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Case Report/Olgu sunumu

Histopathological Classification of Neonatal Sacrococcygeal Teratomas: Case Report

Neonatal Sakrokoksigeal Teratomların Histopatolojik Sınıflaması: Olgu Sunumu

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Abstract

Sacrococcygeal teratomas are tumours that arise from the sacrococcygeal region containing tissue from all three germ layers. Sacrococcygeal teratoma (SCT) is a relatively rare tumour affecting new-borns, infants and children with an incidence of one per 40,000 live births with malignant transformation with increasing age. It is four times more common in women than men. With this case study, it was aimed to evaluate the histopathological evaluation and grading of SCT. Examination of pathological specimens including immature and malignant transformation should be done carefully.

Keywords: Sacrococcygeal teratoma, congenital, histopatology, mature, immature

Öz

Sakrokoksigeal teratomlar, her üç germ tabakasından doku içeren sakrokoksigeal bölgeden ortaya çıkan tümörlerdir. Sakrokoksigeal teratom (SKT), artan yaşla birlikte malign transformasyonu olan 40.000 canlı doğumda bir insidansı olan yenidoğanları, bebekleri ve çocukları etkileyen nispeten nadir bir tümördür. Kadınlarda erkeklerden dört kat daha sık görülür. Bu olgu çalışması ile birlikte SKT histopatolojik değerlendirme ve derecelenmesi değerlendirmek amaçlanmıştır. İmmatür ve malign transformasyon içeren patolojik spesmen incelemeleri dikkatlice yapılmalıdır.

Anahtar Kelimeler: Sakrokoksigeal teratom, konjenital, histopatoloji, mature, immature

INTRODUCTION

Teratoma consists of the embryological origin of germ cell tumours. Its name is derived from the Greek word "teratos", which literally means "monsters". The "oma" added to the end is a neoplasm marker.[1] Sacrococcygeal teratoma (SCT) is the most common solid tumour in the neonatal period and its incidence has been reported as one in 35,000-40,000 live births.[2] The first description of teratoma cases was BC. The first description of teratoma cases was BC. It dates back to 2000 BC.[3] Teratomas are germ cell tumours that consist of tissue elements foreign to the organ or anatomic region.[4] In 1863, the word Teratoma was used by Virchow.[5] The most appropriate and new definition for SCT isan encapsulated tumour with tissue or organ components in which three primordial germ layers ofectoderm, mesoderm and endodermcan be traced.[4] Teratomas are mostly seen in the sacrococcygeal, mediastinal, retroperitoneal and gonads in the childhood age group. SKTs are divided into three classes as mature, immature and malignant. In studies, the frequency of mature teratoma is around 80%. Mature teratomas mainly consist of different tissues. With this case report, it is aimed to discuss the histopathological evaluation and classification of SCT accompanied by the rare SCT case in a patient who underwent surgical treatment in the neonatal period.

CASE

The patient was delivered by caesarean at the age of 40 weeks as a third living from the 4th pregnancy of a 36-year-old mother with a weight of 3800 g. No problem of the mother was detected during pregnancy follow-up in our hospital.

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No sacral mass was detected in the detailed ultrasonography examination performed on the 21st week of pregnancy and the ultrasound controls performed during pregnancy. When our patient was born, the head circumference was measured to be 35 cm (50-75 percentile), height 51 cm (50-75 percentile) and 3800 g (50-75 percentile). The first minute apgar score after birth was evaluated as 9, and the 5th minute apgar score as 10. The baby, who was found to have swelling in the sacral area during a postpartum physical examination, was consulted to the Paediatric Surgery Clinic (**Figure 1**). In the first physical examination, a 5x5 cm soft mass with a palpable bone and cystic tissue was palpated, starting from the right gluteal region to the sacral region. Superficial tissue ultrasonography was performed for differential diagnosis of the mass detected in the patient. Superficial tissue ultrasonography performed on the patient was reported as "fluid collection with as size of 19x7 mm having homogeneous structure, dense in content, smooth contoured starting from the level of the coccyx and extending under the skin was observed, a second area with a size of approximately 22x8 mm having similar characteristics was observed in the right paravertebral area" Tumour markers (AFP, B-HCG, LDH) were requested from the patient for the differential diagnosis of the mass. Considering that the patient's sacro-gluteal mass might be SCT, a computed tomography examination was requested to evaluate the intraabdominal extension. Computed tomography report was reported as "Cystic image of 29 mm width was observed in the distal of the right gluteal shadow" (**Figure 2**). AFP value of tumour markers examined in the patient preoperatively were measured as 44756 ng/ml, B-HCG 6.5 mIU/ml, and LDH 726 U/L. As sacrococcygeal teratoma was considered in the patient, it was decided to remove the mass completely. A 7x6 cm diameter cystic mass and coccyx were totally removed by making a chavron incision made around the gluteal mass. The patient was followed up in the neonatal intensive care unit after surgery. Oral feeding was started on the first postoperative day. The patient was discharged on the 7th postoperative day with the recommendation for the control of the paediatric surgery outpatient clinic with stable vital signs, full oral nutrition, stool discharge, clean incision and no problem. The AFP control result obtained on the postoperative 6th day of the patient was 15611 ng/ml. Pathology result was reported as "Sacrococcygeal teratoma with immature neuronal component in microcopic foci showinghigh proliferative activity with 67 in teratoma, accompanied by coccyx containing cartilage and bone marrow" (**Figure 3**, **Figure 4a-f**). The postoperative follow-up and treatment of the patient is maintained with paediatric oncology.

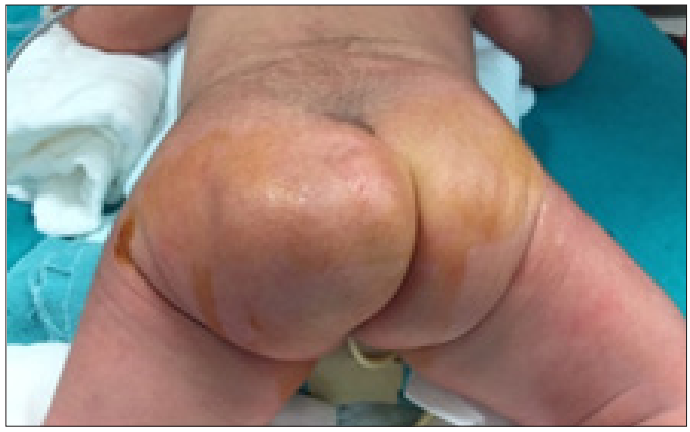

Figure 1. Neonatal Sacrococcygeal teratoma Gluteal Mass Image

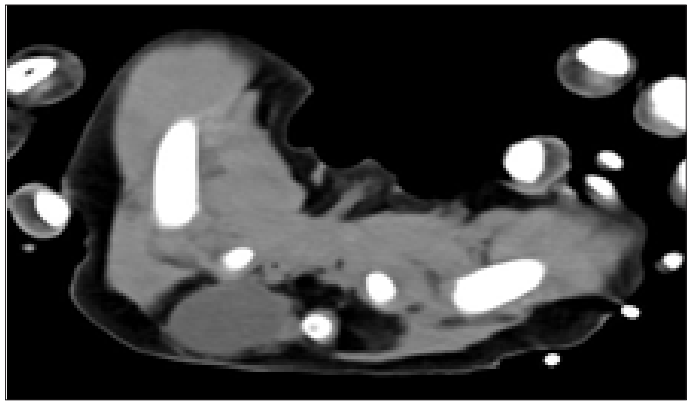

Figure 2. Sacrococcygeal teratoma Computed Tomography Image

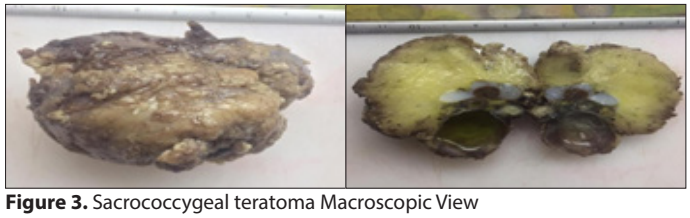

Figure 3. Sacrococcygeal teratoma Macroscopic View

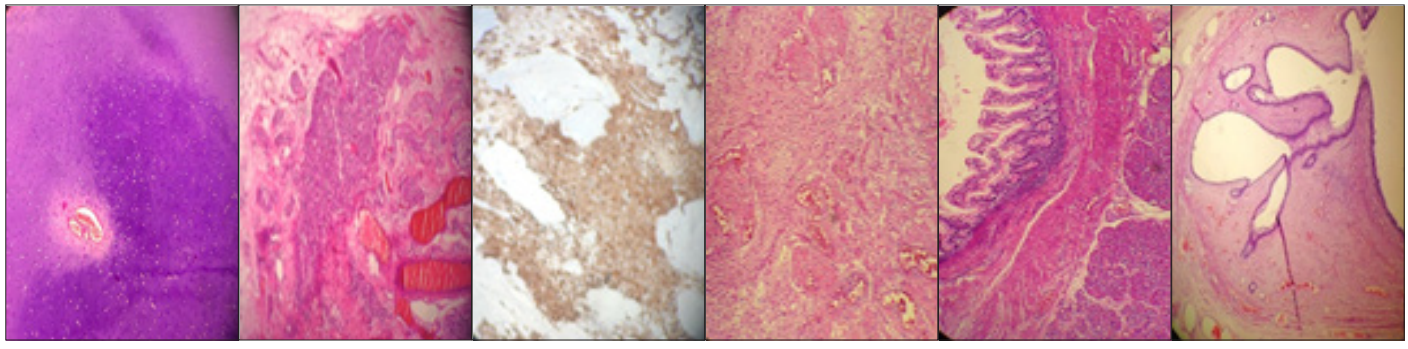

Figure 4. Sacrococcygeal teratoma Histopathological evaluation findings. 4a: Cartilage Tissue, 4b: Peripheral nerve tissue, 4c: Neuronal Component Glial Tissue, 4d: Gastrointestinal mucosa Epithelium, 4e: Glial Tissue Ki-67, 4f: Squamous epithelium tissue

DISCUSSION

Sacrococcygeal teratoma is a relatively rare tumour in the neonatal period. When SCT is present at birth, excisional surgery should be performed as early as possible to prevent malignant transformation that may occur at later ages.[12] The surgical approach in SCTs is total removal of the tumour with the coccyx by using a sacral or sacro-abdominal approach.[13] Most of the SCTs appear at birth as a mass in the sacral region. While most new-borns do not have any other accompanying symptoms, some new-borns may need intensive care due to premature birth, high heart failure and tumour rupture or bleeding within the tumour. Infants with mass lesions in the pelvis may have difficulty in urinating, constipation and signs of an abdominal mass.[14] Although SCTs contain tissues of ectoderm, endoderm and mesoderm origin, their embryological origin is still unclear.[6,7] It is believed that the totipotent cells of the Hensen's node or the remnants of the primitive streak in the coccygeal region emerge in early pregnancy.[15-17] This primitive streak appears as a linear thickening of the ectoderm on the caudal edge of the bilaminar embryonic disc, and subsequently disappears with degenerative changes.[6,7] As the mesoderm reproduces rapidly, the primitive line extends more caudally. The remains of the Hensen's node descend to the coccyx end or anterior surface.[15,17] The growth of these primitive pluripotential cells escapes from the control of embryonic inducers and organizers and results in teratoma as a result of transformation of foreign cells in the anatomical region.[18] Therefore, SCT often occurs near the coccyx, where the highest concentration of primitive cells is present for a longer period of time.[17] SCT consists of multiple neoplastic tissues that are foreign to the sacrococcygeal region and do not contain a specific tissue derived from multiple germ layers.[17] SCT is more common in girls and infants under 3 months.[8-11] According to the Altmann classification, Type 1 and Type 2 SCT (87%) are the most common. Type 3 and Type 4 are extremely rare (**Table 1**). Type 4 SCTs are generally diagnosed at advanced ages.[8] While mixed type (80%) with predominant cystic areas is seen in SCTs, immature solid type can be seen in 20%.[11] Histologically, 80% of SCTs are mature while 20% are immature (**Table 2**). In our case, there are the dominant components of the mature teratoma of ectodermal and endodermal origin, followed by mesodermal, neuroectodermal and organoid endodermal elements. While the ectodermal component usually contains skin and hair follicles and their derivatives, gastrointestinal epithelium, urothelium, and respectively respiratory epithelium are seen most frequently as the endodermal component. The mesodermal component consists of fat, connective tissue, cartilage, bone and muscle tissue with lymphoid tissue. The organoid endodermal component, on the other hand, forms mainly parotid and saliva and pancreatic tissue. Neuroepithelium is seen in all mature teratomas. Immature non-malignant neuroepithelium is seen in immature teratomas. In immature teratomas, rosette formations of primitive tissue are seen. Differential diagnosis of SCTs includes coccygeal meningocele in fetus, myelomeningocele and fetus in fetus.[12,18]

Tablo 1. SCT Altman Classification

Altman classification		Frequency of prevalence
I	The mass is completely outside the pelvis	57.6%
II	Part of the mass is inside the pelvis, most of it outside the pelvis	38.6%
III	Most of the mass is inside the pelvis, some outside the pelvis	3.8%
IV	The mass is completely in the pelvis	Rare

Table 2. Histological diagnosis and tumor grade classification in Sacrococcygeal teratoma

SCT Tumor Maturation	SCT Tumor Grade
Mature Teratoma	Grade 0: Tumor consists only of mature mature tissue
Immature Teratoma	Grade 1: There is a rare immature area in the tumor Grade 2: There are immature areas in the tumor Grade 3: Most of the tumor consists of immature areas

CONCLUSION

Sacrococcygeal teratoma is a relatively rare tumour in the neonatal period. When SCT is detected during delivery, total surgical excision is required as soon as possible to prevent malignant transformation. Histological evaluation is the gold standard in the differential diagnosis of mature or immature SCT.

ETHICAL DECLARATIONS

Informed Consent: Written informed consent was obtained from all participants who participated in this study.

Referee Evaluation Process: Externally peer-reviewed.

Conflict of Interest Statement: The authors have no conflicts of interest to declare.

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Author Contributions: All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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