

Signet ring cell carcinoma arising from sacrococcygeal teratoma: a case report and review of the literature

Journal of International Medical Research

2019, Vol. 47(5) 2234–2239

© The Author(s) 2019

Article reuse guidelines:

sagepub.com/journals-permissions

DOI: 10.1177/0300060519831574

journals.sagepub.com/home/imr



Pengfei Zhou^{1,#} , Shiju Liu^{2,#}, Huiju Yang²,
Yaxin Jiang², Xiang Liu² and Dianwen Liu²

Abstract

We report here a rare case of adult sacrococcygeal teratoma (SCT) that was pathologically diagnosed as signet ring cell carcinoma (SRCC). A 26-year-old man complained of lower abdominal distension and discomfort and difficulty in urinating, and he was admitted to our hospital. Pelvic magnetic resonance imaging showed multiple oval, solid-cystic masses around the anterior sacrococcygeal region that measured approximately $96 \times 114 \times 89$ mm. Magnetic resonance imaging also showed irregular cysts around the posterior sacrococcygeal region that measured approximately $34 \times 72 \times 60$ mm. The preliminary diagnosis was cystic SCT. The patient then underwent surgery, during which we incised the cysts. A large amount of viscous, jelly-like liquid was present in the anterior sacrococcygeal mass. Furthermore, a large amount of light yellow, porridge-like secretion was present in the posterior sacrococcygeal mass. A pathological examination and immunohistochemistry confirmed teratoma, specifically SRCC.

Keywords

Sacrococcygeal teratoma, cyst, signet ring cell carcinoma, magnetic resonance imaging, mucinous adenocarcinoma, carcinoembryonic antigen, tumor

Date received: 26 September 2018; accepted: 24 January 2019

¹He Nan University of Traditional Chinese Medicine, Henan, P.R. China

²Department of Surgery of Traditional Chinese Medicine, The Third Hospital to He Nan University of Traditional Chinese Medicine, Henan, P.R. China

[#]These authors contributed equally to this work

Corresponding author:

Dianwen Liu, The Third Hospital to He Nan University of Traditional Chinese Medicine, No.63, Dongming Road, Jinshui District, Zhengzhou City, Henan Province, 450008, P.R. China.

Email: henry258369@126.com



Introduction

Primary anterior sacral teratoma mainly occurs in infants and is rare in adults. Most sacrococcygeal teratomas (SCTs) are found at birth and less than 10% of cases are found at the age of ≥ 2 years. The incidence rate of SCT in adults ranges from 1/40,000 to 1/63,000, and women are three to four times more likely than men to develop this disease.^{1,2} Among children, 85% of SCTs are benign. The risk of malignant transformation increases with age.³ Studies have shown that the risk of malignant transformation among adult SCT is 1% to 12%.^{4,5} However, transformation of SCT into signet ring cell carcinoma (SRCC) has not been reported. In the present case, the patient was initially diagnosed with an anterior sacral cyst. However, an intraoperative examination showed that the anterior sacrococcygeal cyst contained a large amount of viscous, oozing jelly-like liquid. The postsurgical pathological examination findings were highly suggestive of a teratoma combined with a mucinous tumor. However, immunohistology confirmed a malignant teratoma, and the malignant component was SRCC.

Case report

We experienced a case of a 26-year-old man with a 40- × 50-mm sacrococcygeal lump that had been present since birth. He had no symptoms and had not received any treatment. One week before the current presentation, he sought treatment for lower abdominal distension and discomfort at our hospital. Pelvic magnetic resonance imaging showed a solid-cystic mass in the pelvic cavity. Anterior sacrococcygeal cysts that measured approximately 96 × 114 × 89 mm were present and reached as high as the first sacrum. Posterior sacrococcygeal cysts were also present and they were irregular and measured approximately



Figure 1. Magnetic resonance imaging shows multiple cystic lumps around the sacrococcygeal region. The anterior sacrococcygeal cysts were oval and reached as high as the first sacrum, and measured approximately 96 × 114 × 89 mm.

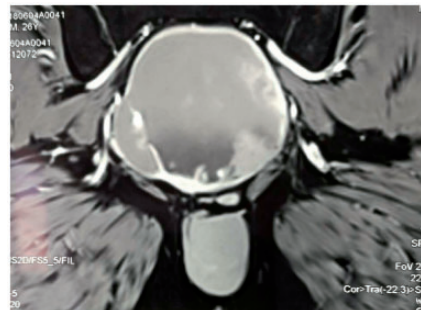


Figure 2. Magnetic resonance imaging shows that the posterior sacrococcygeal cysts are irregular and stratified inside. These cysts measured approximately 34 × 72 × 60 mm.

34 × 72 × 60 mm. The preliminary diagnosis was SCT (Figures 1–3).

The patient was admitted to our hospital and underwent surgery. The surgery started from the sacrococcygeal region, with a curved cut of 10 cm in length and 7 cm

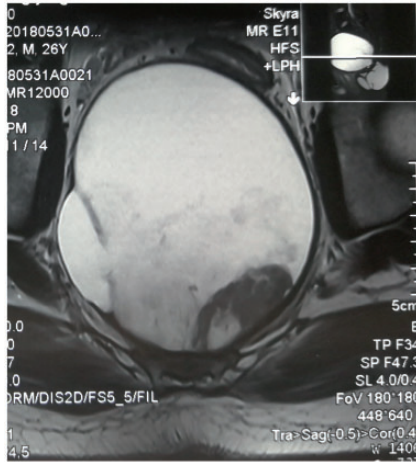


Figure 3. The anterior sacrococcygeal cysts are larger than the posterior cysts, with an intact capsule. The preliminary diagnosis was sacrococcygeal teratoma.

above the anus. The skin and subcutaneous tissue were cut open to show the cystic wall of the retrosacral mass. The mass was completely dissected along the cystic wall, and a large amount of light yellow, porridge-like secretion was present in the sacrococcygeal mass. To separate the larger cyst, which was in front of the sacral tail, the fifth sacral vertebra was removed to further expose the surgical field and complete resection of the cyst. Decompression of the cyst showed a large amount of mucinous jelly-like liquid and the anterior sacral tumor was completely stripped. Because the cyst was closely adhered to the posterior rectal wall, a 1-cm² portion of the posterior rectal wall was removed. Sutures were applied to repair the rectal wall, and sigmoid colostomy was performed.

A postsurgical pathological examination showed a large amount of sticky liquid within the cyst and signet ring cells floating within the liquid. A diagnosis of teratoma combined with a mucinous tumor was considered. The immunohistological results were as follows: AE1/AE3 +, epithelial

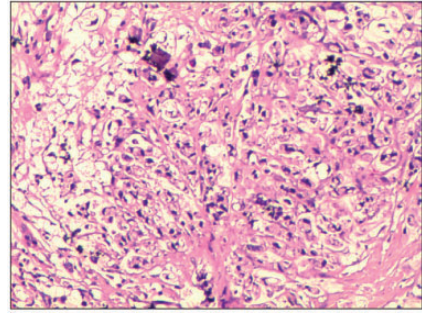


Figure 4. Microscopically, a large amount of mucus can be seen and signet ring cell carcinoma-like cells are floating in mucus. A pathological examination indicated a teratoma combined with a mucinous tumor. Hematoxylin–eosin stain, $\times 100$.

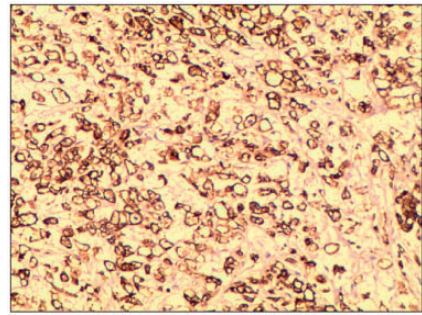


Figure 5. Immunohistochemistry with diaminobenzidine stain ($\times 100$) shows positivity for AE1/AE3, suggesting a malignant teratoma.

membrane antigen +, CDX-2 +, villin +, cytokeratin 7 –, cytokeratin 20 +, synaptophysin –, CD56 –, chromogranin A –, S-100 –, and Ki-67 50%+. These findings were consistent with a malignant teratoma. The malignant cells belonged to SRCC (Figures 4–6). Tumor marker levels were also measured. The CA724 level was 124.784 U/mL and the carcinoembryonic antigen level was 21.53 ng/mL. All of these results suggested high-grade cancer requiring chemotherapy. The patient then underwent chemotherapy.

Written consent was obtained by the patient and his parents for publication of

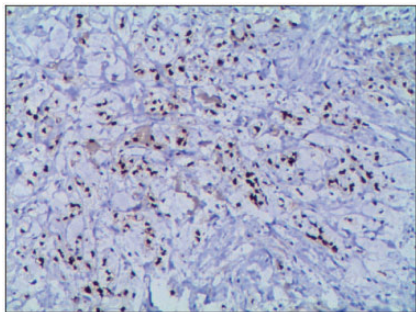


Figure 6. Immunohistochemistry shows that CDX-2 is positive. The malignant component was a signet ring cell carcinoma.

this report and any accompanying images. This case was based on actual routine clinical treatment, and did not involve any ethical human studies. Therefore, we did not need to apply for review by the Ethics Committee or Institutional Review Board.

Discussion

SCT is a congenital embryonic tumor arising from multiple primordial blastoderm cells during human embryonic development. SCT is composed of complex components, such as bone, hair, and teeth.⁶ SCT can be benign or malignant. Altman's classification criteria of neonatal SCT divide this cancer into the following four types.⁷ Type I tumors are exposed to the outside of the fetus. Type II includes tumors that grow outside the fetus, and most are exposed to the body. Types I and II tumors account for 80% of all SCT types and are most common in the neonatal period. Type III tumors are located in the pelvic or abdominal cavity with a small part *in vitro*. Type IV SCT is defined as tumors that are entirely located in the abdominal cavity. Type IV SCT is rare, with an incidence of approximately 10%. The symptoms of type IV SCT appear late, the tumor is found late, and this type has a poor prognosis. Most SCTs in

neonates are benign with a low rate of malignant transformation.⁸ Serum alpha-fetoprotein levels can be used as a reference for detecting malignant transformation of neonatal SCT and to monitor therapeutic effects. Benign solid tumors in neonatal SCT include external, middle, and internal germ layers. Most cystic teratomas contain cystic walls, which are covered by columnar or squamous epithelium. The structure of SCT is disordered and squamous epithelium, and sebaceous glands, and hair follicles are more common in cysts. Striated muscle, smooth muscle, fat, a large amount of fibrous tissue, proliferating blood vessels, multinucleated giant cells, monocytes, lymphocytes, and lymphatic vessels can be seen in the tumor stroma. Neonatal immature teratoma contains a differentiated mature tissue structure, which is often mixed with immature embryonic tissue. This embryonic tissue is most often nerve tissue. According to the maturity level of neonatal immature teratoma, this type is divided into three levels.⁹ The higher the grade, the higher the degree of malignancy. The benign tumor capsule is intact, and most malignant tumors are incomplete. A large amount of immature dermoid tissue, nerve fibers, glandular tissue, cartilage, fat, and lymphatic vessels are present. Multiple germ layers appear, and the tumor cells vary in size and irregular arrangement.⁷

In adults, the benign cyst wall may contain epidermal cells, as well as hair follicles and sweat glands. Within the cyst, fallen epithelial cells, sebaceous glands, keratinized substances, and cholesterol crystals form a white or light yellow porridge-like mixture. The malignant SCT rapidly grows, easily ruptures, invades the surrounding tissue, and may migrate along the blood or lymph circulation. The tumor might be solid or cystic. Microscopic examination may show immature nerve tissues, such as primordial neural tubes, a daisy-like mass structure, and even neuroblasts.

A total of 90% of neonatal SCTs are visible from the patient's body surface. In adults, SCT more frequently manifests as a pelvic cyst without any clinical symptoms and is thus difficult to detect.¹⁰

In the diagnosis of SCT, B-ultrasound can indicate the location of the cyst, but computed tomography and magnetic resonance imaging can show the location of the tumor more clearly. Imaging examinations can also assess characteristics of tumors, including size, morphology, blood vessels, growth rate, and tumor volume. The contents of the cyst usually include different tissues, fats, and fluids. These components can be identified on magnetic resonance-based images on the basis of image signal intensity.¹¹

SCT, regardless of whether it is benign or malignant, requires surgery for complete removal. The tumor type and the corresponding range of invasion determine the surgery protocol and postsurgical management. Benign neoplasms and malignant neoplasms without obvious peripheral invasion can generally be completely excised, while local infiltration of adjacent organs can be excised together with tightly adhered and undetachable organs or tissues. Malignant tumors that have extensively invaded the rectum, sacrum, or major pelvic blood vessels and that have developed distant metastasis require surgical treatment followed by radiotherapy or chemotherapy. Long-term follow-up of the patient's survival status is also required.¹²

SRCC is also called mucinous carcinoma, which is a malignant epithelial tumor, and was first reported by Laufamne and Saphir in 1951.¹³ SRCC is pathologically characterized by rich cytoplasm that is full of mucin; the tumor is filled with signet ring cancer cells and is almost free of gland-like structures.¹⁴ The cytoplasmic mucin displaces the nucleus to the cell periphery, resulting in a signet ring-like cellular morphology. SRCC is a unique subtype of

mucinous adenocarcinoma and more malignant than other mucinous adenocarcinomas. Surrounded by mucins, SRCC becomes insensitive to radiotherapy and chemotherapy, and the prognosis is relatively poor. SRCC often occurs in the gastrointestinal tract, mammary gland, bladder, and prostate.¹⁵ SRCC in the sacrococcygeal region is extremely rare. This adenocarcinoma is characterized by rapid growth and a high level of malignancy, and it rapidly invades the intestinal wall and infiltrates the surrounding organs.

SRCC is a subtype of mucous adenocarcinoma and has unique clinical characteristics. The tumor markers CA724, CA199, and carcinoembryonic antigen are highly recognizable in SRCC, and are thus useful for diagnostic and prognostic evaluation. SRCC has a poor prognosis, and the prognosis is related to the patient's age, sex, tumor stage, and treatment protocol. Low-grade carcinoma without metastasis into the blood and lymph system has a more favorable prognosis.¹⁶

In conclusion, we describe a rare case of adult cystic SCT that was diagnosed as SRCC. Clinically, SRCC has not been previously reported in the sacrococcygeal region. This tumor has a special structure and location that makes definitive diagnosis challenging using imaging and ultrasound technology. The final diagnosis of SRCC depends on a histopathological examination. In the present case, the tumor had a high position, clear margin, and smooth boundary as shown by magnetic resonance imaging. Therefore, the surgery could be started from the sacrococcygeal region. SCT is derived from germ cells (i.e., totipotent cells). These germ cells develop into gonads under normal conditions. However, under certain factors, these cells remain in the sacrum, coccyx, and ovary. Therefore, removal of the coccyx is necessary to prevent recurrence. The patient's serum tumor marker levels and

postsurgical pathological and immunohistological findings suggested potential metastasis. The patient underwent chemotherapy. However, SRCC is highly malignant and insensitive to radiotherapy and chemotherapy. Therefore, the prognosis is guarded.

Author contributions

Pengfei Zhou, Shiju Liu, Huiju Yang, Yaxin Jiang, Xiang Liu, and Dianwen Liu contributed to the diagnosis, acquisition of data, and preparation and drafting of the manuscript. Pengfei Zhou and Shiju Liu were the major contributors for writing the manuscript. Dianwen Liu supervised the present study.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

ORCID iD

Pengfei Zhou  <http://orcid.org/0000-0001-7741-7785>

References

- Batukan C, Ozgun MT and Basbug M. First trimester diagnosis of sacrococcygeal teratoma using two-and three-dimensional ultrasound. *J Clin Ultrasound* 2011; 39: 160–163.
- Szylo K and Lesnik N. Sacrococcygeal teratoma-case report and review of the literature. *Am J Case Rep* 2013; 14: 1–5.
- Mazreku A, Karaj A, Avdia I, et al. The presentation and management of presacral tumors. *Acta Chir Jugosl* 2010; 57: 55–59.
- Zhang F, Yu X, Zeng J, et al. Mucinous tumor arising in a giant sacrococcygeal teratoma: a rare case report. *Medicine (Baltimore)* 2017; 96: e8759.
- Dede M, Gungor S, Yenen MC, et al. CA19-9 may have clinical significance in mature cystic teratomas of the ovary. *Int J Gynecol Cancer* 2006; 16: 189–193.
- Merchea A, Larson DW, Hubner M, et al. The value of preoperative biopsy in the management of solid presacral tumors. *Dis Colon Rectum* 2013; 56: 756–760.
- Padilla BE, Vu L, Lee H, et al. Sacrococcygeal teratoma: late recurrence warrants long-term surveillance. *Pediatr Surg Int* 2017; 33: 1189–1194.
- Göbel U, Schneider DT, Calaminus G, et al. Multimodal treatment of malignant sacrococcygeal germ cell tumors: a prospective analysis of 66 patients of the German cooperative protocols MAKEI 83/86 and 89. *J Clin Oncol* 2001; 19: 1943–1950.
- Sumiyoshi S, Machida J, Yamamoto T, et al. Massive immature teratoma in a neonate. *Int J Oral Maxillofac Surg* 2010; 39: 1020–1023.
- Hobson KG, Ghaemmaghami V, Roe JP, et al. Tumors of the retrorectal space. *Dis Colon Rectum* 2005; 48: 1964–1974.
- Firszt OP, Myga-Porosiło J, Pośpieszny K, et al. Radiological features of sacrococcygeal teratomas in fetal magnetic resonance imaging and computed tomography – a case report. *Pol J Radiol* 2018; 83: e19–e23.
- Graves CE, Idowu O, Zovickian J, et al. Use of intraoperative lateral pelvic X-ray to localize and ensure coccyx removal during sacrococcygeal teratoma resection. *Pediatr Surg Int* 2017; 33: 389–392.
- Laufman H, Saphir O. Primary linitis plastica type of carcinoma of the colon. *AMA Arch Surg* 1951; 62: 79–91.
- Kang H, O'Connell JB, Maggard MA, et al. A 10-year outcomes evaluation of mucinous and signet-ring cell carcinoma of the colon and rectum. *Dis Colon Rectum* 2005; 48: 1161–1168.
- Kitajima K, Fujimori T, Fujii S, et al. Correlations between lymph node metastasis and depth of submucosal invasion in submucosal invasive colorectal carcinoma: a Japanese collaborative study. *J Gastroenterol* 2004; 39: 534–543.
- Arifi S, Elmesbahi O and Amarti Riffi A. Primary signet ring cell carcinoma of the colon and rectum. *Bull Cancer* 2015; 102: 880–888.