Introduction

Neonatal sacrococcygeal teratoma is an unusual tumor that develops before birth and arises from embryologically multipotent cells from the Henson node, which is located in the coccyx.

Objectives

1. Describe a sacrococcygeal teratoma and the two distinct ways in which they generally present.
2. Explain the classification system used to categorize sacrococcygeal teratomas.
3. List two reasons post-operative follow up is needed for the first several years of life.

Incidence

Neonatal sacrococcygeal teratoma is rare, only occurring in approximately 1 in 35,000 births, with a female predominance (3:1-4:1 ratio).

Clinical Presentation

Sacrococcygeal teratomas generally present in two distinct fashions:

- **Newborns**
  - Large predominately external masses found in utero or at the time of delivery.
  - Rarely malignant.

- **Older Infants and Young Children**
  - Presents with tumors confined to the pelvis.
  - Experience a much higher rate of malignancy.

Diagnosis

Most sacrococcygeal teratomas are now diagnosed prenatally due to the routine obstetric ultrasonography. If detected prenatally, the neonate should be considered for abdominal delivery if the mass is greater than 5cm in size. Newborns generally present with the mass protruding from the sacral region.

Altman Classification of Sacrococcygeal Teratoma

- **Type I** tumors are predominately external (47%).
- **Type II** tumors present with both internal and external components (35%).
- **Type III** tumors are predominately hidden in the pelvis with very small external portion (9%).
- **Type IV** tumors are entirely hidden within the pelvis (10%).

Pre-Operative

Post-Operative

Long-term Sequelae

Long-term studies have demonstrated significant post-operative functional complications including constipation (30-36%) and urinary incontinence (19-22%). A neurogenic bladder has also been reported in 12% of children.

Post-Operative Follow Up

1. Constipation and urinary incontinence risks make it crucial for patients diagnosed with sacroccygeal tumor to have very close outpatient monitoring during the first few years of life to ensure bowel and bladder function is normal as they go through toilet training.
2. A recurrence rate of 10-20% should be taken into consideration. More importantly, the average malignancy rate is around 50%, making post-operative follow up essential.
3. Follow up until age 5 is recommended. Alpha feta protein levels and physical exams including rectal exams to evaluate for mass are recommended routinely.

Acknowledgements

Drs. Deborah Billmire and Fredrick Rescorla for providing surgical images.

References
