

Primary yolk sac tumor in the abdominal wall in a 20-year-old woman: A case report

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Abstract

BACKGROUND

Extragenital yolk sac tumors (YSTs) are rare, with only a low reported tumor occurrence outside the gonads locally and abroad. Extragenital YSTs are usually a diagnostic challenge, because they are infrequent, but also because a thoughtful and detailed differential diagnostic process must be performed.

CASE SUMMARY

Here we present a case of an abdominal wall YST in a 20-year-old woman admitted with a tumor in the lower abdomen close to the umbilicus. The tumorectomy was performed. The histological examination revealed characteristic findings such as Schiller-Duval bodies, loose reticular structures, papillary structures, and eosinophilic globules. According to the immunohistochemical staining, the tumor tissue was positive for broad-spectrum cytokeratin, Spalt-like transcription factor 4, glypican-3, CD117, and epithelial membrane antigen. Based on the clinical information, histological features, and immunohistochemical staining profile, the tumor was diagnosed as a YST present in the abdominal wall.

CONCLUSION

Based on the clinical information, histological features, and immunohistochemical staining profile described above, the tumor was diagnosed as a primary YST in the abdominal wall.

Key Words: Yolk sac tumor; Abdominal wall; 20-year-old woman; Case report

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Core Tip: Extragenadal yolk sac tumors (YSTs) are usually a diagnostic challenge, because they are infrequent, but also because a thoughtful and detailed differential diagnostic process must be performed. In this case, since the tumor presented classic yolk sac tumor features, and immunohistochemistry was concordant, the diagnostic process was more straightforward, but not simplistic. And this is the first reported case of a primary YST in the abdominal wall of an adult.

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INTRODUCTION

Yolk sac tumor (YST), also known as endodermal sinus tumor or primitive endodermal tumor, is a rare and highly malignant tumor of germ cell origin. It is common in infant and adolescent children with a median age at diagnosis of 19 years, with 40% of patients diagnosed in the prepubertal period[1,2]. This tumor occurs mostly in the ovary and the testis, and it rarely occurs outside the gonads, with only 10%-15% of YST cases reported locally and abroad[3]. Extragenadal YST can occur in the mediastinum [4,5], retroperitoneum[6], cervix[7], endometrium[8], sacrococcygeal region[9], vagina[10,11], omentum [12], urachus[13], and liver[14]. Moreover, the clinical manifestations vary according to the site of tumorigenesis, which often makes the clinical diagnosis difficult. Some cases lack typical structures for proper morphological observation, while others have diverse pathological morphology.

YSTs arising in the abdominal wall are even rarer, with less than 2 cases of primary abdominal wall YST reported in the literature[15,16]. Here, we report a special case of primary YST in the abdominal wall of a 20-year-old woman, which to the best of our knowledge, is the first report of this disease in an adult.

CASE PRESENTATION

Chief complaints

A 20-year-old Chinese woman was admitted to the First Affiliated Hospital of Jinzhou Medical University in January 2022 with a lower abdominal mass, which had progressively enlarged since November 2021. It was not painful or itchy.

History of present illness

Palpation revealed that the mass was located in the hypogastric region, with a size of about 5 cm × 3 cm, The lesion was well defined, was not capsulated, normal skin without redness or swelling, poor range of motion, and no tenderness.

History of past illness

The patient has no past medical history.

Personal and family history

The patient denied any family history. Healthy parents, no siblings.

Physical examination

On physical examination, the vital signs were as follows: Body temperature, 36.3 °C; blood pressure, 113/63 mmHg; heart rate, 86 beats per min; respiratory rate, 20 breaths per min. Palpation revealed that the mass was located in the hypogastric region, with a size of about 5 cm × 3 cm, no redness and tenderness.

Laboratory examinations

The results such as routine hematological testing, blood sedimentation rate, and tumor-associated markers were normal.

Imaging examinations

Because the location of the tumor was relatively superficial, there was no imaging information at the beginning. The Enhanced Computed tomography (CT) examination revealed no abnormalities in the

abdominal organs and other regions, so this case was considered a primary tumor in the abdominal wall (Supplementary Figure 1).

FINAL DIAGNOSIS

A primary YST in the abdominal wall.

TREATMENT

The patient underwent subcutaneous tumor resection. The mass was found to be located in the deep layer of subcutaneous fat, and the entire mass was completely dissected and sent for biopsy. We found the cut surface of the tumor was greyish-red and greyish-yellow in colour, soft to medium in quality, with visible necrosis in some areas (about 10%) (Supplementary Figure 2). It was first considered as a possible soft tissue tumor or metastatic carcinoma.

After the tumor tissue was resected, it was fixed with 10% neutral-buffered formalin and then embedded in paraffin blocks. The tissue blocks were cut into 4- μ m sections. Then, the hematoxylin and eosin-stained sections were used for histological evaluation. Histological findings from the resected tumor specimen showed classic micro vesicular, solid trabecular, papillary morphology and cystically area at low power magnification (Figure 1). The surgical margins were negative, and it had visible necrosis, with no vascular invasion.

The morphology of the tumor tissues was diverse (Figure 2), and the main microscopic manifestations were as follows: (1) Characteristic Schiller-Duval bodies, which had central capillaries surrounded by a layer of cuboidal or low columnar embryonal epithelioid cells attached to the periphery; (2) Loose reticular structure, composed of a basophilic myxoid matrix, reticular microcysts, with or without a slit-like structure; the capsule wall was covered with flat, pleomorphic, or mesothelioid cells, which often had large and dark nuclei and cysts of varying sizes; (3) Papillary structure, with papillae composed of connective tissue axes and overlying epithelioid cells, with significant cellular and nuclear pleomorphism; the connective tissue had varying degrees of hyalinization; (4) Solid structure, with an area composed of aggregates of small epithelioid polygonal cells with clear cytoplasm, large nuclei, and prominent nucleoli; (5) Mucoïd structure. A few cords and adenoids composed of epithelioid cells arranged in myxoid stroma were observed; and (6) Eosinophilic globules or hyaline bodies.

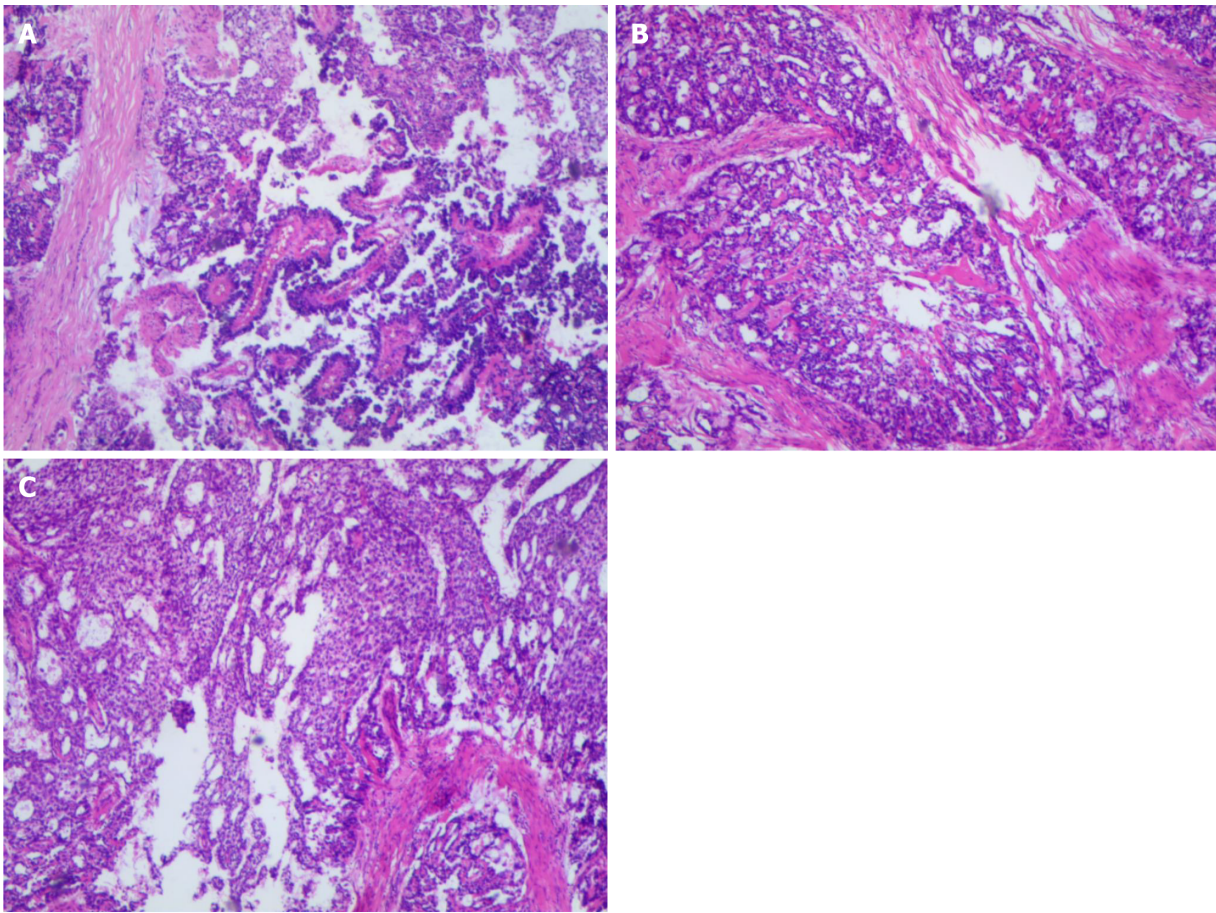
Immunohistochemistry were performed to establish a definitive diagnosis. The detection of the antibodies was accomplished using the streptavidin peroxidase method[17]. Immunostaining showed that broad-spectrum creatine kinase (CK), glypican-3 (GPC3) was strongly positive in almost all neoplastic cells. Spalt-like transcription factor 4 (SALL4) was also strongly positive in the nuclei of the neoplastic cells (Figure 3), while CD117 and epithelial membrane antigen (EMA) were positive in a little range of neoplastic cells [Because the positive range was minimal (around 5%), we have reason to suspect that they were false positives]. The percentage of cells that stained positively for Ki67 was about 40%. The expression of alpha-fetoprotein (AFP), Vimentin, CD31, CD34, D2-40, S100, Octamer-binding transcription factor 3/4 (OCT3/4), Desmin, Smooth Muscle Antibody, Paired box protein 8, Wilms Tumor gene 1, Placental alkaline phosphatase, Estrogen receptor, Progesterone receptor, and human chorionic gonadotrophin were negative in almost all neoplastic cells. Because it was shown in the literature that nearly all YST expressed AFP, we performed the second stain of AFP, and the result was also negative. In addition, we sent the samples to "Cancer Hospital, Chinese Academy of Medical Sciences" for counterstaining, AFP was still negative in expression.

OUTCOME AND FOLLOW-UP

The tumor was then resected without significant bleeding, and the patient recovered well without any signs of recurrence up to now. Remission was achieved after radical surgery, the patient was in good health. At the 3-, 6-, and 9-mo follow-up, the patient did not show local recurrence or distant metastasis, and the serum AFP was within normal levels. The patient will be followed up with clinical examinations and serum AFP level measurements every 3 mo for the first 2 years and every 6 mo thereafter for the next 3 years.

DISCUSSION

Based on the clinical information, histological features, and immunohistochemical staining profile described above, the tumor was diagnosed as a primary YST in the abdominal wall. YST is a common



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Figure 1 Histological findings from the resected tumor specimen showed at low power magnification. A: Histological findings from the resected tumor specimen showed papillary morphology areas at low power magnification; B: Classic micro vesicular and cystically areas at low power magnification; C: A solid trabecular areas at low power magnification. Original magnification $\times 100$ (Hematoxylin & eosin).

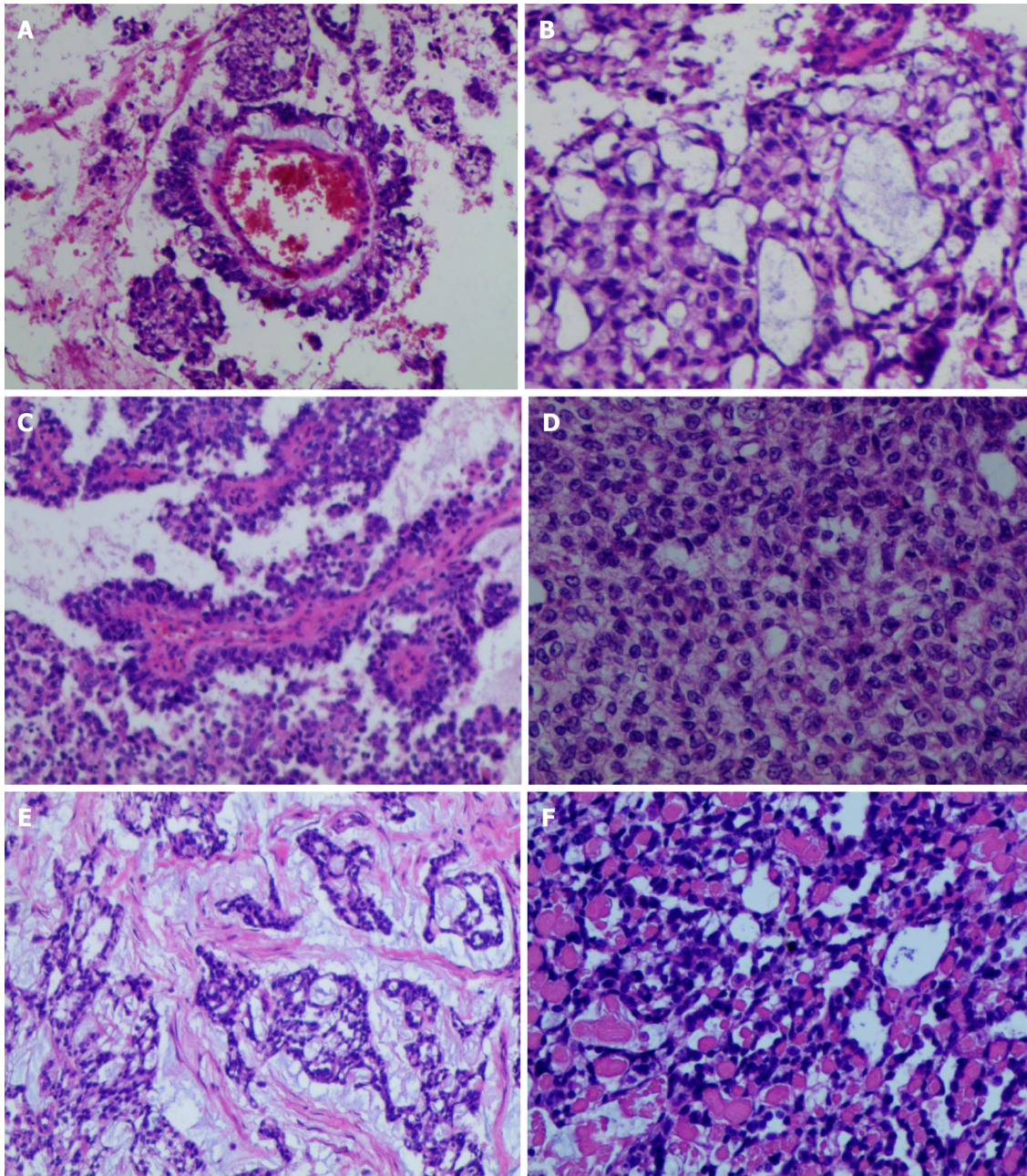
tumor in the gonads, especially in the ovary and testis, and is rarely found in regions other than the gonads, according to a few documented reports. Extragonadal YST may have derived from the malposition of germ cells during embryogenesis. During embryonic development, germ cells migrate from the yolk sac *via* the midline dorsal mesentery to the genital ridge. During migration, some germ cells may remain anywhere along the migration pathway and develop into germ cell tumors[18]. These tumor locations include the liver and sacrococcygeal, gastric, retroperitoneal, and other regions[19-22]. Even fewer cases have been reported of YST occurrence in the abdominal wall. To date, only 2 cases of primary abdominal wall YST have been reported in the medical literature (Table 1)[15,16]. Both reports describe tumors in female infants, while our case occurred in a 20-year-old adult. And the 2 cases all expressed AFP, while in our case AFP was negative in expression. Ultimately, we believe that this was also a special feature of our case that is different from previous YST cases.

Previous reports, including this case, have summarised the features of the primary abdominal wall YST, which often occurs in younger children. Macroscopically, these tumors are typically well-circumscribed, round, or spherical. The cut surface can appear greyish-white, greyish-red, or greyish-yellow. The histological pattern of this tumor is similar to the patterns found in the gonads. In this case, because the mass was located in the lower abdomen, the initial consideration of the clinicians was a sebaceous gland tumor, soft tissue tumor, or metastatic carcinoma. Thus, the patient in this case was not tested for AFP before the surgery. After resection, the patient's serum AFP level was 2.33 ng/mL (normal range: 0–8.04 ng/mL).

To make a correct diagnosis, the metastatic YST from the ovary should first be excluded through detailed clinical examination and evaluation. Moreover, the enhanced CT examination revealed no abnormalities in the abdominal organs and other regions, so this case was considered a primary tumor in the abdominal wall. Other differential diagnoses include embryonal carcinoma, dysgerminoma, clear cell carcinoma, teratoma, and other types of germ cell tumors. These different types of tumors can be distinguished by their respective histological features and immunohistochemical results. Embryonal carcinoma may show a solid structure, and the CK, SALL4, and GPC3 expression can be measured using immunohistochemistry. However in embryonal carcinoma, the cells were more atypical, with large nuclei, prominent nucleoli, and no basement membrane-like material. The immunomarkers CD30

Table 1 The case reports of abdominal wall yolk sac tumor

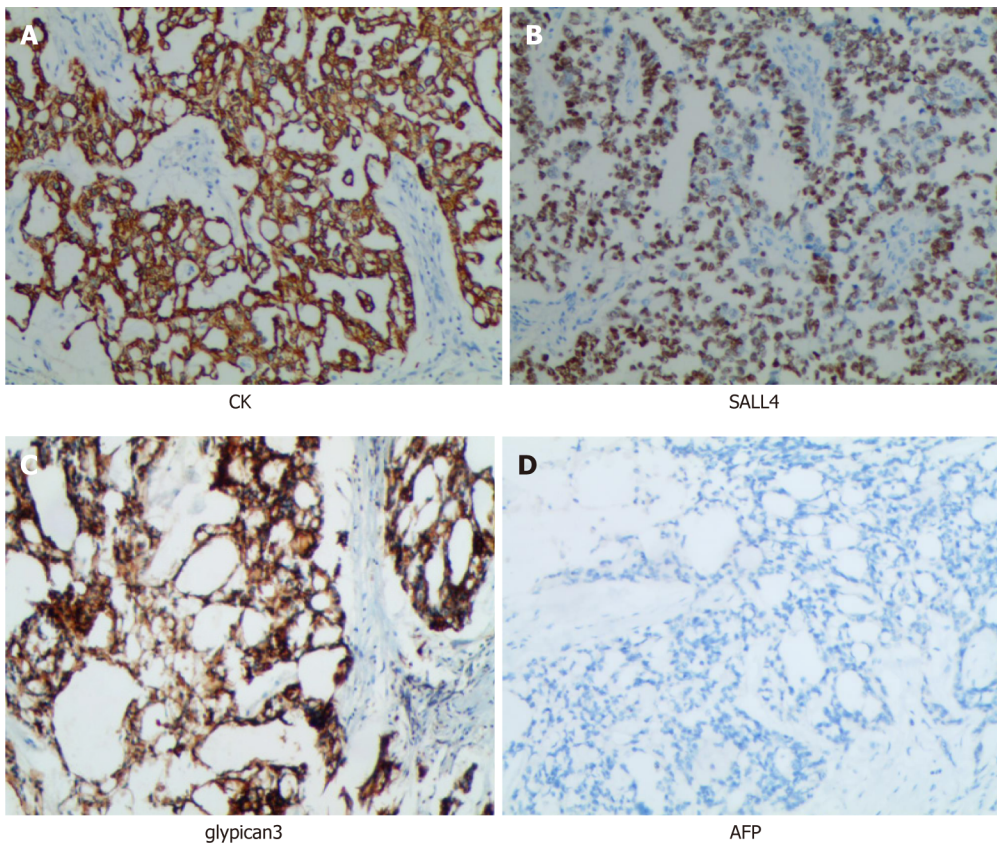
Case	Sex	Age	Location	Size	Clinical presentation	Management	Ref.
1	Female	20-mo	Upper abdominal	4 cm × 4 cm	Abdominal wall mass	Tumor resection	Reddy <i>et al</i> [15], 2022
2	Female	18-mo	Right hypochondriac region	1.5 cm × 1.3 cm × 0.8 cm	Abdominal wall mass	Tumor resection	van den Akker <i>et al</i> [16], 2017



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Figure 2 Histological findings from the resected tumor specimen showed at high power magnification. A: Characteristic Schiller-Duval bodies; B: Loose reticular structure; C: Papillary structure; D: Solid structure; E: Mucoïd structure; F: Eosinophilic globules or hyaline bodies. Original magnification × 200 (Hematoxylin & eosin).

and OCT3/4 were positive in embryonal carcinoma, even though these were rarely observed in YST. Compared with YST tumor cells, dysgerminoma cells were more polygonal, the nucleoli were more obvious, and most were arranged in solid, cord-like, small tubular, and insular patterns. Meanwhile, the YST mostly showed cystic reticular structures with single solid structure. As for the immunohistochemical analysis, D2-40 and OCT4 were positive, and GPC3 and CK were negative in dysgerminoma, while D2-40 and OCT4 were negative, and CK and GPC3 were positive in YST. Clear cell carcinoma had a more regular tubular structure, often accompanied by papillary projections, cells overlying the tubules



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Figure 3 Immunostaining of tumor cells. Immunostaining showed that broad-spectrum creatine kinase and glypican-3 were strongly positive in almost all the neoplastic cells; Spalt-like transcription factor 4 was strongly positive in the nuclei of the neoplastic cells; alpha-fetoprotein was negative in the neoplastic cells. Original magnification $\times 200$ (Immunohistochemical). CK: Creatine kinase; SALL4: Spalt-like transcription factor 4; AFP: Alpha-fetoprotein.

were cuboidal, clear cytoplasm, or boot stud-like cells, and lacked the typical perivascular structure of YST.

Immunohistochemically, clear cell carcinoma was diffusely strong positive for EMA and negative or partially positive for GPC3, while our results showed that very few areas of EMA are positive, while GPC3 was diffusely strong positive. Cystic mature teratomas were generally composed of three well-differentiated germ layers (endoderm, mesoderm, and ectoderm), and tumors were often cystic with a relatively intact capsule. Not consistent with the tumor we saw.

At present, there is no specific tumor marker for YST. The immunostaining of YST often displays the broad-spectrum CK, GPC3, and SALL4 positivity, which can be helpful for its diagnosis. YST has variable morphological features: microcystic/reticular, glandular, and solid structures are the more common patterns, while papillary and hepatoid structures are less common. Mixed histologic patterns were present in two-thirds of the cases, while Schiller-Duval bodies were seen in one-fifth of the cases [6]. As for the solid patterns, the neoplastic cells usually have mostly pale to clear abundant cytoplasm, with frequent intercellular basement membrane deposits, rare microcysts, nuclear pleomorphism, and hyaline globules. Thus, the present case showed a typical structure of YST.

CONCLUSION

Extragenital YSTs are usually a diagnostic challenge, because they are infrequent, but also because a thoughtful and detailed differential diagnostic process must be performed. In this case, since the tumor presented classic YST features, and immunohistochemistry was concordant, the diagnostic process was more straightforward, but not simplistic. And this is the first reported case of a primary YST in the abdominal wall of an adult. As for the prognosis of the disease is generally poor, the authors could follow up the patient closely.

FOOTNOTES

Author contributions: Wang Y contributed to manuscript preparation and published work; Yang J contributed to data acquisition and analysis.

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