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Extragonadal Perirectal Mature Cystic Teratoma in the Adult Male

Pranamaya Mahankali
Rowan University

Liam Trimble
Rowan University

Diana Panciera
Rowan University

Hui Li
Rowan University

Thaer Obaid
Rowan University

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Mahankali P, DO; Trimble L, DO; Panciera D, DO; Li H, MD; Obaid T, MD
Jefferson Health – New Jersey

Background

A teratoma is a mass that originates from the germ cells of the three primitive embryonic layers and commonly arises from the ovaries or testicles, thereby characterized as gonadal in origin. These masses comprise different types of tissues such as fat, hair, muscle, and bone[1]. Extragonadal teratomas are uncommon and arise from anatomical locations other than the gonads, such as the pineal gland, anterior mediastinum, retroperitoneum, and sacrococcygeal regions[2]. The most common is the presacral region, as it contains tissue derived from all germ layers. Tumors in this specific location are far more common in neonates, infants, and children rather than adults, and females rather than males[3]. The patient in this case, an adult male, was found to have a sacrococcygeal teratoma, making this case exceptionally unique. The teratoma was appropriately excised, and done so laparoscopically – an approach with little published literature regarding these specific tumors. Given the exceedingly rare nature of sacrococcygeal teratomas in adults, especially males, there is little literature on the subject in regard to management.

Case Presentation

A 49-year-old male initially presented to a urology office following an episode of asymptomatic hematuria. CT urogram was obtained, noting the presence of a large, 8.8 x 6.1 x 5.9 cm extraperitoneal perirectal mass within the left perirectal space, abutting the left aspect of the rectum and displacing it to the right (Figure 1). This was noted to be distinct and separate from the rectum, prostate, bladder, and ureters. The genitourinary tract was unaffected, prompting referral to Colorectal Surgery for further evaluation.

Apart from hematuria the patient remained asymptomatic, denying any pelvic pain, sexual dysfunction, bowel, or bladder symptoms. The patient's past medical and surgical history were only pertinent for Clostridium difficile infection along with two unremarkable colonoscopies in 2013 and 2018. Physical examination was unremarkable without any abdominal tenderness or fullness. Digital rectal examination revealed no external pathology nor visible or palpable masses. A magnetic resonance imaging (MRI) pelvis was obtained to further characterize the mass, which again demonstrated a large, well-defined extraperitoneal perirectal mass located in the left perirectal space, abutting and displacing the rectum to the right (Figure 2). 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 3 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.

The patient subsequently underwent a laparoscopic excision of this perirectal mass. The anterior peritoneal reflection was dissected to encounter a large tan cystic mass which was circumferentially dissected from the surrounding tissues and completely removed. There was no pelvic or abdominal adenopathy, ascites, or additional intra-abdominal gross abnormalities. Grossly, the specimen appeared tan with an outer rough and irregular surface (Figure 3a). Intracystic components were copious, thick and yellow cheese-like with multiple hair shafts (Figure 3b). Microscopic tissue examination revealed a cystic lesion lined by keratinized stratified squamous epithelium with skin appendages including hair shafts without any evidence of malignancy or immature components, consistent with abdominopelvic extragonadal mature cystic teratoma (Figure 4a, 4b).

Imaging

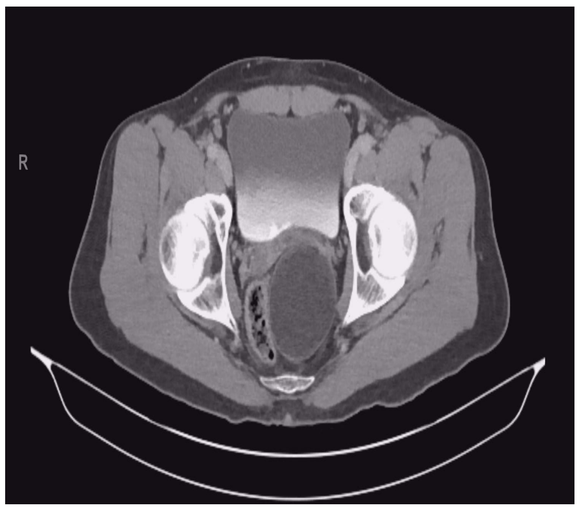

Figure 1: Computed tomography of the abdomen and pelvis (axial view) demonstrating an 8.8 x 6.1 cm homogenous, extraperitoneal perirectal mass located within the left perirectal space abutting the rectum, without invasion, with an approximately 3 mm layer of separation. No peripheral enhancement, calcifications, or lymphadenopathy is noted.

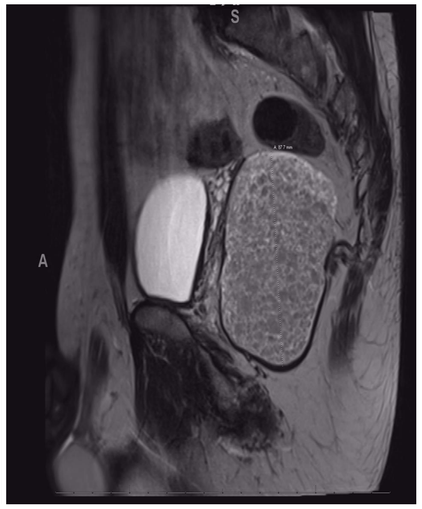

Figure 2: T2 weighted magnetic resonance imaging of the pelvis (sagittal view) demonstrating the large mass inferior to the anterior peritoneal reflection, confirming extraperitoneal origin of the mass and it's location within the left perirectal space.

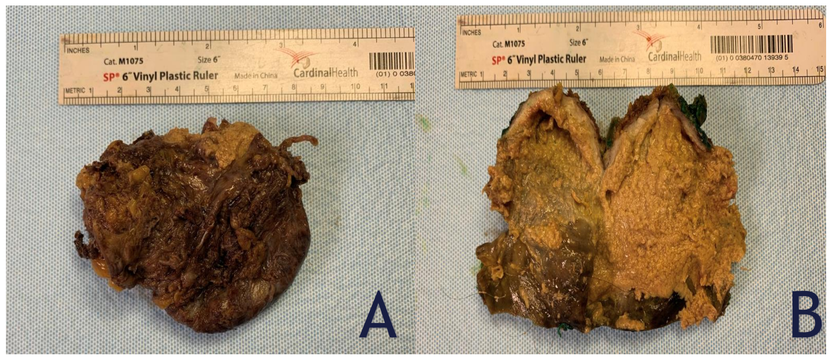

Figure 3: Gross pathological examination of extragonadal mature cystic teratoma. A) Rough tan colored outer surface. B) Cyst predominantly filled with sebaceous material and hair follicles.

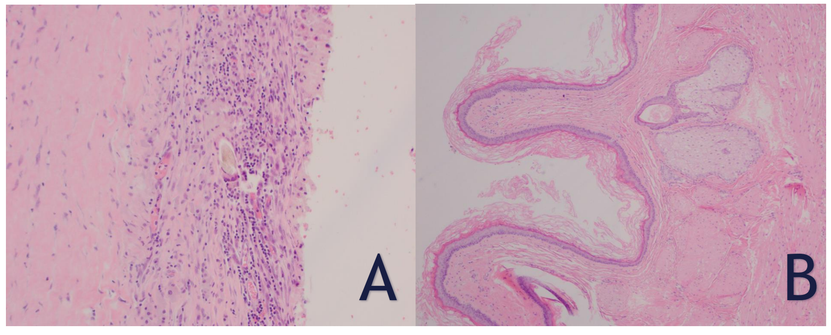

Figure 4: Microscopic examination of extragonadal mature cystic teratoma. A) H&E, hair shaft. B) H&E, 4x, keratinized stratified squamous epithelium.

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. Sacrococcygeal teratomas (SCTs) have an incidence of 1 in 35,000 - 40,000 live births, making them common tumors in the neonatal, infant and child population, but a rare entity in the adult population[4]. In accord with cases reported in the literature, there is also a female predominance of 3-4:1[3]. Although the majority are benign, there still persists a 20-30% risk of infection and a 1-12% risk of malignant transformation, which increases with age. The standard of care is surgical excision[5]. The role of surgery in this case is two-fold, as it is diagnostic as well as curative.

Adult patients with SCTs will typically present with nonspecific or subtle symptoms of a mass effect, otherwise the majority will be diagnosed incidentally via imaging studies or 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 in literature include rectal, back, or pelvic pain, paresthesias, saddle anesthesia, urinary urgency or retention, frequent urge to defecate, incomplete evacuation, constipation, and loss of bladder or rectal control[6].

Radiological imaging aids in the diagnosis of these masses along with assisting in surgical planning by defining their relationships with surrounding tissues. Initial CT identified a pelvic mass in our patient, further characterized with MRI to aid with our differential diagnoses. As previously mentioned, our differential diagnoses included dermoid cyst, epidermoid cyst, liposarcoma, and teratoma. Definitive diagnosis of SCT was obtained after surgical excision and histological examination of the mass.

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. 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[7]. Regardless of the approach undertaken by the surgeon, complete excision without rupture ensures that the risk of recurrence is extremely low.

Conclusion

Albeit rare in the adult population, and much more rare in the male sex, sacrococcygeal mature cystic teratoma is a plausible differential diagnosis for a pelvic mass. Detection of these masses is often first discovered on imaging after presentation with vague symptoms, secondary 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. Our case demonstrates that laparoscopic excision is a feasible and promising approach to management of a sacrococcygeal teratoma.

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