistakenly diagnosed as the secondary opening of an anorectal fistula or, if MRI is performed, as complex or recurrent suppuration [18—19]. This was the case of the patient in our present series whose infection was treated by puncture and antibiotics before resection was performed.

Rectal examination is essential, and its findings should be supplemented by EUS — especially in the case of small lesions [20, 21] — CT and MRI [22—26]. EUS may be useful for defining the limits of small lesions, their relation to the anal sphincter and the nature of their contents [20], and Sogni et al. [21] have shown that it may be better than CT at revealing any connection to the rectal wall. On the other hand, it cannot visualize any attachments to local muscle

<table>
<thead>
<tr>
<th>Source</th>
<th>Study period</th>
<th>No. of retrorectal tumors</th>
<th>No. of congenital retrorectal tumors</th>
<th>No. of vestigial cystic tumors</th>
<th>No. of malignant vestigial cystic tumors</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lee et al. [4] 1988</td>
<td>1965—1980</td>
<td>70</td>
<td>34</td>
<td>33</td>
<td>4</td>
</tr>
<tr>
<td>Killingsworth et al. [7a] 2005</td>
<td>1988—2004</td>
<td>NA</td>
<td>NA</td>
<td>43</td>
<td>17b</td>
</tr>
<tr>
<td>Present study 2007</td>
<td>1989—2005</td>
<td>NA</td>
<td>NA</td>
<td>30</td>
<td>1</td>
</tr>
</tbody>
</table>

NA, data not available.

a Review of the literature.
b With neuroendocrine tumors.
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It is not ideal for very large lesions and it is highly operator-dependent.

In the present series, CT was used early on in the study, but was soon replaced by MRI because of its poor resolution of spaces and with contrast. CT spatial resolution is not optimal because of the surrounding bony environment and because only an axial plane is possible, whereas MRI allows a number of different planes to be scanned. As for resolution with contrast, because of variations in the signal depending on the chosen sequence, MRI allows a likely identification of the nature of lesional contents, which is not as clear with CT.

MRI should be performed with T2-weighted sequences in three spatial planes, including a fast T2-weighted sequence and two T1-weighted sequences, with and without fat saturation. The latter three sequences permit the study of the contents of the lesion, and allow educated guesses to be made as to its histopathology and the possibility of cancer. The fast T2-weighted sequence allows good contrast imaging of the lesion compared with the other soft tissues (Fig. 1). The T2 sequence can equally show the course of a tract toward the sphincter, or reveal several cysts in the intersphincteric zone or lying close to the skin (Fig. 2D).

The lesions often have an intermediate or even high intensity signal on T1-weighted sequences (Fig. 3 and Fig. 4A) and are usually homogeneous, but can vary in intensity from one cyst to another. The high intensity signal with T1 may be due to the fatty contents of teratomas, and should disappear on T1-weighted fat-saturated images. Another clue to the diagnosis of teratoma is the presence of heterogeneity within the lesion due to clusters of exoskeleton held in suspension (Fig. 4B). However, the high intensity signals on T1-weighted sequences are most often due to the contents being rich in protein, as is the case with most tailgut cysts, where the contents are macroscopically pasty (Fig. 1A).

The signal on T2-weighted images is usually high intensity and homogeneous, similar to that of the adjacent cerebrospinal fluid even though the contents are macroscopically pasty (Fig. 4C).

Locating the rectum during MRI is possible by applying ultrasound or paramagnetic gel inside it. If cancer is suspected, then doing an MRI scan after injection of gadolinium should permit differentiation of an adenocarcinoma from a carcinoid tumor [27,28].

A preoperative MRI is essential for deciding on the best surgical procedure, for localization of the upper extremity of the lesion in relation to the third sacral vertebra, for showing the number and location of the cysts and the volume of the largest one, and for extending the examination to the intersphincteric space (which may contain an associated lesion).

There is little information in the literature concerning the significance of MRI in such lesions and only small series have been reported. In a recent retrospective work, Yang et al. [26] presented the MRI study of five tailgut cysts, which showed that the signals from these lesions are always low intensity on T1- and high intensity on T2-weighted sequences. However, they also observed that the presence of mucinous or protein contents, or bleeding, can produce hyperintense signals on T1-weighted sequences. In addition,
Figure 2  Multilocular tailgut cyst, $5 \times 4 \times 2.5$ cm, lies the right paramedian region in front of the sacrococcygeal junction, and has thick greenish contents. A: MRI, axial T1-weighted echo-gradient (EG) image with fat saturation (FSPGR/80, TR: 200, TE: 1.9): the lesion is hyperintense, confirming the lack of fatty contents. MRI, median sagittal fast T2-weighted image (FRFSE, TR: 3940, TE: 132): the lesion appears to be unilocular and continues downward towards the intersphincteric space, ending in a cluster of small cysts (arrows). B: MRI, median sagittal T1-weighted EG image with fat suppression post-gadolinium (FSPGR/80, TR: 275, TE: 2): the components of the cluster are hypointense, confirming their cystic nature and liquid contents. Appearance of the anal edge: postanal dimple (arrow).


they found that a T2-weighted sequence was better than a T1-weighted sequence or a CT scan for revealing small cystic lesions lying close to the main one. Glasgow et al. [8] compared the diagnostic sensitivity of CT and MRI, and found the latter to be superior; however, this finding was not statistically significant because of the small scale of the series. Hobson et al. [10] and Chêne et al. [29] both found that MRI was able to identify the risk of malignant degeneration, particularly by the solid appearance of the lesional contents and/or extension to bone or nerves. Recently, Woodfield et al. [9] studied 27 cases of retrorectal tumors (comprising 12 vestigial tumors) and found that MRI was of great benefit in the diagnosis of malignancy and in the choice of surgical approach.

Preoperative explorations make it possible to rule out other types of retrorectal tumors such as chordomas, rare neural or bony tumors derived from the anal glands [30], and inflammatory pseudotumors, which are also very uncommon.
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Figure 3 Unilobular epidermoid cyst, 6 \times 5 \times 3.5 cm, right paramedian location, is precoccygeal and has pasty contents. A: MRI, axial T1-weighted image (FSE, TR: 600, TE: 10.4): homogeneous intermediate signal, the same intensity as the adjacent muscles (gluteus major). B: MRI, right paramedian sagittal T2-weighted image (FSE, TR: 3300, TE: 117): heterogeneous signal from the lesion due to hypointense pseudonodular signals within it.

Kyste épidermoïde, 6 \times 5 \times 3.5 cm, uniloculaire, paramédian droit précoccygien, à contenu mastic. A: IRM coupe axiale Spin Écho T1 (FSE, TR: 600, TE: 10.4): signal intermédiaire, homogène, iso-intense quand comparé avec les muscles adjacents (grand fessier). B: IRM coupe sagittale paramédiane droite Spin Écho T2 (FSE, TR: 3300, TE: 117): signal hétérogène marqué par des images pseudonodulaires en hyposignal au centre de la lésion.

[23]. The association of a vestigial retrorectal cystic tumor with a meningocele is a hazard for the surgeon, and has to be carefully investigated to avoid, in the case of infection, the risk of death associated with resection in such a case. In a case-report by Krivokapic et al. [31], the surgical resection of an enteroid cyst that lay in close proximity to a meningocele led to meningitis that was difficult to treat with antibiotics. In one patient in the present series, there was a medical history of meningocele, which was surgically cured two years before our surgical removal, by the anterior approach, of a huge teratoma that had probably not been diagnosed at the time of the earlier operation.

The surgical removal of such lesions are necessary, even when no symptoms are present, because of the possible development of an infection, which is especially likely with rectal duplications and tailgut cysts (three cases, or 10%, of the present series). Surgical removal of the lesion is also necessary because of its potential for malignant degeneration, as described in the literature (see Table 3) (occurring in 7—10% of enteroid cysts [27,32—34] and 15—25% of teratomas [1,4,35]); indeed, one patient in the present series had degeneration into an epidermoid carcinoma.

Prasad et al. [36] studied 10 cases of malignant degeneration of retrorectal cysts into six adenocarcinomas and four carcinoid tumors. Such degeneration has also been reported by others [28,37,38]. Mourra et al. [39] described the degeneration of a tailgut cyst that was less than 2 cm in diameter into a neuroendocrine carcinoma. Yamaguchi et al. [40] described a particular form of rectal adenocarcinoma invading a tailgut cyst through a fistula, but without degeneration of the cyst, and Killingsworth et al. [7] described 17 cases of degeneration of 43 tailgut cysts, including 11 adenocarcinomas, five carcinoid tumors and one endocrine tumor.

If degeneration is suspected from the imaging information, a metastatic work-up needs to be done, and the resection should be large, such as a posterior pelvectomy. Also, carcinoembryonic antigen and alpha-fetoprotein tests may be positive if the lesion is a teratoma, although this result is not specific for malignancy [1]; nevertheless, it can still be useful for the follow-up of surgical resection [41].

When the lesion is discovered during pregnancy, it may be necessary to perform a caesarean section as a precaution to avoid dystocia due to distortion of the birth canal [42].

The choice of surgical procedure depends on the lesion’s size, its location in relation to the third sacral vertebra and whether it is uni- or multilobulated [9,43,44]. In fact, this is where MRI information regarding the number and location of the cysts, and any possible intrasphincteric extension is especially useful. The transanal approach would have to be considered in the case of a low-lying unilobular lesion under the tip of the coccyx, with a diameter of 2—3 cm, on preoperative MRI. Hjermstad et al. [3] treated nine patients in this way out of a series of 48, and Pidala et al. [45] treated 11 patients out of 14, all with no recurrence during a follow-up of 39 months.

The posterior anoperineal approach, brought back into use by Abel et al. [35], allows a good view of the posterior rectal wall after section of the median raphe and opening of the ischiorectal fossa. Partial or complete section of the coccyx or of the last sacral vertebra is useful, almost routine, if the lesion is located in front of the bony wall and multilob-
Figure 4  Teratoma, 11 cm in diameter, is multilocular and precoccygeal, and is pushing the rectum forward; it also has an unusual subcutaneous extension in the subcoccygeal area. The lesion was pasty, brownish and contained a tuft of hair. A: MRI, axial T1-weighted image (TSE, TR: 700, TE: 12): the lesion appears to be bilocular; its anterior part has an intermediate homogeneous signal, while the posterior part appears to comprise two components, defined by a clear limit, revealing the fatty nature of the anterior part. B: MRI, paramedian sagittal T1-weighted image (TSE, TR: 700, TE: 12): within the posterior component is a heterogeneous signal corresponding to the tuft of hair in suspension (arrow). MRI, median sagittal T2-weighted image (TSE, TR: 4500, TE: 128): the posterior part of the lesion is hyperintense, revealing its liquid contents. The subcoccygeal and subcutaneous parts of the lesion are multilocular.

ular, as there is then a high risk of overlooking a small cyst deep in front of the sacrum, which may lead to recurrence of the disease, as it did in two patients in the present series. In the case of a fistula running towards the sphincter, as suggested by the presence of a retroanal dimple, dissection from top to bottom is easier, and complete removal of the fistula track is not necessary. Such a procedure is generally used for smaller lesions (diameter < 4–5 cm) but, according to Lee et al. [4] and Barthod et al. [11], it may also be used for lesions up to 10 cm of diameter (or more), as in two patients in the present series and one from Chêne et al. [29] (whose lesion was 17 cm in diameter), if there is no sign of malignant degeneration. The anteropelvic approach is associated with very low morbidity.

The abdominal posterior and perineal approach is used for lesions as large as 5 cm in diameter that do not involve the sacrum, but where the upper extremity reaches the third sacral vertebra while the lower part fails to reach or extend beyond the fourth sacral vertebra. In such cases, rectal dissection needs to be performed in the presacral plane as this results in less bleeding. This procedure is recommended by Guillem et al. [46], who used it in a small series on two occasions, once for a teratoma that was 15 cm in diameter and again for a malignant teratoma that was 5 cm in diameter.

The combined posterior and abdominal approach is used for larger lesions (diameter > 5 cm) that are adherent to the posterior bony wall, and where the upper extremity reaches the third sacral vertebra and malignant degeneration is suspected. The operating position is either lateral reclining, or on the back first, followed by the prone position for the lower stage of the operation. This operation can cause considerable impairment, as it requires exposure of the sacral nerve roots, ligature of the dural sac and sacral osteotomy. Such a combined approach was performed twice in the present series, once for a 15-cm teratoma and again for a rectal duplication that was 13 cm in diameter.

Histopathological analyses of our 30 cases showed that, unlike previous reports in the literature, an isolated dermoid cyst was not found. That type of vestigial cyst is composed of malpighian epithelioma and cutaneous sudoriparous or pilary annexes, sometimes surrounded by smooth muscle fibers. The contents are pasty and sometimes calcified. In both the series reported by Jao et al. [1], involving 49 vestigial tumors, and the present series, no dermoid cysts were found. That is in agreement with the observations of Barthod et al. [11] and Chêne et al. [29], who suggested that unilocular dermoid cysts may be simplified dysembryomas that are graded as teratomas, which often have a dermoid constituent.

The follow-up of patients is useful because of the risk of recurrence or malignant degeneration [1,47] due to incomplete resection, particularly with multilobular forms of the lesion. Recurrence was seen in two patients of the present series (7%), and led to repeat surgery using the posterior approach, with coccygeal resection to maximize the exposure of the operation. Hjermstad et al. [3] found four recurrences (11%) in a series of 36 tailgut cysts with a mean follow-up time of 11 years, and Lev-Chelouche et al. [6] found no recurrences in a series of 12 congenital benign lesions with a median follow-up time of 54 months. These were medical and clinical follow-ups and, most important of all, included EUS or MRI explorations one year after surgery. In cases of malignant degeneration, an adjuvant therapy (chemo- or radiotherapy) can be instituted, accompanied by frequent supervision.

Conclusion

This series of 30 vestigial retrorectal cystic tumors in adults, involving a review of the imaging and histopathology findings, clarifies the surgical procedure and the management of any subsequent risks associated with such lesions. MRI appears to be essential for the exploration of these lesions.

References
