

Case Report

Sacrococcygeal teratoma with nephroblastic elements: a case report and review of literature

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Abstract: Sacrococcygeal teratoma with nephroblastic elements is very rare. Only 8 cases have been reported up to date. 3 cases were misdiagnosed as extrarenal nephroblastomas and were administered excessive treatments such as chemotherapy and radiation therapy. It has a great significance to distinguish sacrococcygeal teratoma with nephroblastic elements from extrarenal nephroblastoma for their different biological behaviors, therapies and prognoses. Here we report one case of sacrococcygeal teratoma with nephroblastic elements in a newborn with review of the literature. This is the first case of sacrococcygeal teratoma with nephroblastic elements in Chinese to our knowledge.

Keywords: Sacrococcygeal teratoma, nephroblastic elements, extrarenal nephroblastoma

Introduction

Sacrococcygeal teratoma is one of the most common neoplasms in newborns with an incidence of 1/21,000 births [1]. Although most sacrococcygeal teratomas are mature, a variety of immature components may be present. Occurrence of nephroblastic elements is very rare in sacrococcygeal teratomas. Only 8 cases of sacrococcygeal teratomas with nephroblastic elements (STWNE) have been reported up to now. Here we describe another case of sacrococcygeal teratoma with nephroblastic elements and review the literature.

Case report

A 3700 g boy was delivered by caesarean section at full-term gestation. He was admitted to our hospital at birth for a sacrococcygeal mass. The mass was first detected in the fetus at 24 weeks' gestation by ultrasound. An exogenous mass measuring 10 × 8 × 5 cm in the sacrococcygeal region was found on physical examination which was confirmed by CT scan and MRI scan afterwards (**Figure 1**). Both of the kidneys were normal. The serum α -fetoprotein (AFP) concentration was more than 121000 ng/ml at birth. Given that the mass might be a teratoma,

a tumorectomy was performed on the 8th day after birth. The mass was entirely removed. No additional treatments such as chemotherapy and radiation therapy were given postoperatively. The child is still alive without evidence of tumor recurrence and metastasis 8 months after surgery. Serum AFP levels were monitored in follow-up and fell to 45.1 ng/ml at 8 months after surgery. Written informed consent was got from the child's parents.

Pathological findings

Macroscopically, the specimen was a gray multinodular mass with an intact capsule and a smooth external surface measuring 9 × 8 × 5 cm (**Figure 2**). The cut surface exhibited a gray-tan to light yellow appearance with multicystic and solid components (**Figure 2**). Cystic region contained yellowish clear fluid, and osteocartilaginous tissue was found in the solid area. A lymph node around the tumor measuring about 0.5 cm in diameter was also sent for pathological examination at the same time.

Microscopically, the solid components were composed of a mixture of mature tissues including stratified squamous epithelium (**Figure 3A**), skin appendages (**Figure 3A**), gastrointestinal



Figure 1. Magnetic resonance imaging scan showed a mass in the sacroccocygeal area (arrow).

epithelia (**Figure 3B**), fibrous tissue, vascular tissue, striated muscle tissue, smooth muscle tissue, adipose tissue, peripheral nervous tissue, cartilage, bone, adrenal tissue (**Figure 3C**), pancreatic tissue (**Figure 3D**), and glial tissue. All the mature tissues displayed an organoid arrangement as the normal skin tissue, intestinal wall, pancreatic tissue, adrenal tissue, and so on. There were focal areas of nephroblastic elements under the gastrointestinal epithelia in the form of primitive round cells, immature glomerular structures and tubules (**Figure 3E** and **3F**). The nephroblastic elements were 0.2×0.2 cm comprising approximately 2.5 permillage of the tumor and mitoses could be occasionally found in the areas. A few myxomatous mesenchymal cells surrounded the nephroblastic components. A few foci of the nephroblastic elements resembled the Schiller-Duval bodies in yolk sac tumor (**Figure 3G**). The wall of the cystic area in the tumor was made up of fibrous tissue, fat tissue, and smooth muscle tissue. Immature neuroepithelial components were not observed in the tumor.

Immunohistochemically, the squamous epithelium, the exocrine pancreatic tissue and the adrenal cortical tissue were positive for pan-cytokeratin (AE1/AE3). The glial tissue was positive for glial fibrillary acidic protein. Pancreatic islets and peripheral nervous tissue strongly expressed chromogranin A (CgA) and neuron-specific enolase (NSE). Smooth muscular tissues were positive for α -smooth muscle actin

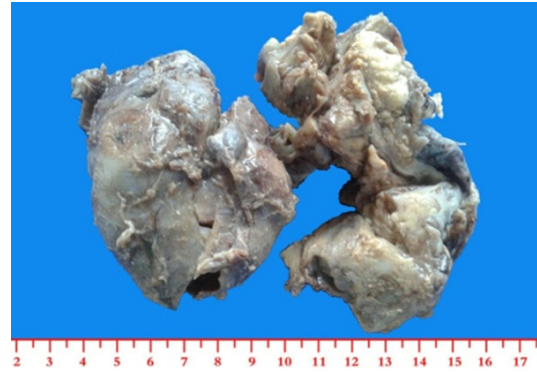


Figure 2. Gross photo of sacroccocygeal teratoma with nephroblastic elements. The mass was gray and multinodular with an intact capsule and a smooth external surface and the cut surface had a gray-tan to light yellow appearance with multicystic and solid components.

(α -SMA). A diffuse and strong nuclear expression for WT1 was observed in the blastemal cells and the immature glomerular and tubular tissues (**Figure 4A**). Tubular structures showed positivity to pan-cytokeratin (AE1/AE3) (**Figure 4B**) and the glomerular and tubular tissue was positive for CD10 (**Figure 4C**). The proliferation index Ki-67 was estimated at 25% in the nephroblastic components compared with 7% in the mature tissues suggesting the nephroblastic components may have a relatively higher proliferative activity (**Figure 4D**). The diagnosis was immature solid-cystic teratoma grade I with nephroblastic elements and no tumor cell was detected in the lymph node.

Discussion

Teratomas are the most common germ cell tumor that consists of tissues derived from the three germ cell layers (ectoderm, mesoderm, and endoderm). The common sites of teratomas are the sacroccocygeal, mediastinal, retroperitoneal and the gonadal organs, etc. Teratomas are classified as mature or immature according to the histological composition and degree of maturation or differentiation. The majority of teratomas are mature and 20%-25% cases are immature [2]. Immature elements usually contain neuroepithelium, cartilage, and primitive mesenchymal tissue. Nephroblastic elements are very rare in the immature elements of teratomas and the detailed occurrence rate of teratomas with nephroblastic elements (TWNE) is unclear. Berry et al reported 1 case of TWNE in his

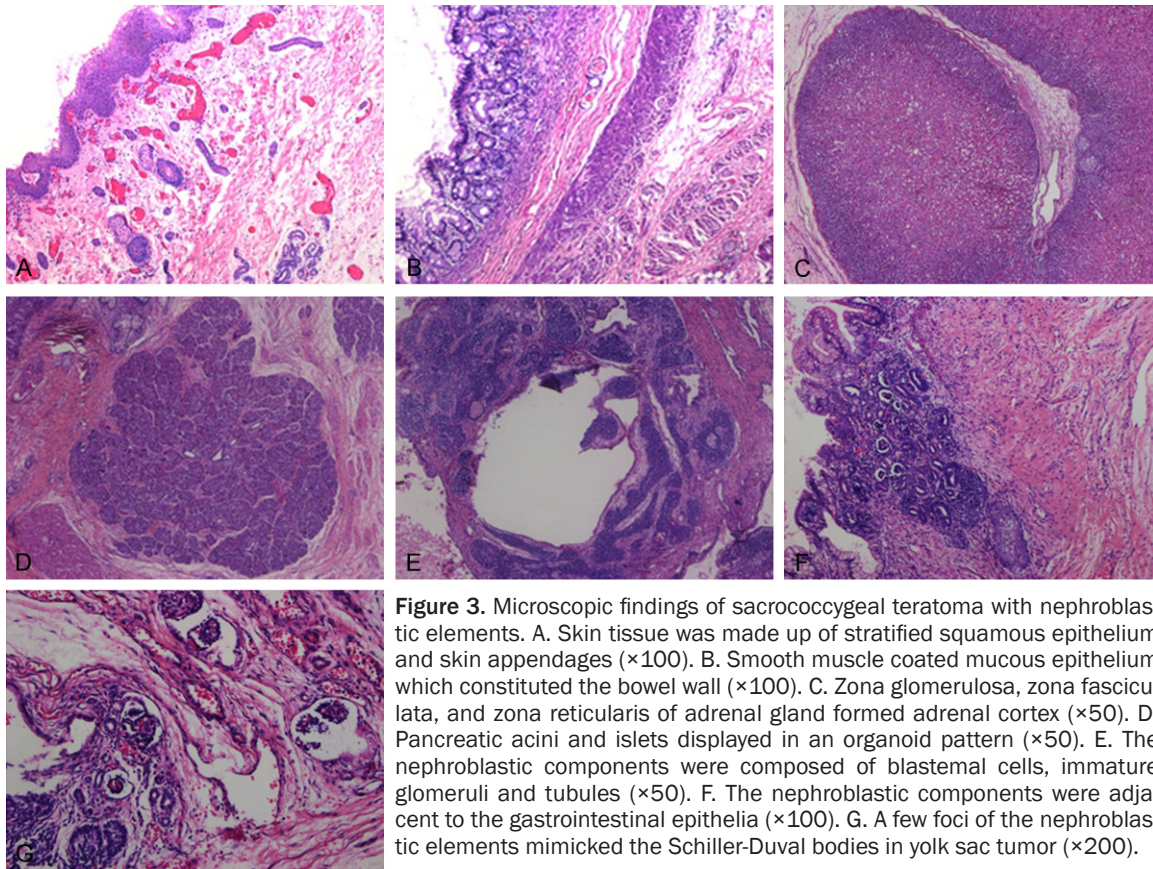


Figure 3. Microscopic findings of sacrococcygeal teratoma with nephroblastic elements. A. Skin tissue was made up of stratified squamous epithelium and skin appendages ($\times 100$). B. Smooth muscle coated mucous epithelium which constituted the bowel wall ($\times 100$). C. Zona glomerulosa, zona fasciculata, and zona reticularis of adrenal gland formed adrenal cortex ($\times 50$). D. Pancreatic acini and islets displayed in an organoid pattern ($\times 50$). E. The nephroblastic components were composed of blastemal cells, immature glomeruli and tubules ($\times 50$). F. The nephroblastic components were adjacent to the gastrointestinal epithelia ($\times 100$). G. A few foci of the nephroblastic elements mimicked the Schiller-Duval bodies in yolk sac tumor ($\times 200$).

series of 91 cases of teratomas [3]. However, Ronald et al detected 2 cases of TWNE in 68 cases of sacrococcygeal teratomas [4]. Only 33 cases of TWNE were reported up to date including our case [5, 6]. 9 of the 33 cases were STWNE [3, 4, 7-11]. Clinicopathologic data of all the 9 cases were collected in **Table 1** though some data were not integrated in the literature. 7 cases were female and our case was a boy though the sex of another case was unavailable. They showed a prominent female predominance which is a typical sex distribution for sacrococcygeal teratomas. They were all young children. Seven were congenital and the other two were unknown. The most common presentations were sacrococcygeal masses. The available maximum diameters of the tumors were 2.0-9.0 cm. All the tumors were composed of differentiated tissues of multidermal origin which arranged in an organoid pattern resembling the normal human organs. The mature tissues contained skin tissue, fat, fibrous tissue, cartilage, respiratory epithelium, gastrointestinal epithelium, neuroglial tissue, pancreatic tissue, and so on. Different proportions of nephro-

blastic elements such as blastemal cells and primitive glomeruli and tubules could be found in the tumors and they were blended with the teratomatous components. 2 of the 9 cases had prevalent nephroblastic components. The nephroblastic components were adjacent to the gastrointestinal epithelia in 3 cases suggesting that they might have some correlations with each other in histogenesis. The nephroblastic components were surrounded by myxomatous mesenchyma in 6 cases. In the nephroblastic elements abundant mitoses were observed in 2 cases and WT-1 nuclear positivity were shown in another 2 cases. Interestingly, immature neuroectodermal tissues were not encountered in the 7 cases and were unclear in the left 2 cases. Ultrastructurally, the immature glomerular structures and tubules were demonstrated to have a differentiation of glomeruli and renal tubules and there was a transition from the myxomatous mesenchymal cells to the blastemal cells [7].

Our case shared the same features with the previous cases: young children, the same posi-

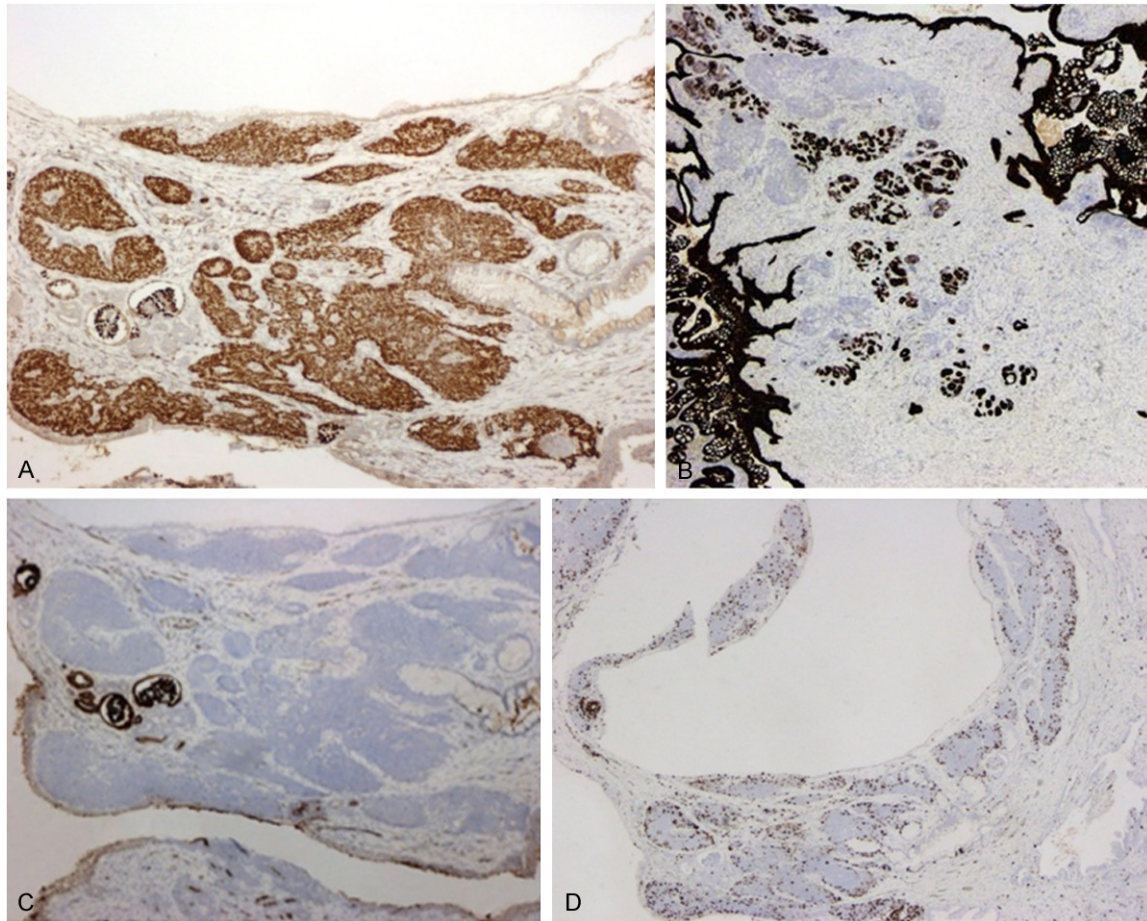


Figure 4. Immunohistochemical features of the nephroblastic elements in sacrococcygeal teratoma. A. WT-1 was diffusely positive in the nephroblastic components ($\times 100$). B. The tubules were positive for pancytokeratin ($\times 50$). C. The immature glomerular structures and tubules were strongly positive for CD10 ($\times 100$). D. Ki-67 index was about 25% in the nephroblastic components ($\times 50$).

tion, mature tissues arising from multiple germ layers in an organoid pattern, and nephroblastic elements. The diagnosis of immature teratoma with nephroblastic elements was optimal. However, some differences could be found between our case and the precedent cases. Our patient was the known only one boy and the amount of nephroblastic elements was so small. A relatively high Ki-67 index and WT-1 positive immunostaining in the nephroblastic elements were detected in our case which had been rarely reported in the previous cases.

The differential diagnosis should be made with extrarenal nephroblastoma and teratoma with yolk sac tumor in the sacrococcygeal area. Extrarenal nephroblastoma was extremely rare. Extrarenal nephroblastoma in the sacrococcygeal area, especially extrarenal teratoid nephro-

blastoma in the sacrococcygeal area, might also include several differentiated tissues, blastemal cells and primitive glomeruli and tubules sometimes and there were some overlaps in morphology between extrarenal nephroblastoma in the sacrococcygeal area and STWNE. 3 cases of STWNE were misdiagnosed as extrarenal nephroblastoma in the total 9 cases and they received excessive treatments such as chemotherapy and radiation therapy. It makes great senses to differentiate these two entities for their different biological behaviors, therapies and prognoses. Teratomatous components of STWNE usually have many varieties of mature tissues and arrange in an organoid pattern which are very rare in extrarenal nephroblastoma in the sacrococcygeal area. Nephroblastic components in teratomas was supposed to be of embryonic origin, however,

Sacrococcygeal teratoma with nephroblastic elements

Table 1. Clinicopathologic features of sacrococcygeal teratomas with nephroblastic elements

Case	Sex	Age at initial discovery	Chief complaint	Age at surgery	Maximum tumor diameter (cm)	Treatment	Follow-up
1 Berry et al [3]	F	birth	Sacrococcygeal mass	2 wo	UA	Operation	Local recurrence and lung metastases 1 y later, lost to follow-up
2 Ward et al [7]	F	birth	Sacrococcygeal mass	3 yo	8.0	Operation, chemotherapy and radiation	AWRM 12 m after surgery
3 Tebbi et al [8]	F	birth	Sacrococcygeal mass	3 yo	8.0	Operation, chemotherapy and radiation	AWRM 46 m after surgery
4 Valdiserri et al [4]	F	birth	Sacrococcygeal mass	3 do	9.0	Operation	AWRM 6 y after surgery
5 Valdiserri et al [4]	F	UA	Sacrococcygeal mass	1.5 yo	2.0	Operation	Died of other diseases 3 y after surgery
6 Andrews et al [9]	F	UA	Sacrococcygeal mass	1 wo-4.3 yo, the detail UA	UA	Operation, chemotherapy and radiation	AWRM 46 m after surgery
7 Heerema-McKenney et al [10]	NA	birth	Sacrococcygeal mass	less than 1 wo	3.8	Operation	AWRM 25 m after surgery
8 Al-Adnani et al [11]	F	birth	Sacrococcygeal mass	15 mo	9.0	Operation	AWRM 9 m after surgery
9 our case	M	24 weeks' gestation	Sacrococcygeal mass	8 do	9.0	Operation	AWRM 8 m after surgery

UA, unavailable; do, days old; wo, weeks old; mo, months old; yo, years old; y, years; m, months; cm, centimeters; M, male; F, female; AWRM, alive without recurrences and metastases.

nephroblastoma was thought to be of mesonephric origin. Emerson et al reported the nephroblastic components shared a clonal origin with the teratoma in an interesting case of mature and immature teratoma with nephroblastic components of the testis [12]. Yolk sac tumor can be found with sacrococcygeal teratoma occasionally. Schiller-Duval bodies in yolk sac tumor mimicked the immature glomeruli. Serum AFP concentration is elevated in sacrococcygeal teratoma with yolk sac tumor and positive immunoreactivity for AFP and negative immunoreactivity for WT-1 would also been shown in the tumor which is different from STWNE.

The standard therapy has not been established for STWNE for their scarcity. Whether fetal STWNE need surgical interventions in utero or not are still unknown. STWNE generally behave indolently and metastases are very rare. Complete resection and close follow-up are optimal. 6 of the 9 STWNE cases received surgery alone and the other 3 cases were given comprehensive therapy including surgery, chemotherapy and radiation therapy because they were misdiagnosed as extrarenal nephroblastomas. 1 case died of other diseases 3 years after surgery, 1 recurred locally with lung metastases 1 year after surgery and was lost to follow-up, the left were all uneventful in the reported follow-up time. All the data suggest the prognoses of STWNE are good.

In conclusion, we described a case of STWNE. This is the first case in Chinese. More data are needed to understand the biology and clinical behavior of this tumor.

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Disclosure of conflict of interest

None.

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