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Retrorectal dermoid cyst

Diagnostic approach and surgical strategies: an updated summary

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Abstract:

Aim

Dermoid cysts of the presacral region are part of the large family of the retrorectal tumours, a rare clinical entity. The roles of different diagnostic approaches,non operative management and the indications for surgical resection are controversial.

Different studieshave recently reviewed the literature on retrorectal tumours and produced an algorithm for contemporary management. The role of MRI and biopsy were been re-evaluated and updated. Our aim is to summarize the different proposals and, if possible, to combine them in onecommon valid approach to these rare tumours pointing out dermoid cyst management.

Method

A systematic literature review was conducted using the PubMed database. Case reports and retrospective reviews made up the majority of articles. Keywords used for the research were: Retrorectaltumours, dermoid, presacral cyst.

Results

Congenital lesions represent 50 to 70% of all the Retrorectal tumours.

Out of them chordoma is the most common solid tumour and developmental cysts (epidermoid, dermoid, teratoma, tailgut cyst) are the most common among the cystic new growth.

MRI can discriminate between solid and cystic masses and from benign and malignant ones with a sensitivity of 95%.

Needle biopsy has a higher sensitivity in diagnosing solid malignancies (sensitivity 96% and specificity 100%) with a minimal risk of seeding. Needle biopsyin cystic masses is not recommended since they may contain malignancy on pathologic final evaluation with previous negative needle aspiration cytology.

Meanwhile the literature widely supports the surgical approach in solid tumours,for cystic masses observation is often the suggested choice.

Conclusions

Retrorectal dermoid cyst,among the retrorectal benign masses, is a rare entity. MRI can differentiate between solid and cystic masses but a needle biopsy cannot rule out potential malignancy in cystic ones (present in 3 to 15% of teratomas and in 2-13% of tailgut cysts).

These tumours can often get infected (30%) and, in fertile females, can lead to obstructed labour.

New mini invasive surgical techniques (laparoscopy, TEMS) havestrongly developed during these last ten years.

All these considerations lead us to suggest, also for this kind of cystic tumours, an early surgical approach in referral centres where minimally invasive surgery can safely be performed soon after diagnosis is done. More multicentric studies are needed to support this suggestion.

What does this paper add to the literature? This paper is a comprehensive updated review of diagnostic and therapeutic approach to retrorectal dermoid cyst.

Introduction:

The retrorectal space –also defined as presacral space – (**Figure 1**) is the anatomical site between the sacrum and the rectum. It contains a wide variety of tissues giving rise to diverse range of tumour types that can develop as solid or cystic neoplasms.

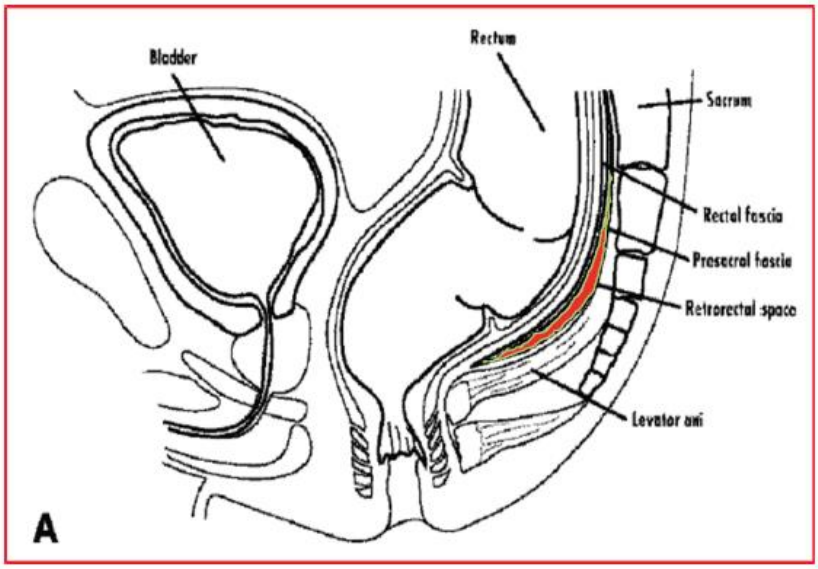

Figure 1 - Retrorectal space

Retrorectal tumours are a rare clinical entity: some studies give an incidence of 1.4 to 6.3 new cases per year, accounting for 1/40000-60000 new admission in big referral hospitals [1]. Their incidence is greater in women during the reproductive period [2] (even if for some Authors itis a bias due to the more frequent US investigations done in women during pregnancy). They are classified as congenital, inflammatory, neurogenic, osseous and miscellaneous [3, 4].

Congenital tumours represent 55 to 70% of all the presacral masses[4].

Development cyst is the most frequently encountered retrorectal tumour, being the 60% of all the congenital lesions. They may arise from all the three embryonic germ layers and so they can be divided in epidermoid, dermoid, duplication (enterogenous), and tail gut cysts. Epidermoid cysts are composed of stratified squamous cells; they are typically benign unilocularlesions that do not contain skin appendages. Unlike epidermoid cysts, dermoid cysts have stratified squamous epithelium with skin appendages (sweat glands, hair follicles, sebaceous cyst) **(Figure 2).**

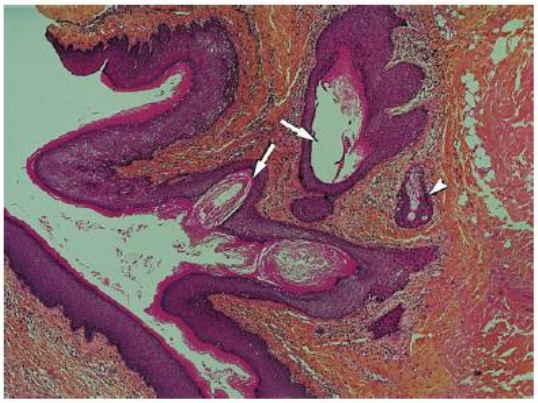

Figure 2. Dermoid cyst – skin appendages

Methods:

A systematic literature review was conducted using the PubMed database. Case reports and retrospective reviews made up the majority of articles.

Keywords used for the research were: Retrorectal tumours, dermoid, presacral cyst.

Duplicated data and abstracts were excluded from this study.

Results:

Clinical presentation Retrorectal cysts are mainly asymptomatic (26-50%). They are often discovered through a rectal examination or a Ultrasonography done for other reasons.Sometimes patients refer specific symptoms related to the mass effect caused by the volume of the lesion (constipation, rectal fullness, dysuria, pelvic pain) or due to some of the most common complications (infections that can occur up to 30%; or chronic non-healing recurrent anal fistula [5]; obstructed labour).

The malignant potential is more defined for teratomas (5-12%) and tailgut cysts, whileit seems to be negligible in other cystic tumours like dermoids.

Diagnosis

As reported in Literature MRI is the gold standard to reach a preoperative diagnosis [6, 7]. Hopper compared MRI and CT and showing “an overall diagnostic accuracy of imaging in distinguishing benign and malignant disease of 82%“. In this study MRI resulted more accurate in the diagnosis of malignant lesion than CT (94% vs 64%) with a positive and negative predictive values of 78% and 90% [8].

According to published studies [6, 8, 9] MRI axial images are the most useful tool in determining involvement of the pelvic side wall. T1, T2 and fat satured T1 images help to determine the internal composition of the tumour, whether cystic, solid, fatty or mixed [9].

The role of TRUS (trans rectal ultrasound) in diagnosing these tumours is controversial and someAuthors limit its use in preforming trans rectal guided biopsies [10] but it is mandatory to underline the basic positive characteristics of this exam: it does not give radiation to the patient, it is rapid, it can be repeated safely as many times as we need, and it is inexpensive so it could be also proposed during the follow-up of these patients.

Needle biopsy has a clear role in diagnosing solid masses giving the possibility of a more accurate therapeutic plan (i.e. in GIST and in B-cell lymphoma where preoperative chemotherapy can play an important role). It is absolutely proscribed in Tarlov perineural cysts, whose management is entirely different. A recent review of the Literature performed by Toh and Morgan, from Sidney, highlightsthat its accuracy as a pre-operative exam in cystic lesions is almost unreliable [11].

Otherwise needle biopsy has a higher sensitivity in diagnosing solid malignancies (sensitivity 96% and specificity 100%) with a minimal risk of seeding [12]. Needle biopsy in cystic masses is not recommended since they may contain malignancy on pathologic final evaluation with previous negative needle aspiration cytology [13].

Treatment

Once a retrorectal tumour is diagnosed, most patients require surgery [10, 14,15]. For benign tumours, this is for the risk of misdiagnosis, malignant degeneration and risk of disease and symptoms progression [11].

The surgical approach to presacral tumours classically depends on their position, their volume, aggressiveness and malignant potential.

The sacrococcygeal sinus angle (SSA) is the angle formed by a tangential line from anterior surface of S1 and a line drawn from the sacral promontory to coccyx. As suggested by Kaplan et Al (16) SSA and S3 are the most important landmarks for the posterior approach of presacral lesions. To obtain this landmarks a sagittal T2-weighted MRI image is commonly used, in order to determine the upper level of the sacrum reached by the tumour.

When an adequate localization of the neoplasm is obtained with MRI, the classical posterior perineal (Kraske’s) approach is used for masses below S3, while an anterior abdominal approach is recommended for tumours above the middle of S3, or if it is present a pelvic or pelvic sidewall involvement; for bigger masses a combined approach is often the only chance. Recent studies show that

laparoscopic removal is safe for resection of retrorectal lesions, with no increase in morbidity or intraoperative complications [15,17], the conversion rate is low and the main predictors of open conversion include huge retrorectal tumours, obesity, narrow pelvis and high ASA.

TEM (Transanal Endoscopic Microsurgery) seems safe for the resection of retrorectal benign masses [11, 18, 20].

Approaching a retrorectal tumour through the rectal wall does not follow oncological principles for malignancies.

The concern is that it may be difficult to exclude malignant degeneration of a benign appearing cyst (12.9% of cystic lesions were malignant in a recent study[19]); it has been argued that even with careful pre-operative assessment, malignancy cannot be completely excluded [21]. On the other side a recent retrospective study from New Zealand [8], in a series of 69 patients, supports the non-operative treatment if an MRI is defining a benign appearance of the new growth. (**Figure 3**)

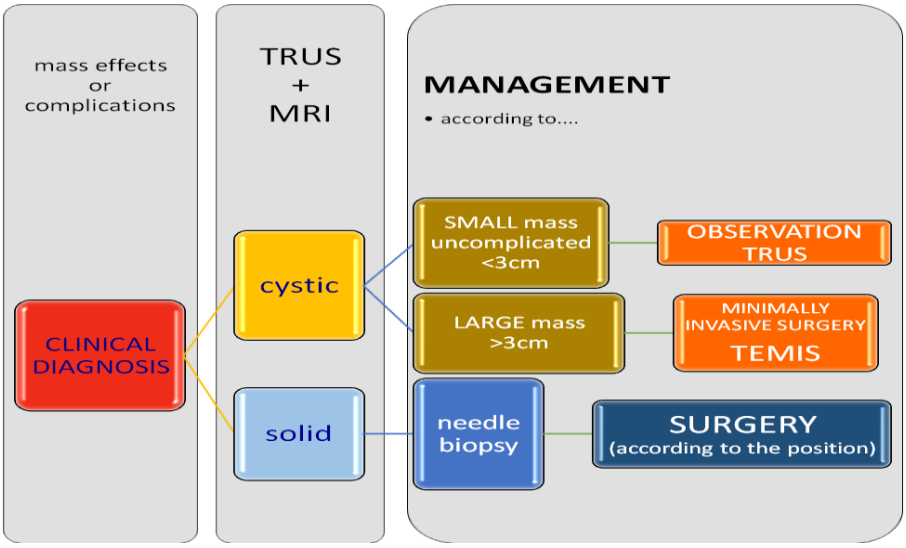

Figure 3 – Possible Flow-chart to approach retrorectal tumours: some of the above mentioned assumptions need larger consensus from multicentric studies (i.e dimension do define "small or large"; or the use of TRUS)

Discussion

The literature produces some incontestable assumptions on retrorectal tumours:

– They are a rare clinical entity
– Congenital lesions represent up to 60% of these masses
– Among cystic lesions dermoids are often multiloculated masses with skin appendages
– MRI is the best preoperative investigation that can easily distinguish between solid and cystic lesions, its sensitiveness in diagnosing malignancies reaches 94%
– Needle biopsy is useful in solid masses to plan the treatment, in cystic lesions it can miss a malignancy

– Surgical treatment is well stated for solid malignant tumours with different approaches according to the position (below or above the middle S3 line), their volume and the invasion of other pelvic structures.
– Cystic lesions can became infected up to 30% of cases.

A lot of limitations are present in different literature reviews: most of the studies were retrospective, or case series or case reports, so this situation, due mainly to the rarity of these tumours, brings us to other topics still under discussion and it shows the needs for further evaluation:

– The role of TRUS: is it only a support to needle biopsy or may it help in reaching a final diagnosis being useful also during the follow up after removal of benign or locally malignant masses?
– Development cyst management can be only strict observation or would the surgical removal be better as soon as diagnosis is done? Or –again- the approach has to be guided by the size of the mass?
– In cystic masses is an early minimally invasive surgery a valid option in order to avoid complication, future growing or malignant changes?

In our opinion to still all doubts some more good multicentric studies have to be planned to evaluate the impact of minimally invasive surgery in solving the problem when a cystic, apparently benign lesion, is diagnosed in retrorectal space.

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