

Extragonadal Perirectal Mature Cystic Teratoma in the Adult Male

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ABSTRACT

Background: Extragonadal abdominopelvic teratomas in adults are extremely rare, and those in males are exceedingly rare. These masses are most commonly found incidentally and require surgical excision for diagnostic confirmation after a thorough workup.

Case Presentation: This is a case report of a 49-year-old male who presented to a urology office with symptoms of hematuria, incidentally, found to have a pelvic mass on computed tomography urogram prompting colorectal surgical evaluation and subsequent laparoscopic complete excision. The clinical presentation, radiographic findings, and histopathological findings are described along with a literature review of extragonadal abdominopelvic mature cystic teratoma, also referred to as a sacrococcygeal teratoma.

Discussion: A broad differential diagnosis was generated for this patient with a pelvic mass after complete work-up, consisting of a dermoid or epidermoid cyst, liposarcoma, or sacrococcygeal teratoma. Radiological features can aid in

the diagnostic confusion that may present in the adult patient.

Conclusion: Albeit rare in the male and adult population, sacrococcygeal teratoma is a plausible differential diagnosis for a pelvic mass. Underrepresented in the literature in regard to guidelines on management, complete surgical excision is the gold standard, with laparoscopy being a reasonable approach.

Key Words: Sacrococcygeal teratoma, Extragonadal abdominopelvic mature cystic teratoma, Mature cystic teratoma.

BACKGROUND

A teratoma is a mass that originates from the germ cells of the three primitive embryonic layers (ectoderm, mesoderm, and endoderm) and commonly arises from the ovaries or testicles, thereby characterized as gonadal in origin. In the literature and clinical practice, these are well studied and described. These masses comprise different types of tissues such as fat, hair, muscle, bone, etc.¹ Extragonadal teratomas are uncommon and as the name declares, arises from anatomical locations other than the gonads. Common locations for extragonadal teratomas are within midline structures such as the intracranial pineal gland, anterior mediastinum, retroperitoneum, and sacrococcygeal regions.^{2,3} The most common is the presacral region as it is an area of embryonic fusion of the hindgut, proctodeum (lower anal canal), neural tissue, and bone thereby containing tissue derived from all germ layers. These are either congenital or acquired, with congenital tumors accounting for the majority.^{4,5} There are no known genetic causes for sacrococcygeal teratomas.⁶ These tumors in this specific location are far more common in neonates, infants, and children rather than adults, and females rather than males.⁷⁻⁹ In the adult population this is a rare case with only case reports found in the literature describing their unique presentation. An estimated adult incidence of 1 in 40,000 – 63,000 and 1.4 – 6.3 patients per year has been described in major referral centers.^{4,10} These masses are likely to be congenital

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Acknowledgements: none.

Conflict of interests: none.

Disclosure: none.

Funding sources: none.

Informed consent: Dr. Pranamy Mahankali declares that written informed consent was obtained from the patient/s for publication of this study/report and any accompanying images.

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DOI:10.4293/CRSLS.2022.00035

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when found in the adult population. Their malignant potential varies greatly, depending on the patient's age at presentation, with malignant teratomas being more aggressive in children and much less common beyond the second decade of life.¹¹ Complete surgical excision is recommended to establish a diagnosis given the broad differential that is present in the adult population along with ruling out any malignant transformation. Common malignancies, albeit rare, identified are squamous carcinoma, neuroendocrine tumor, and adenocarcinoma. The prognosis for complete surgical excision for benign teratomas is excellent.¹²

CASE PRESENTATION

Our patient is a 49-year-old male who initially presented to the urology office for asymptomatic hematuria for which a computed tomography (CT) urogram was obtained noting the presence of a large extraperitoneal perirectal mass measuring $8.8 \times 8.1 \times 1.9$ cm (cc x ap x tr) within the left perirectal space abutting the left aspect of the rectum displacing it to the right. The mass is predominantly homogenous, with low attenuation, Hounsfield units around -40 without peripheral enhancement, calcifications or surrounding enlarged lymph nodes present [Figures 1A and 1B]. This was noted to be distinct and separate from the rectum, prostate, bladder, and ureters. The genitourinary tract was unaffected, prompting referral to the colorectal surgery department for further evaluation.

The patient was seen in our office and was found to be asymptomatic from this incidentally discovered mass. Apart from hematuria, he denied any pelvic pain, sexual

dysfunction, bowel, or bladder symptoms. The patient's past medical and surgical history were only pertinent for two prior colonoscopies in 2018 and 2013, which were reportedly negative. Physical examination was unremarkable without any abdominal tenderness or fullness appreciated and a digital rectal examination with no external pathology, visible, or palpable mass. A magnetic resonance (MR) imaging of the pelvis was obtained to further characterize this mass, which redemonstrated this large well-defined extraperitoneal perirectal mass located in the left perirectal space. [Figures 2a and 2b] Innumerable small globules of T1 hypointensity were noted within the mass consistent with fatty tissue along with interspersed fluid. Again, no areas of signal void were noted suggesting calcifications. The mass was inferior to the anterior peritoneal reflection, confirming its extraperitoneal origin, abutting and displacing the rectum anteriorly and laterally. No rectal invasion was visualized, although only a thin layer of tissue measuring approximately three mm was separating the mass from the rectum. The prostate and seminal vesicles were also displaced by this mass to the right without any evidence of infiltration. There was no evidence of adenopathy or metastatic disease within the abdomen, pelvis, or chest. Our differential included dermoid or epidermoid cysts given predominance of hypoattenuating fat as well as liposarcoma or sacrococcygeal teratoma given variable appearance with mixed solid and cystic components albeit without visible invasion.¹ Routine biochemical testing was all within normal limits.

Surgical options were discussed with the patient, and after explanation and understanding of the risks and benefits the patient was consented for a laparoscopic excision of this perirectal mass. The day before surgery the patient completed a course of Nichols prep and Golytely. The

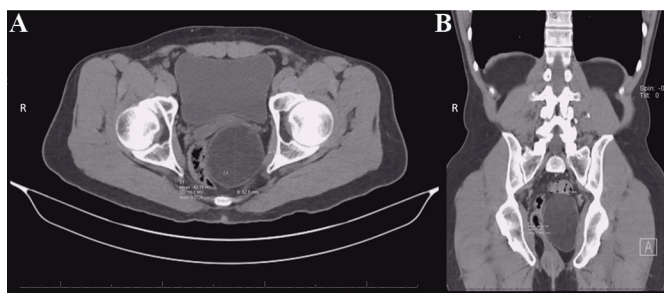


Figure 1. (A) Axial view of the extra-peritoneal teratoma located within the left perirectal space abutting the rectum, without invasion, with an approximately 3 mm layer of separation. (B) Coronal view demonstrating the $8.8 \times 6.1 \times 5.9$ cm mass effect with resultant displacement of the rectum by the teratoma anteriorly and laterally.

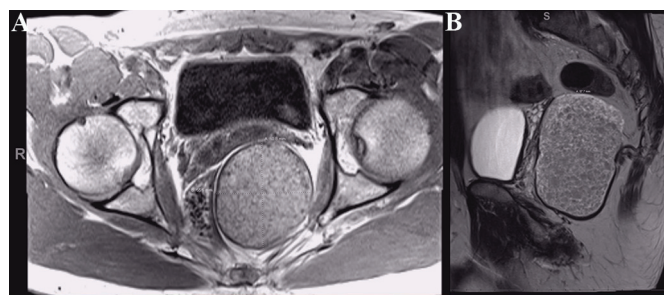


Figure 2. T2 weighted magnetic resonance imaging of the pelvis demonstrating the mature cystic teratoma. (A) Axial view revealing an innumerable number of hypointense small globules suggesting fatty tissue with interspersed fluid. (B) Sagittal view shows a large mass inferior to the anterior peritoneal reflection confirming extraperitoneal origin of the mass and its location within the left perirectal space.

abdomen was prepped with betadine before the procedure went underway. The anterior peritoneal reflection was dissected laparoscopically to encounter a large tan cystic mass, which was circumferentially dissected from the surrounding tissues and completely removed without rupture. There was no pelvic or abdominal adenopathy, ascites, or additional intra-abdominal gross abnormalities. Postoperative gross examination of the perirectal mass is seen in **Figures 3A – 3C**. Grossly, the specimen appeared tan with an outer rough and irregular surface. Intracystic components were copious, thick, and yellow cheese-like with multiple hair shafts. Microscopic tissue examination [**Figures 4A – 4C**] revealed a cystic lesion lined by keratinized stratified squamous epithelium with skin appendages including hair shafts with no evidence of malignancy or immature components, consistent with abdominopelvic extragonadal mature cystic teratoma.

DISCUSSION

The case that we describe in this report is an extremely rare tumor found in the adult population, particularly in the male sex. An extensive literature review was undertaken which revealed only unique case reports or small single-institutional case series.^{13–21} Sacrococcygeal teratomas (SCTs) have an incidence of 1 in 35,000 – 40,000 live births, therefore making them a common tumor found in the neonatal, infant, and child population. In the adult population, this is a rare entity.^{9,19} In accord with cases reported in the literature, there is also a female predominance of 3 – 4:1³. This case we present is unique in that our patient is male. Although the majority are benign, there still persists a 20% – 30% risk of infection and a 1% –

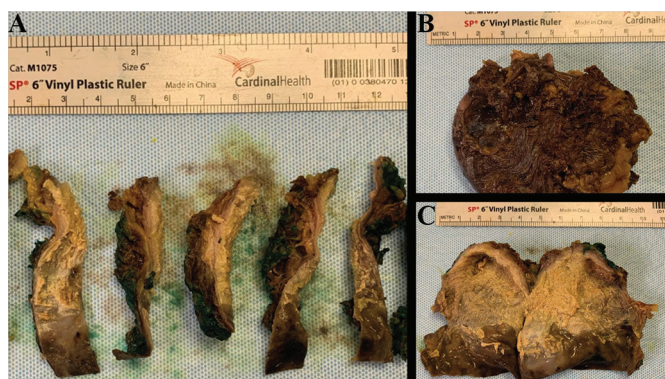


Figure 3. Gross pathological examination of extragonadal mature cystic teratoma. (A) Cross sectional cuts. (B) Rough tan colored outer surface. (C) Cyst predominantly filled with sebaceous material and hair follicles.

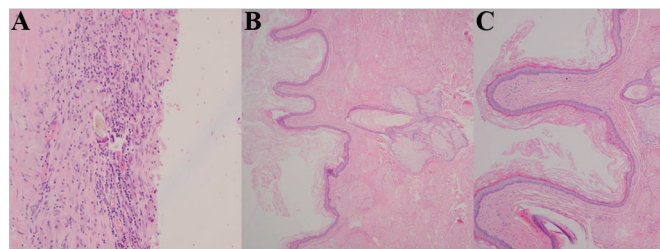


Figure 4. Microscopic examination of extragonadal mature cystic teratoma. (A) Hematoxylin and Eosin, Hair shaft. (B) Hematoxylin and Eosin, cyst lined by squamous epithelium. (C) Hematoxylin and Eosin (4 ×), keratinized stratified squamous epithelium.

12% risk of malignant transformation.²¹ The standard of care is surgical excision and the role of surgery is two-fold diagnostic as well as curative.

The Altman classification of the Surgical Section of the American Academy of Pediatrics divides SCTs into four categories based on their anatomical location. Type I (46%) is completely external, Type II (35%) is predominantly external but with significant intrapelvic extension. Type III (9%) is apparent externally but with a predominantly pelvic mass that extends into the abdomen. Type IV (10%) is a presacral mass with no external components. The most common in adults is Type III, although in our patient this is a true Type IV with no tissue connection to the coccyx.²² Histopathologically, these teratomas are divided into three categories: mature, immature, or malignant. The incidence of malignancy increases with age.^{3,16,23}

Adult patients will typically present with nonspecific or subtle symptoms of a mass effect, otherwise the majority will be diagnosed incidentally, by chance during imaging studies or by clinical examination. Our patient presented with symptoms related to a mass effect on the genitourinary tract resulting in hematuria which prompted further work-up revealing this teratoma. Other symptoms reported include rectal pain, back or pelvic pain, paresthesias, saddle anesthesia, urinary urgency or retention, frequent urge to defecate, incomplete evacuation or constipation, and loss of bladder or rectal control.²⁴ In women there are additional symptoms that may lead to an earlier time to diagnosis, although this is speculation, as most cases found in the literature present a female patient.

Radiological imaging aids in the diagnosis of these masses along with assisting in surgical planning by defining their relationships with surrounding tissues. Initially, a CT was obtained for our patient which identified a pelvic mass. It

was further characterized using MR imaging to help us differentiate between our working differential diagnoses: dermoid cyst, epidermoid cyst, liposarcoma, and teratoma. Dermoid and epidermoid cysts vary based on their composition on histopathology but also based on their characteristic appearance on MR imaging. A dermoid cyst contains at least two of the three germ cell layers, appearing as a complex, well-circumscribed cystic mass containing hypoattenuating fatty nodules and fluid density along with calcifications. In comparison, an epidermoid cyst lacks additional skin appendages such as hair that may be present within dermoid cysts. Epidermoid cysts commonly appear as unilocular masses containing homogenous and hypoattenuating fluid material with a distinct thin wall.¹ Liposarcoma is a mesenchymal tumor with a wide spectrum of radiographic appearance based on type: well-differentiated, dedifferentiated, myxoid, pleomorphic, and mixed-type.²⁵ As this diagnosis is more common within the age group of our patient, this was also considered as part of our differential.

Definitive diagnosis is only possible after histopathological examination, given the broad differential diagnoses present. As previously stated, surgery is the standard of care. CT and MR imaging can be used to provide details of the tumor's characteristics and assist in predicting whether the tumor is benign or malignant. Alarming features include poorly defined soft tissue planes, local invasion, predominantly solid components, and regional lymph node enlargement. When these features are present, a pre-operative biopsy can be considered to aid in determining if neo-adjuvant or adjuvant therapy is necessary.²⁶ However, biopsies carry a high risk of infection and tumor seeding, and should never be done transperitoneally, transretroperitoneally, transrectally, or transvaginally.²⁷ A cystic lesion, as seen in our case, does not warrant a pre-operative biopsy as these lesions are usually benign in nature and do not require additional therapy. α -fetoprotein, carcinoembryonic antigen, and human chorionic gonadotropin are not useful markers to differentiate between benign and malignant lesions. However, malignant teratomas have high rates of recurrence and these tumor markers could be used to monitor for recurrence after surgical excision.²⁸ Negative margins (R0) should be achieved when excising malignant teratomas.²⁶ If the pathology report demonstrates a malignant teratoma it is recommended that the patient undergoes adjuvant chemotherapy and radiotherapy. However, limited data exists on the use of adjuvant therapy in these patients, and there are currently no proven benefits nor guidelines

regarding the use of adjuvant therapy in malignant teratomas.²⁶ As such, surgical excision remains the gold standard of treatment.

Due to the rarity of this diagnosis, there is no published consensus on its management apart from surgical excision. Location of the mass will dictate the selection of the surgical approach. An operation can be performed via the open or laparoscopic abdominal approach, perineal approach, or a mixed approach based on the location of the mass and the surgeon's preference and experience. The laparoscopic excision of a retrorectal teratoma was first reported in 1995; however, there have been scarce reports since then describing a laparoscopic approach to these specific tumors.²⁹ A robot-assisted laparoscopic approach is even more scarcely reported in the literature.³⁰ While a robot-assisted laparoscopic approach has been successfully reported in previous literature, our facility did not have a robotic surgical system in order to consider this option. The laparoscopic approach does have drawbacks, including the need for advanced surgical training and the lack of direct touch to discern tumor boundaries. Noting its challenges, we recommend a laparoscopic approach as the advantages are statistically significant across the literature comparing open to laparoscopic approaches. There is less postoperative pain, faster patient recovery, better cosmetic appearance of scars, lower mortality rates as well as lower morbidity rates when looking at bleeding, adhesions, and infections along with additional complications.^{10,29,31,32} Our case demonstrates that laparoscopic excision is a feasible and promising approach to management of a sacrococcygeal teratoma. Regardless of the approach undertaken by the surgeon, complete excision without rupture ensures that the risk of recurrence is extremely low.

LESSONS LEARNED

Mature cystic teratomas are rare in the adult population, and much rarer in the male sex with detection only on imaging after presentation with vague symptoms related to mass effect. Imaging only yields further diagnostic uncertainty given the broad differential. Surgical excision is the standard of care despite no strong evidenced based literature on its management. Laparoscopic excision is a feasible and promising approach to management of these tumors.

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