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A Case Report of Neuroendocrine Tumor in Presacral Region: How Can It Be Managed? Laparoscopy versus Laparotomy

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Abstract

Presacral or retrorectal tumors are rare, usually asymptomatic, and diagnosed accidentally during physical examination or imaging. Symptomatic tumors may present with perianal pain, bowel dysfunction, and urinary symptoms due to the mass compression or invasion of the surrounding tissues and organs. Surgical resection is the first choice for treating presacral tumors. Clinicians should choose surgical procedures based on the location and size of the tumors. We presented a 43-year-old woman who suffered from pelvic pain and primary infertility from two years ago. A large mass between the posterior vaginal wall and the rectum was found on recto-vaginal examination. Magnetic resonance imaging (MRI) revealed a large 120×115 mm benign multiloculated cystic mass. Eventually, the mass was removed through laparoscopic surgery. The pathology report indicated a carcinoid tumor (grade I) with no lymphovascular invasion. Thus, presacral tumors are resectable through laparoscopy with lower complications than open surgery.

Keywords: Carcinoid Tumor, Case Reports, Laparoscopy, Neuroendocrine Tumors

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Introduction

Presacral or retrorectal tumors are rare, with 1.4 to 6.3 reported cases per year, mostly discovered accidentally. Patients with presacral tumors are usually asymptomatic and are diagnosed during physical examination (digital rectal examination) or imaging. Moreover, symptomatic tumors may present with perianal pain, bowel dysfunction, and urinary symptoms due to the mass compression or invasion of the adjacent tissues. As a result, it is challenging for clinicians to diagnose and treat them in a timely manner (1).

The first choice for treating presacral tumors is surgical resection. Clinicians should choose surgical procedures based on the location and size of the tumors according to the preoperative examination and imaging (2).

Herein, we present a case diagnosed with a bulky presacral carcinoid tumor resected through laparoscopy.

Case report

A 43-year-old woman was referred to us from the infertility center with a history of pelvic pain and primary infertility from two years ago. The last menstrual period (LMP) of the patient was 15 days before the visit, and she had an irregular menstrual cycle with increased bleeding volume. Initially, the patient denied any complaint of abdominal pain and urinary symptoms. She did not complain of sweating, weight loss, or anorexia. The patient's familial history, past medical history, and drug history were negative. She was hemodynamically stable with a blood pressure of 110/80 mmHg, a pulse rate of 85/minutes, an axillary temperature of 37ºC, and a respiratory rate of 19 /minutes. The patient's body mass index (BMI) was 27.7 kg/m^2. Physical examination of the abdomen revealed a surgical scar resulting from a previous ovarian cystectomy about 11 months ago, without a palpable mass, and there was an exophytic wart on the labia major. On recto-vaginal examination, a large mass between the posterior vaginal wall and the rectum was found. On speculum examination, the cervix was invisible.

The patient's laboratory tests were normal (WBC: 6300 Mill/cumm, Hemoglobin: 12 g/dL, PLT: 299000×1000/cumm, Creatinine: 0.9 mg/dL, AFP: 3.4 IU/mL, CA-125: 19 IU/mL, CA19-9: 5.8 IU/mL). Ultrasound revealed a large multiloculated cystic 119×82 mm mass near the left ovary. magnetic resonance imaging (MRI) illustrated a large 120×115 mm benign multiloculated cystic mass, but some were restricted with thin peripheral enhancement (Fig.1). Eventually, the mass was removed through laparoscopic surgery. The pathology report indicated a grade I carcinoid tumor with mitosis <2/10 high-power fields (HPFs), with no lymphovascular

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invasion. The immunohistochemistry (IHC) result showed synaptophysin-positive/chromogranin: positive (Fig.2). The patient was discharged after the surgery with no complications, and after six months the patient had no complaints.

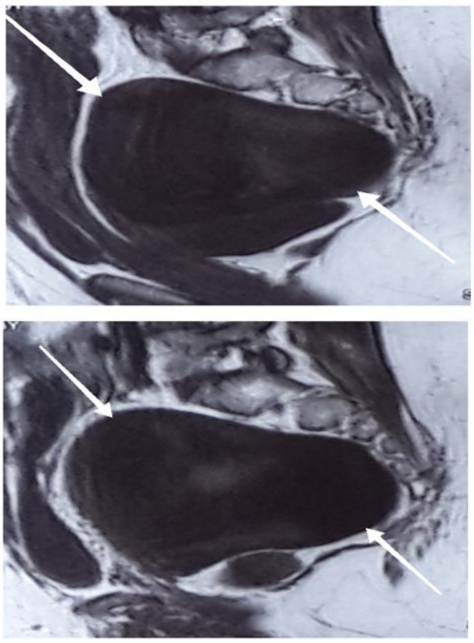

Fig.1: Magnetic resonance imaging (MRI) presents a large 120×115 mm benign multiloculated cystic mass that is attached to the thecal sac, some portion of the lesion is restricted with thin peripheral enhancement.

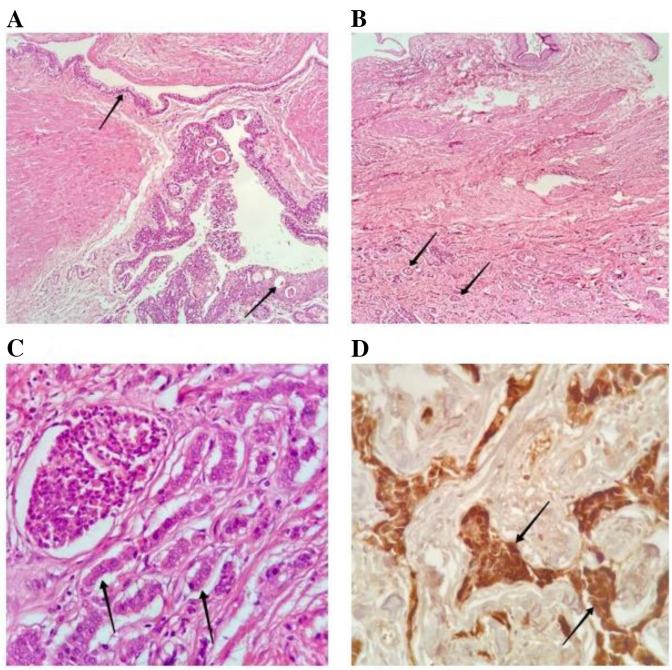

Fig.2: Pathological images of the mass. **A.** shows the cyst lining consists of cuboidal to columnar epithelium with intracytoplasmic mucin and foci of squamous metaplasia (100x, H&E staining). **B.** Focal neoplastic proliferation in the cyst wall is seen (40x, H&E staining). **C.** The neoplastic cells are composed of trabecular structures and nests of uniform polygonal cells with round to oval nuclei, salt and pepper chromatin, and moderate cytoplasm (100x, H&E staining). **D.** The neoplastic cells show immunopositivity for chromogranin (IHC study, 100x) and so the diagnosis of well-differentiated neuroendocrine tumor, grade 1 (carcinoid tumor) is confirmed.

This case report was approved by the Institutional Research Ethics Committee and Vice Chancellor in Research Affairs of Shahid Beheshti University of Medical Sciences (IR.SBMU.RETECH.REC.1401.638).

Discussion

Presacral space is an extraperitoneal fossa between the upper two-thirds of the rectum and the sacrum (3). There are several main vascular and neural structures in this fossa; thus, their injury may have considerable physiologic rectoanal, neurological, and musculoskeletal consequences (4). Due to the improvements in diagnostic methods, the number of reported cases of neuroendocrine tumors (NETs) has increased during the past decades (5).

NETs originate from neuroendocrine cells and usually occur in the gastrointestinal and bronchopulmonary tract (approximately 70 and 25%, respectively) (6). NETs account for about 0.5% of all newly diagnosed malignancies and are more common in women than in men. Also, NETs in the retroperitoneal space are extremely rare. They can arise from primary presacral neoplasms or metastasis of rectal carcinoids (7). The association between presacral carcinoid tumors and tailgut cysts suggests that both originate from the hindgut (8). Presacral NETs are usually asymptomatic; however, they may manifest with pelvic pain, rectal fullness, and constipation due to the mass effect (9). They usually present no carcinoid syndrome, such as flushing, sweating, or hypertension. Our patient suffered from chronic pelvic pain without other mass-related symptoms (e.g., constipation and rectal fullness) or carcinoid syndrome. Since NETs are usually asymptomatic, they are typically diagnosed accidentally through physical examination or imaging. Our patient's recto-vaginal examination showed a large mass between the posterior vaginal wall and the rectum (10).

Differential diagnoses of presacral NETs include presacral hypervascular masses, such as paraganglioma, extraintestinal gastrointestinal stromal tumor (GIST), and solitary fibrous tumor, distinguished by imaging. On MRI, paraganglioma is presented by a well-circumscribed, intensely enhanced mass. Extraintestinal GIST typically contains necrotic centers, hemorrhage, or cystic degeneration, tending to be aggressive. The solitary fibrous tumor presents as a well-circumscribed solid mass with intense heterogeneous enhancement (10).

On MRI, our case had a 120×115 mm benign multiloculated cystic mass with thin peripheral enhancement in the presacral region. MRI is the preferred modality for diagnosis before surgery because it designates local invasion and neural involvement with advanced contrast resolution compared to computed tomography (CT) (11).

In NETs, well-differentiated tumors, the cells produce neurosecretory granules, reflected in the strong and diffuse immunoexpression of neuroendocrine markers (e.g., chromogranin A and synaptophysin) (12). Our patient's mitotic index was<2/10 HPFs, so she had a G1

carcinoid tumor.

Presacral NETs are usually treated by resecting the primary tumor and closely following the patient after the procedure (5). Due to the location of presacral tumors, the treatment of choice for them is usually open surgery (13). There are three approaches for resecting presacral tumors: the anterior (transabdominal) approach, the posterior (transsacral or transanal) approach, and a combination of both (2). Nedelcu et al. (14) suggested laparoscopic surgery for presacral tumors that are supposed to be benign with no invasion to other organs or bones with a size of less than 6 cm. Laparoscopic surgery in presacral tumors has the following advantages: smaller incisions, milder postoperative pain, better field of vision, and the ability to precisely differentiate between the tumor and adjacent structures (15). Although our case had a 12 cm mass in the presacral region, the tumor did not invade other organs based on MRI results. Thus, we decided to resect it through laparoscopy surgery. Informed consent was obtained from the candidate patient for the laparoscopic anterior approach. Pre-surgery procedures, including bowel preparation and thromboprophylaxis were performed. The patient was placed in the lithotomy position. Prep and drape done, a 10 mm trocar was inserted from the umbilicus by a direct entry method; a 5 mm trocar was inserted into the lateral rectus and supra pubic muscles. Using the camera, the right side of the pelvis was observed. According to the mass location on the MRI, the surgeon noticed a brief bulging in the posterior cul de sac, which had caused the deviation of the rectum to the left side. Then, a peritoneal dissection was performed on the right side of the pelvis above the prominence of the mass. After identifying the right ureter, the right pararectal space and the recto-vaginal septum were dissected. When the mass became observable, an attempt was made to separate it from the space in front of the sacrum and behind the rectum.

At the bottom of the mass, a solid area of about 6-7 cm was visible, which was separated from the sacrum with difficulty. Despite careful hemostasis, venous bleeding occurred. Thus, the hemostatic powder was used, and tranexamic acid was injected. After the fixation of the hemovac drain, the peritoneum was repaired. The amount of bleeding during surgery was estimated to be 1000 cc. Due to a severe drop in hemoglobin (from 12 to 8.4 mg/dl), two units of packed red blood cells and two units of fresh frozen plasma (FFP) were injected. After 48 hours, she was discharged with good general condition. At the outpatient visit for follow-up, she complained of pelvic pain, which was formed in the re-MRI of an organized hematoma at the site of the mass, which was absorbed in the subsequent follow-ups.

Due to the anatomical proximities, bleeding from the tumor-feeding vasculature and the presacral venous plexus should be considered in presacral tumor surgeries. Previously, several patients with malignancies underwent laparoscopic surgeries. These cases are followed for possible uncontrolled bleeding and metastasis (2). The most common complications include bleeding, wound infection, and injury of the urethra or rectum (13). Another study showed among 11 reported cases of primary presacral NETs with a mean follow-up time of 41.8 months, the patients with well-differentiated NETs and local disease had good prognoses (5). Similarly, we followed up of our case for six months, and her conditions turned out to be good, with no complaint or recurrence.

Conclusion

We reported a rare presacral carcinoid tumor of about 12 cm in size. Finally, the patient underwent laparoscopic resection without any complications. Our case revealed that laparoscopic surgery is a feasible option for resecting presacral tumors because of its advantages (less invasive and small skin incision); however, this finding requires further investigations.

Acknowledgments

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Authors' Contributions

B.N.; Conception, design, and supervision. H.N.; Drafting of the manuscript and critical revision of the manuscript. Sh.H.Ch; Analysis and interpretation of data. All authors read and approved the final manuscript.

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