Case Report
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Disseminated Metastasis after Resection of Sacrococcygeal Teratoma with Mucinous Adenocarcinoma: A Case Report

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Abstract

Mature teratoma is a common tumor that can undergo malignant transformation, either in ovarian or extragonadal sites. While adenocarcinoma superimposed on sacrococcygeal teratoma is rare, mucinous variants have been reported in only five cases. Here, we present a case of a young girl with disseminated metastasis of mucinous carcinoma, initially of unknown primary origin. Further investigation by a dedicated multidisciplinary team (MDT) revealed a focus of mucinous carcinoma (intestinal type) in a sacrococcygeal teratoma incompletely resected five years earlier. The patient is currently undergoing second-line chemotherapy after experiencing side-effects on the first-line regimen. Pathologists, gynecologic oncologists, and surgical oncologists should exercise caution when dealing with locally aggressive teratomas, thoroughly searching for malignant components and conducting short-term follow-up.

Keywords: Teratoma, Adenocarcinoma, Mucinous, Unknown primary, Sacrococcygeal

Introduction

Mature teratoma of the ovary is the most common neoplasm, representing 20% of all ovarian tumours. Although rare, malignant transformation of teratoma has been described in 1%-2% of cases.[1] The commonest associated malignancy was squamous cell carcinoma, adenocarcinoma and carcinoid tumour, in this order.[2, 3] In addition, it has been recognized that the prognosis of carcinomas associated with teratoma is worse than that of epithelial ovarian carcinomas, regardless of whether postoperative chemotherapy or radiotherapy is given. This may be explained by the

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rarity of this tumour, posing a significant challenge to developing standardized adjuvant management protocols.[4]

Sacrococcygeal teratomas are relatively uncommon, and most are generally present since birth. However, when sacrococcygeal germ cell tumours are predominantly intrapelvic, they may be asymptomatic for years and are at risk for malignant transformation.[5]

We herein report what might be the sixth case in literature of malignant transformation arising in a sacrococcygeal teratoma.

Case Presentation

A 22-year-old female patient presented with left supraclavicular, left pelvic, and upper thigh swellings to the outpatient clinics of the Oncology Center at Mansoura University in late December 2019. There was no medical history, but the patient had undergone excision of a cystic teratoma in the sacrococcygeal region five years ago. Revising the computed tomography (CT) dating back to 2014 of this lesion, the CT showed a well-defined large hypodense cystic lesion in the presacral space posterior to the rectum, compressing and displacing it anteriorly and contacting the sacrum posteriorly, measuring 14 × 13.5 × 11 cm. The lesion also showed a small projection in its posterolateral wall with fine calcifications. Additionally, post-contrast magnetic resonance imaging (MRI) showed a well-defined lesion in the presacral region, mainly cystic, measuring

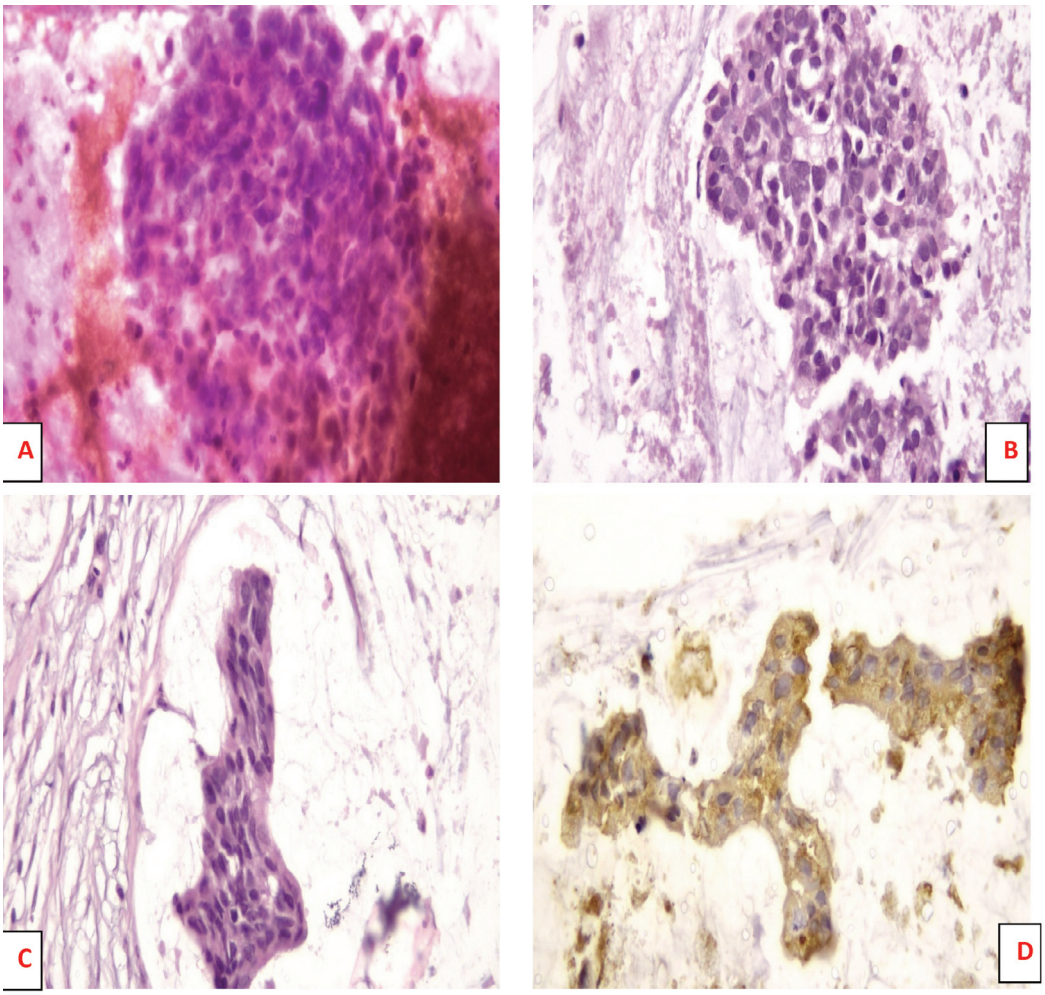

Figure 1. Fine needle aspiration cytology from a cervical node showing aggregates of malignant cells (A, smear, H&E, 40×) with associated pools of mucin (B, cell block, H&E, 40×). Histopathologic examination of the true-cut biopsy of an iliac mass showed neoplastic cells floating in pools of extracellular mucin (C, H&E, 40×). Immunohistochemical expression for CK20 was detected in tumor cells (D, H&E, 40×).

15 × 12.5 × 11.5 cm with a small solid component measuring 4 × 3.5 × 3.3 cm with the same relations described in the CT. The lesion was incompletely excised with extensive bleeding from the presacral plexus, leaving residue on the sacrum five years ago. The post-resection pathological analysis revealed a benign cystic teratoma with atypical epithelial proliferation within the mucoid background (no further immunohistochemistry (IHC) was done at that time).

In December 2019, the patient came because of neck swelling, and ultrasonography revealed suspicious lymph nodes on the neck in the left lower deep group, irregular in shape with lost hilum, measuring 2.6 × 1.6 cm. Non-contrast pelvic and upper thigh MRI showed a large soft tissue lesion displacing the intestinal loops and the urinary bladder, extending to the upper abdomen superiorly and inferiorly to the upper left thigh through the left obturator foramen, with the possibility of recurrence of the previous mass or a malignant left ovarian mass.

Fine needle aspiration cytology (FNAC) from the suspicious left deep cervical lymph node showed a malignant smear of metastatic mucoid adenocarcinoma (Figures 1A and 1B). Core needle biopsy from the left iliac mass showed a picture compatible with invasive mucinous carcinoma that showed a positive immunohistochemical reaction for CK20 with a negative reaction for CK7 and CDX2 (Figures 1C and 1D). The tumor was MSI stable/MMR proficient.

Tumor markers were as follows: CA-125 = 17.05, CA-19-9 ≤ 0.6, and carcinoembryonic antigen (CEA) = 113, where CEA was later raised to 147 while on chemotherapy.

Three months later, a CT scan showed generalized lymphadenopathy, bilateral external and internal iliac, inguinal, and abdominal lymph nodes measuring 3.8 × 2.4, 2.5 × 2.3, and 3.5 × 2 cm, respectively, with amalgamated lymph nodes in the left lower deep cervical and supra-clavicular region measuring 4.5 × 2.3 cm. Additionally, multiple bilateral pulmonary nodules, the largest

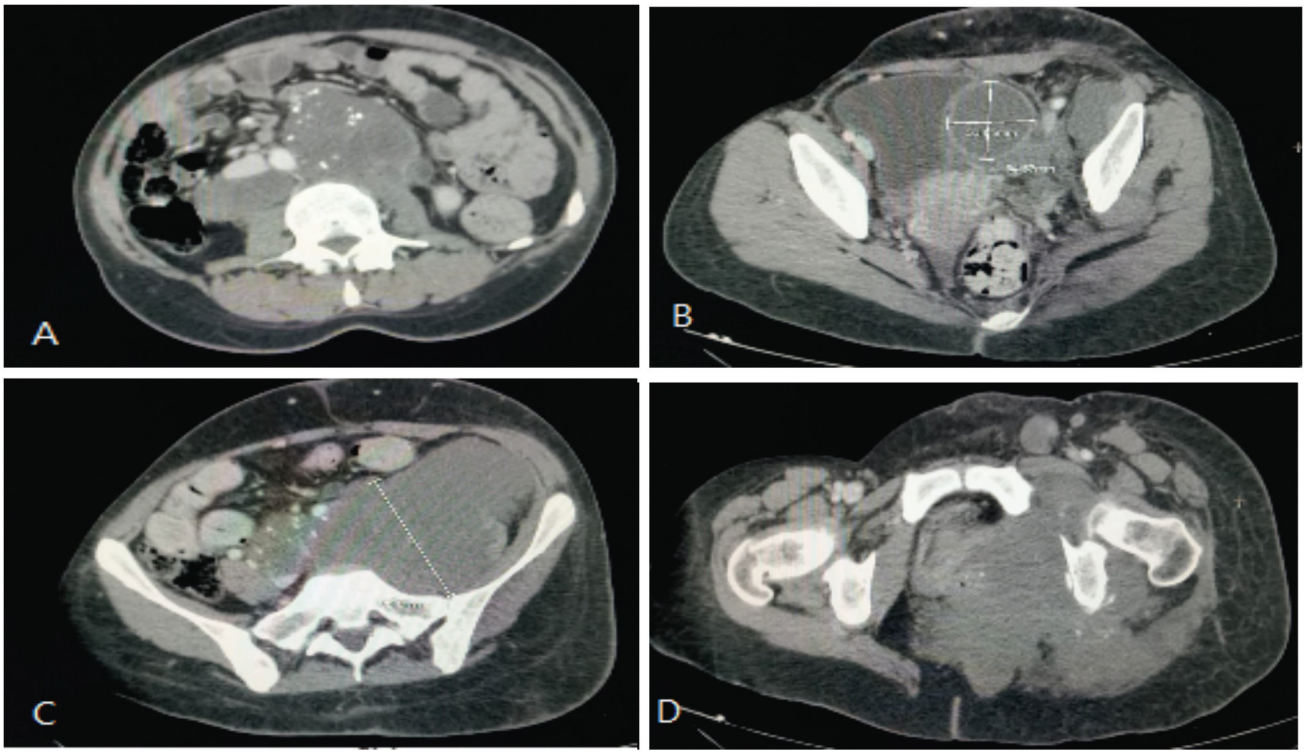

Figure 2. Computed tomography scan showing A) a hypodense soft tissue retroperitoneal mass in the pelvis with foci of calcification, B) a mass inseparable from the bladder, indenting its left wall, C) a mass inseparable from the left ilio-psoas muscle, and D) another similar mass seen in the left side of the pelvic wall and left peritoneal region, inseparable from the endocervix, ischial and pelvic bones, and inseparable from the left gluteus, obturator, and left levator-ani muscles.

being 9 mm on the left lower lobe, were identified, along with a hypodense soft tissue retroperitoneal mass that was inseparable from the left ilio-psoas muscle, encasing the lower abdominal aorta, its bifurcation, and left iliac vessels, showing foci of calcification and measuring 17.9 × 11.4 × 8.6 cm. Another similar mass was seen in the left side of the pelvic wall and left peritoneal region, inseparable from the internal cervix, ischial and pelvic bones, inseparable from left gluteus, obturator, and left levator-ani muscles, measuring 13.2 × 12.7 × 9.5 cm. The masses were associated with mild to moderate left hydronephrosis (Figure 2 A-D).

The patient was treated as a case of metastasis of unknown origin (MUO) of probably gastrointestinal tract (GIT) origin with XELOX for three cycles, where a sensitivity reaction to oxaliplatin developed, and the patient refused to continue. Thereafter, the patient was shifted to weekly paclitaxel + carboplatin as a case of metastasis of unknown primary of germ cell tumor origin, as the patient clinically progressed on XELOX and CEA levels increased.

Follow-up CT scans showed a stationary course on chemotherapy. Considering the stationary

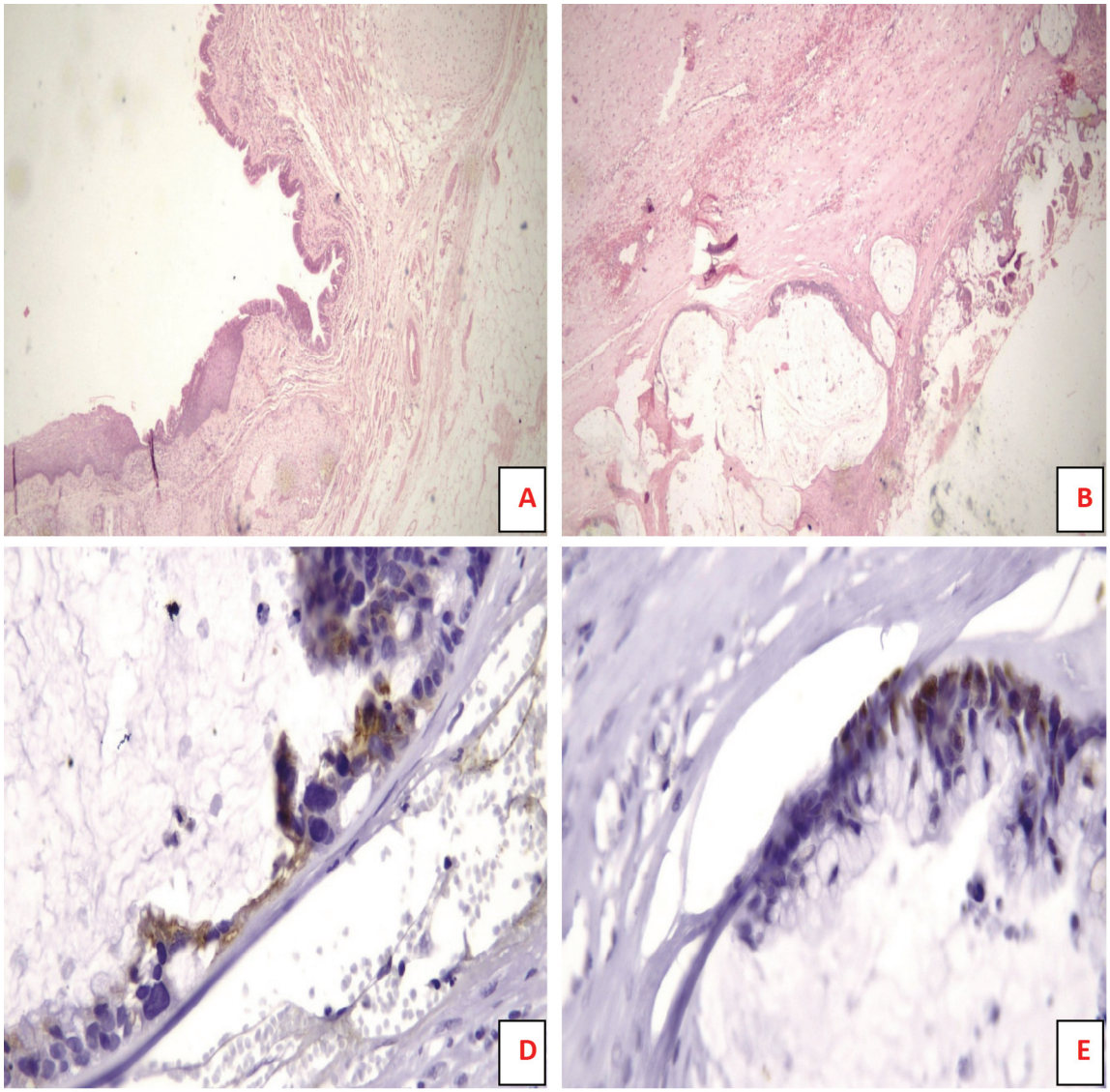

Figure 3. The mature teratoma was composed of cysts lined by mature squamous epithelium simulating skin, with adnexa, as well as foci of benign respiratory-type epithelium and cartilage (A, H&E, 4×). Associated foci of invasive mucinous adenocarcinoma (B, H&E, 4×) showed neoplastic cells floating in pools of extracellular mucin (C, H&E, 40×). Immunohistochemical expression for CK20 (D, H&E, 40×) and CDX2 were detected in tumor cells (E, H&E, 40×).

course of the disease and the unrivaled site of origin by imaging, immune stain, and markers, the old pathology of presacral teratoma was retrieved and re-examined. Surprisingly, a small focus of invasive mucinous carcinoma (intestinal type) was detected with IHC for CK20 and CDX2, revealing focal positive reaction (Figure 3 A-E). The differential diagnosis was ovarian mucinous carcinoma on top of the teratoma or metastatic mucinous carcinoma of GIT origin. However, based on the long history, the presence of incompletely resected teratoma, the lack of response to GIT type chemotherapy, and the known lower CDX2 positivity in ovarian than GIT mucinous tumors, the panel concluded that the diagnosis of primary carcinoma superseded teratoma.

Ethical approval

This case report has been approved by the Institutional Review Board (IRB) of the Mansoura Faculty of Medicine with code R.22.12.1981.

Discussion

The authors herein report the 6th case with this pathology up to our knowledge. The long follow-up sequence of the patient is a point of strength; however, the non-accurate diagnosis and incomplete cytoreduction done at the initial surgery for the patient may be a point of weakness.

Germ cell derivative neoplasms commonly involve sites such as ovaries, followed by testis, anterior mediastinum, retroperitoneum, and sacrococcygeal area. Sacrococcygeal teratoma comprises the most common congenital tumor in the neonatal group with a female predilection; a 4-fold increased incidence is observed in females. However, its incidence in the adult population is relatively uncommon.[6, 7] Malignant transformation occurs in either the embryonic or somatic components, with a low incidence rate of 1%-3% both.[8]

Depending on the tumor extension, the sacrococcygeal tumors are divided into four types. Type I sacrococcygeal tumor has a minimal presacral component with a predominant external component; type II tumors are also predominantly external with definitive intra-abdominal extensions; type III tumors are located mostly in the abdomen or comprise of a dominant pelvic mass extending into the abdomen with a small external component; and type IV are entirely in the presacral region with no external component.[7]

There have been only 2 reported cases of extragonadal teratoma-associated mucinous tumors with pseudomyxoma peritonei, and both had an adverse outcome, suggesting that they may be more aggressive than their gonadal counterparts.[9, 10]

It is difficult to make a diagnosis of malignant transformation of a teratoma preoperatively. However, MRI findings may be helpful in distinguishing malignant transformation from benign neoplasms. Several reports have revealed that an important feature of the malignant transformation of teratoma was the existence of an enhanced solid component,[11] which was also noted in our patient. In addition, a rising CEA was detected in a handsome number of patients,[12] together with our patient, making it a useful aid in diagnosis.

The outcomes for patients with advanced stage mucinous adenocarcinoma superimposed on teratoma are generally poor, and are claimed to be even worse than those of squamous carcinoma arising on teratoma.[13] Surgical resection aimed at complete cytoreduction and avoiding artificial rupture of the tumor should be opted for to prevent recurrence.[12] Unfortunately, this was not the scenario in our patient, given the lack of such knowledge at the time of surgery.

Previously, there were reports of three females[14-16] and two males[7, 17] with mucinous carcinoma arising in a sacrococcygeal teratoma, along with the current report of a female patient. The mean age of the five patients was 49.4 years old, with a median size of 13.5 cm, ranging from 7 to 30 cm in largest diameter. Manifestations vary from pelvic pain, difficult micturition and/or defecation, abdominal swelling, and weight loss. The pathology was mucinous carcinoma in four patients[7, 14, 16, 17] and signet ring carcinoma in one of them.[15] Three received adjuvant chemotherapy;[7, 15, 17] however, no long-term outcome was reported except in the current patient

(seven and a half years since detection of the pelvic teratoma until the last visit).

Conclusion

In conclusion, mucinous carcinomas in sacrococcygeal teratomas are extremely rare. They usually present with non-specific symptoms and are only detected when they have grown to a significant size. Currently, there is no clear guidance regarding adjuvant therapy; however, chemotherapy extrapolating experience in colorectal cancer is commonly used. Long-term outcomes are unknown, although we reported a disseminated metastasis years later. Familiarity of pathologists with this entity and patient counseling with a clear follow-up plan in case of non-straightforward resection of teratomas should be encouraged.

Informed Consent

Informed consent was obtained from the patient.

Conflict of Interest

None declared.

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