


Sacrococcygeal Teratoma With Multiple Recurrences by Intradural Extension in the Neonate: A Common Neonatal Mass With an Unusual Course

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Case Report

D.P. was born at 32 weeks and 2 days via emergent C-section due to a 0/10 biophysical profile. D.P., twin B (twin A healthy), was prenatally diagnosed with a large sacrococcygeal teratoma (SCT) and bilateral hydronephrosis. Apgars were 4¹ and 8⁵, with a birth weight of 3.720 kg (100% percentile), length of 41.0 cm (20.9 percentile), and a head circumference of 30.5 cm (66.6 percentile). Physical exam noted large sacrococcygeal mass and flaccid lower extremities with no active noted response and decreased/absent primitive reflexes. Computed tomography (CT) scan confirmed Altman type II mass (13.7 cm × 12.6 cm × 15.6 cm) with suggestion for intraspinal extension to level T12. Subsequent magnetic resonance imaging (MRI) scans confirmed this finding with extension to the upper lumbar spine; an intrapelvic component causing mass effect on the ureters with hydronephrosis was also noted. Alpha-fetoprotein (AFP) was >120 000, and human chorionic gonadotrophin (HCG) was 63. On day of life 1, D.P. underwent surgical excision of the tumor (Figure 1), except the extension to the peritoneal and spinal region. Histopathology showed an immature teratoma without malignant elements (Figure 2). Postoperative MRI disclosed residual mass extending intraspinally to the upper thoracic spine. The patient underwent a thoracolumbar laminectomy and subtotal resection of the intradural and intramedullary extension of the SCT at the end of the first week of life. Intraoperatively, the tumor appeared to extend from T3-T4 to T8; below this level there appeared to be no spinal cord. Patient was discharged at 37 weeks 1 day of life. At that time AFP declined to 13 219.

Final Diagnosis

Sacrococcygeal teratoma of nonmalignant origin with immature element with multiple intradural recurrences

Hospital Course

At 2 months of age the patient was diagnosed with a urinary tract infection, and her head circumference had significantly increased. Brain MRI showed a right frontal mass. AFP had increased to 15 000. She underwent resection of the brain mass. Pathology was consistent with the previous mass: immature teratoma without malignant elements. AFP levels were followed and steadily declined to 33.2 (1 months postoperatively).

Two months later the patient presented to the emergency room with a fever. Evaluation included a CT/MRI, which showed new soft tissue mass lesion at the craniocervical junction extending into both internal auditory canals and the foramen of Luschka, resulting in hydrocephalus. MRI showed an amorphous, intermediate signal abnormality at the medullocervical junction region, lateral and anterior to the medulla, at the base of the fourth ventricle, extending into the upper cervical canal with no evidence of enhancement.

Two weeks after presentation, D.P. returned to the operating room for VP shunt and biopsy of the soft tissue mass. Histopathology confirmed mature teratoma, neural elements only; no malignant elements were noted. Due to persistent growth, various treatment options were considered and the decision was made to start daily α -interferon (3 million units/m²) and weekly Vinblastine (6 mg/m²). The patient received 1 year of chemotherapy. Prior to discontinuing chemotherapy, the patient underwent a syringopleural shunt for surgical decompression for an enlarging cervical syringomyelia. Routine surveillance imaging and AFP were followed

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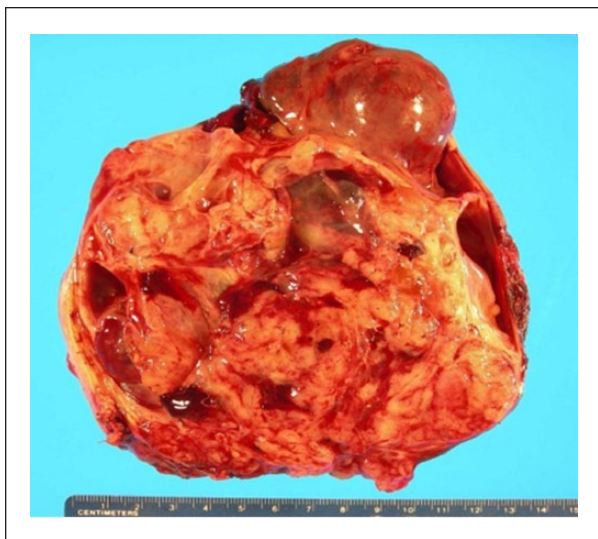


Figure 1. Gross pathology of removed sacrococcygeal teratoma (left).

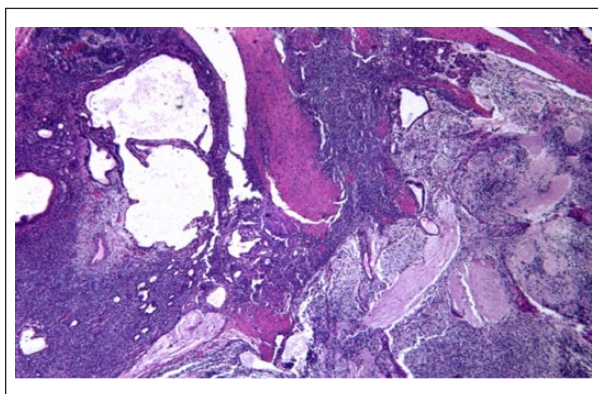


Figure 2. Pathology of removed sacrococcygeal teratoma with multiple tissue types, including glandular, primitive neural tubules, pancreatic, and neural white matter.

and remained stable. Four months after chemotherapy was completed her AFP and β -HCG normalized.

Follow-up with the patient at 2 years and 2 months of age (and 6 months off therapy) showed the patient in stable health with no concerning changes. The patient continued to have good upper extremity strength, with flaccid paralysis of the lower extremities, scoliosis, and a neurogenic bladder.

Discussion

Sacrococcygeal teratomas are one of the most common tumors of the newborn, occurring 1/21 000 births, and

are the most common site of germ cell tumors.¹ SCTs are benign, but can have malignant or immature characteristics, and they have increased risk of malignancy as age of diagnosis increases. They are more common in females but have higher rates of malignancy in males.² Elevated serum markers—classically AFP and HCG—suggest the presence of malignant elements. SCTs are often classified using the Altman classification.¹ SCTs are now more commonly detected prenatally with advancements in perinatal imaging.

These can be complicated by relapse and are found to occur with varying incidence. Overall survival is significantly lower in those who relapse.^{1,3} Risk factors for recurrence include immature or malignant histology, tumor spillage, and incomplete resection of the coccyx.^{1,3,4} Recurrence occurs in both mature and immature histology, with immature being more common. While recurrence does occur, to our knowledge multiple recurrences are seemingly rare, and even more so, via intradural extension.

On rare occasions these tumors lead to growing teratoma syndrome. This is defined as a nonseminomatous germ cell tumor that presents with enlarging metastatic masses during appropriate chemotherapy, or after resection, in the context of normalized serum markers (hallmark feature). The etiology of this syndrome has been debated, with the 2 most common explanations being the following: (a) chemotherapy destroys only immature malignant cells, leaving the mature benign teratomatous elements and/or (b) chemotherapy alters the cell kinetics toward transformation from a totipotent malignant germ cell toward a benign mature teratoma.⁵ While this case did not meet criteria for growing teratoma syndrome, these multiple recurrences were also unique as they were distant from the original site. Review of this case gives insight into the differing presentations of recurrence and a more complex course, as well as the importance of strict, regular, and prolonged follow-up.

Treatment of SCTs varies; however, surgery seems to be universal—removal of the coccyx leads to both a better prognosis and decreased risk of recurrence.^{1,3} With surgery, complications can inevitably arise, including preoperative and intraoperative tumor spillage, perforation of the rectum, and bleeding.

Conclusion

While many SCTs are successfully resected without subsequent recurrence, our case proved to have an unusual course. Multiple distant intradural recurrences were found over the course of 2 years. To our knowledge

no case has been reported with multiple recurrences of intradural extension, without meeting the classification of growing teratoma syndrome. In addition, intradural extension of teratomas proved to be rare in our search.^{6,7}

Growing teratoma syndrome is persistence of the germ cell tumor with enlarging metastatic masses during chemotherapy, with normal serum markers. Our case did not meet this definition as pathology was not malignant, tumor markers were elevated with each recurrence, and chemotherapy was not pursued until the third and final recurrence. This case did demonstrate the importance of long-term follow-up.^{3,8} It validated that follow-up is just as crucial even without initial malignancy or chemotherapy administration. While this scenario is seemingly rare, one must always be prepared for the possibility of a complicated course when managing and treating an SCT in the newborn.

Declaration of Conflicting Interests

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