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REVIEW
ΑΝΑΣΚΟΠΗΣΗ

Management of sacrococcygeal teratoma and imminent complications

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2025, 42(6):727–731

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Διαχείριση ιεροκοκκυγικού τερατώματος και επικείμενες επιπλοκές

Περίληψη στο τέλος του άρθρου

Sacrococcygeal teratomas (SCT) are the most common congenital tumors in fetuses and neonates; approximately 1/35,000 to 1/40,000 live births are reported, and they are extremely rare in adults, with a female-to-male ratio of 3:1. Despite the scarcity of SCT incidents, their management seems to be primarily important so that a dignified quality of life is provided. Surgical resection is the preferred treatment opinion for SCT and can determine the pathological type. There are different approaches, according to the resection: transabdominal, transsacral, and transabdominal-sacral. Whether the coccyx should be removed during surgery is controversial. As a result of the coccyx removal, abnormal bladder function is observed in contrast with the sexual function, which is not affected to both males and females. However, the risk of recurrence still exists and the risk of malignancy transformation as well.

Key words

Malignant
Neonatal
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1. INTRODUCTION

Sacrococcygeal teratomas (SCT) are known as rare tumors, but they are the most common congenital tumors in fetuses and neonates, reported in approximately 1/35,000 to 1/40,000 live births,[1–3] whilst adult SCT are extremely rare, with an incidence of 1 in 40,000 to 1 in 63,000 patients and a female to male ratio of 3:1.[4] A teratoma is a common germ cell tumor that is mainly composed of totipotent stem cells.[5] SCTs were first described by Virchow in 1869 and, because of their complexity, were called "monstrous tumors".[6] SCTs are usually large enough to have a significant mass effect on adjacent structures, and the mass effect begins during fetal development. That is the reason why SCTs are generally diagnosed at birth or in infancy.[2] It has also been reported that fetal mortality is higher if the gestational week at the time of diagnosis is early in SCT.[7] SCTs diagnosed postnatally have been associated with an excellent prognosis after surgical excision;[8] fetuses with a prenatally diagnosed SCT still have a high risk of death even if the prenatal diagnosis may have contributed to the improvement of outcome, though.[9] The main reason for the poor prognosis in fetal SCT is a high-output cardiac failure caused by increased blood flow, according to the amount of solid components present in the tumor, and rupture of the tumor during delivery with a massive hemorrhage. Generally, the vast majority of SCT cases are sporadic and rare in twin pregnancy,[10] whilst familial presacral teratomas are inherited in an autosomal dominant pattern.[11]

2. ETIOLOGY AND STRUCTURE

The etiology of teratomas remains unknown. Teratomas are thought to originate from totipotent stem cells, which are usually present in the ovaries and testes and are sometimes abnormally presented in isolated midline embryos. In rare cases, teratomas may be solid tumors composed of a variety of heterogeneous tissues and organizational structures that appear benign. These structures were derived from all three germ cells.[5] In particular, SCTs are thought to arise from the migration of totipotent stem cells to the coccygeal region.[12]

Typical mature teratomas are mostly cystic and fatty, with few parenchyma.[13] SCTs contain varying proportions of soft tissue, fat, calcification, and fluid components and often have no capsule or pseudocapsule, unlike teratomas at other sites.[5] Giant teratomas such as SCTs compress the pelvic cavity and pelvic floor structure, occupy most of the pelvic cavity, and protrude to the tail or the abdominal cavity and tail, compressing the rectum and uterus to move forward, and the anus to move forward and downward. In addition, by definition, teratomas are composed of more than one germ cell layer (ectoderm, mesoderm, and endoderm) and so SCTs contain a variety of tissues of more than one germ cell layer.[11]

The size of SCT (average 8 cm, range 1 to 30 cm) does not predict its biological behavior.

3. TREATMENT OF SACROCOCCYGEAL TERATOMA

Surgical resection is the preferred treatment opinion for SCT and can determine the pathological type. Three surgical approaches are primarily used: transabdominal, transsacral, and transabdominal-sacral.[5] The transabdominal approach is usually used for higher lesions because it exposes the tumor to the back of the rectum, whereas the transsacral approach is preferred for small tumors located in the lower pelvis. The transsacral approach is considered a good option for tumors ≤10 cm in diameter and located below the third sacral vertebra, although specific criteria have not been established.[14] When the presence of malignant components is confirmed, the treatment model should be adjusted according to the pathological type.[5]

Recent reports have shown that a small number of SCTs can be safely resected by laparoscopy,[15,16] and some surgeons have also used endoscopy to remove tumors through the sacral approach.[17]

Provided that common sacrococcygeal malignancies are mainly immature teratomas and mature teratomas, upon confirmation of the presence of malignant components after postoperative pathology, the treatment mode should be adjusted according to the pathological type, and appropriate chemotherapy and radiotherapy are required.[5]

4. COCCYX REMOVAL

Owing to the increased malignancy rate of teratomas, early excision is recommended.[5] Whether the coccyx should be removed during surgery is controversial. The main reasons for coccyx resection are based on the following considerations: For large tumors, exposure during surgery may be required; SCT may originate from pluripotent cells of the coccyx; and tumors may tend to attach to the coccyx, and resection of the coccyx can prevent relapse.[18] After birth, early and complete surgical resection is key to the management of mature and immature SCTs, regardless of the size of the tumor or the patient's age.[19–22] The following investigations are recommended before surgery: X-rays of spine, abdomen, chest, and pelvis to detect evidence of tumor or metastasis; ultrasound to define the extent, nature (cystic or solid) of the mass, the presence and size of solid elements or calcification, and to recognize intrapelvic extension; computed tomography (CT) to delineate the bony pelvic structures and to identify small areas of intrapelvic extension; combined use of Doppler sonography and magnetic resonance imaging (MRI) to facilitate the antenatal diagnosis of SCT; angiography to demonstrate the main blood vessels supplying SCT – the middle and lateral sacral and gluteal branches of the internal iliac artery and a branch from the profundus femoris artery; as hypervascularity, irregularly encased vessels, arteriovenous shunting, and serpentine dilated veins may be seen in malignant, as well as in benign tumors, angiography is not conclusive in distinguishing benign from malignant disease; myelography to evaluate the intraspinal of SCTs.[1]

Therefore, regardless of the method, complete resection should be the primary treatment goal based on individualization, and the quality of life should be improved as much as possible. Unlike teratomas at other sites, SCTs often have no capsule or pseudocapsule, and complete resection of the lesion is difficult.[5]

5. RECURRENCE

Immature and malignant histology, incomplete surgical resection, and failure of *en bloc* resection of the coccyx are associated with a high risk of recurrence.[23] The overall recurrence rate differs between mature and immature teratomas; specifically, recurrence rate is approximately 10% for mature teratomas and 33% for immature teratomas.[24] For mature teratomas, recurrence rates range from 0% to 26% (mean 10%). High rates have been observed in patients with immature teratoma, that is from 12% to 55% (mean 33%).[24] The postoperative recurrence rate of lesions with incomplete resection is as high as 37%.[25] Notably, the coccyx may contain foci of totipotent cells, and if the coccyx is not resected simultaneously, the risk of tumor recurrence is 30% to 40%.[26]

Research confirms recurrence rates: 11 cases showed

recurrence, with an average recurrence time of 10 months after the operation, out of 173 cases of SCT of which 143 cases were benign,[17] a female patient showed recurrence at 14 months after the operation out of 12 patients of SCT,[27] and 4 cases showed recurrence after surgery with an average recurrence time of 11 years out of 5 cases of SCTs.[28]

6. OUTCOMES

Abnormal bladder function was reported in 18% (7 cases out of 39) Altman type I, 37.7% (23 cases out of 61) type II, 32.4% (11 cases out of 34) type III, and 60% (15 cases out of 25) type IV cases. Abnormal bladder function was also more commonly associated with malignant tumors versus benign or immature tumors. Patients who required more than one surgical resection episode were also more likely to develop urinary complications.[28]

Concerning sexual function, scores on the Female Sexual Function Index (FSFI) domains pain, orgasm, and satisfaction did not significantly differ between populations. In general, females treated for SCT reported good orgasmic function and low pain scores during intercourse and none reported severe dyspareunia, suggesting adequate functioning of the pelvic floor during sexual activity, even though pelvic floor anatomy was compromised because of the tumor pressure or operative treatment in a large proportion of patients. International Index of Erectile Function (IIEF) scores of males treated for SCT did not significantly differ from those of the control population. In research, males reported normal erectile function and penetration ability with normal ejaculation, but a larger sample size would be needed to conclusively establish the findings.[29]

7. RISK OF MALIGNANT TRANSFORMATION

The percentage of malignant transformation within 2 months after birth is 20% and 40% after 4 months, so it should be completely removed as soon as possible after birth to prevent malignant transformation.[10] Malignancy of any component may occur; however, ectodermal squamous cell carcinoma is the most common component.[30] Others include basal cell carcinoma, melanoma, adenocarcinoma, sarcoma, and thyroid cancer. Metastases of malignant teratomas include lymphatic metastasis to the retroperitoneal lymph nodes, hematogenous metastasis to the lung and bone, and poor prognosis after distant metastasis.[31] Poor prognosis exists, also, in cases where SCT transforms into carcinoma or sarcoma.[5]

Regarding adult SCT, the risk of malignant transformation ranges from 1 to 12%.[32]

8. PRENATAL COMPLICATIONS

Occasionally, a large SCT may cause an abnormal position of the fetus in utero; therefore, hip dislocation may occur. These conditions may be associated with potential long-term sequelae despite successful surgical resection of the primary tumor.[33,34] When the tumor is too large, dystocia or difficulty in delivery can occur. After birth, the tumor is present as a skin-covered tail mass. If the tumor is quite large, the skin covering the mass can be ulcerated and necrotized.[2] The neonatal death rate of prenatal diagnosed SCT is as high as 24% and the dead cases had higher tumor volume index and concentrations of NT-pro-BNP and cTnT than the survivors.[35] These perinatal high death rates of SCT are caused mostly by preterm labor, placentomegaly, the development of hydrops, and cardiac failure.[36,37]

Complications include urologic complications (the most common cause of morbidity), high output cardiac failure from arteriovenous (AV) shunting, which in turn can cause hydrops fetalis, gastrointestinal tract obstruction, compression of underlying nerves leading to urinary/fecal incontinence, anemia, dystocia, tumor rupture. Tumor compression of the bladder outlet caused retention in the urinary system, followed by renal impairment, oligohydramnios, and pulmonary hypoplasia, as well as tumor compression or infiltration of the sacral nerves and intraspinal extension of the tumor.[3]

9. CONCLUSIONS

This research confirms the already recommended treatment for the SCT: resection of SCT. A series of tests need to be done before the resection, but regardless of the method, complete resection should be the primary treatment goal based on individualization, so that the quality of life should be improved as much as possible. Coccyx removal does not affect sexual function in both males and females but is considered to provoke abnormal bladder function. SCT can, finally, create prenatal complications and this is the reason why extended research needs to be done to facilitate delivery and ameliorate the quality of life of both the gravida and the fetus and neonate.

ΠΕΡΙΛΗΨΗ

Διαχείριση ιεροκοκκυγικού τερατώματος και επικείμενες επιπλοκές

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Το ιεροκοκκυγικό τεράτωμα (SCT) είναι ο πλέον συχνός όγκος σε έμβρυα και νεογνά, με συχνότητα 1/35.000 έως 1/40.000 γεννήσεις και είναι εξαιρετικά σπάνιο σε ενήλικες με αναλογία γυναικών προς άνδρες 3:1. Παρά τη σπανιότητα των περιστατικών SCT, η αντιμετώπισή τους κρίνεται σημαντική για την εξασφάλιση αξιοπρεπούς ποιότητας ζωής. Η χειρουργική εκτομή είναι η προτιμώμενη θεραπευτική επιλογή για το ιεροκοκκυγικό τεράτωμα και μπορεί να καθορίσει τον παθολογικό τύπο αυτού. Υπάρχουν διαφορετικές προσεγγίσεις, σύμφωνα με την εκτομή: διακοιλιακή, διαϊερή, διακοιλιακή-ιερή. Η αφαίρεση του κόκκυγα κατά τη διάρκεια της επέμβασης είναι αμφιλεγόμενη. Ως αποτέλεσμα της αφαίρεσης του κόκκυγα παρατηρείται μη φυσιολογική λειτουργία της κύστης, σε αντίθεση με τη σεξουαλική λειτουργία, η οποία δεν επηρεάζεται ούτε στους άνδρες ούτε και στις γυναίκες. Ωστόσο, ο κίνδυνος υποτροπής και επανεμφάνισης εξακολουθεί να υπάρχει, καθώς επίσης ο κίνδυνος εξαλλαγής σε κακοήθεια.

Λέξεις ευρετηρίου: Εκτομή, Επανεμφάνιση, Ιεροκοκκυγικό τεράτωμα, Κακοήθης, Νεογνικός

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