Sacroccygeal Teratomas in Children

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The earliest record of a sacroccygeal tumor was inscribed on a Babylonian cuneiform tablet, probably made by the Chaldeans who inhabited Mesopotamia from 625–539 B.C. The Chaldeans apparently regarded this protuberance on a newborn infant as an omen of prosperity rather than a medical curiosity. Today, much more is known of this interesting tumor: it is comprised of all three germ layers, contains a variety of mature and immature tissue, may be benign or malignant and is most commonly located in the sacroccygeal region of children. In a report of the Great Ormond Street Hospital experience from 1934–1969, Berry noted that 58 of 91 teratomas were located in the sacroccygeal region compared to 10 in the ovary and seven in the testis. Rarer locations include the mediastinum, thyroid gland, stomach and retroperitoneal space.

Several theories have been proposed to explain the origin of a tumor which occurs in newborn infants and contains cells from all germ layers. Bonet believed that the teratoma represented a displaced fertilized ovum while other investigators attribute its origin to incomplete twinning or so-called fetus-in-fetu.

However, according to the currently accepted theory, the sacroccygeal teratoma is probably derived from the totipotent cells of the primitive knot (Hensen’s node). As the somites develop, the primitive knot migrates in a cephalad direction and locates at the tip of the tail in the six-week-old embryo. As the tail retracts, the primitive knot comes back to rest at the tip of the coccyx in the ten-week-old fetus. It seems probable that some of the totipotent cells from Hensen’s node then develop independently from the rest of the fetus, thus forming the teratoma.

Epidemiology

Higher Incidence of Teratoma in Females than Males

The teratoma is seen predominantly in female children with a ratio of four females to one male. Gross reported that 32 of the 40 patients in his series were girls. Donnellan described 69 females with teratomas compared to only 34 males and noted an incidence of four females to one male in the newborn age group. This ratio of 4:1 has been substantiated by Hickey, Waldhausen and Chretien.

Higher Incidence of Cancer as Child Grows Older

One of the striking features of the teratoma is the increased incidence of cancer as the child grows older, Gross
reported 32 newborn patients with teratomas, all of which were benign. However, two of these children subsequently underwent excision for recurrent teratomas which were found to be malignant. In the eight older children in his series, five patients had malignant teratomas at first examination. Donnellan noted that in his 79 patients who were under two months of age, 90 percent were benign; 90 percent of the tumors in the 24 children over two months of age were malignant. Hickey reported that 95 percent of the teratomas removed before the patients were four months old were benign, whereas 61 percent of the teratomas removed after the child was over four months old were malignant. Waldhausen supported this evidence when he noted that 13 tumors removed before the children reached four months of age were all benign.

Eight patients between four months and 16 years old showed a 50 percent incidence of malignancy, although two patients over 16 years old both had benign tumors.

**Higher Incidence of Twins in the Immediate Family**

The incidence of twins in the immediate family is higher in children with sacrococcygeal teratoma than in normal children. Gross noted that three of his 40 patients were twins and a careful study of the families of his patients revealed a 50 percent incidence of twins. Hickey reported a four percent incidence of twins in his patients and a 14 percent incidence in the immediate family.

**Higher Incidence of Associated Congenital Abnormalities**

As would be expected in a tumor derived from primitive developing cells, the incidence of congenital abnormalities in this group of patients is high. Hickey noted that 11 percent of 112 patients had associated congenital abnormalities which occurred in the long axis of the child, such as spina bifida, cleft palate, patent urachus, meningocoele and undescended testicles. Waldhausen reported four cases of spina bifida and two of meningocele in his series and Gross observed two patients with associated abnormalities—cleft palate and thoracic spine deformity.

**Pathology**

In gross appearance, teratomas vary in size from a few centimeters in diameter to huge tumors which may interfere with the delivery of the child. The covering skin may be thinned out, shiny and show a bluish discoloration mimicking a hemangioma, or the skin may be wrinkled, rough and appear loose and redundant. Teratomas may be solid, cystic or, as is commonly the case, a combination of the two. The surface may be either smooth or grossly irregular. The cut specimens reflect this wide variety of gross appearance, namely, some are solid, some cystic, and others show a mixture of solid and cystic components arranged in an irregular, disorderly fashion. The cystic spaces often contain clear yellow or cloudy fluid but may also contain sebaceous material. In general, the more cystic the tumor, the greater the likelihood that it is benign. The solid areas may be hard or fleshy in consistency and it is here that the malignant elements are found. Occasionally, the cut surface is gritty and areas of calcification are common. Thus, the presence of calcium does not necessarily indicate that the entire tumor is benign.

Microscopic examination usually reveals a large variety of tissues from all germ layers. One germ layer may predominate, although a careful search will usually identify cells or structures from the other two layers as well. Muscle, cartilage, bone and nerve tissues may be found along with skin, alimentary tract, glandular and respiratory epithelium in the benign tumors. Malignant tumors present largely as embryonal adenocarcinoma. Benign and malignant, mature
Fig. 1. Clinical presentation of the primary tumor.*

1a. Congenital midline protuberance.

1b. Large presacral tumor with midline protuberance.

1c. No external tumor mass.

1d. Buttock enlargement.

*Adapted from Chretien, P. B.; Milan, J. S.; Foote, F. W., and Miller, T. R.: Embryonal adenocarcinoma (a type of malignant teratoma) of the sacrococcygeal region. Clinical and pathologic aspects of 21 cases. Cancer 26: 522-535, 1970.
and immature elements may be found side by side in the same tumor. Thus, careful histological examination of the whole tumor is necessary to rule out the presence of cancer.

Clinical Findings
The most common finding is a mass in the sacral-buttock area, varying in size from a few centimeters to as large as the baby's head. Almost all teratomas noted in the newborn period show some degree of external mass and many have presacral extensions of varying sizes which displace the anus and rectum forward. (Fig. 1.) Occasionally, especially in older children, no external mass is present; however, the tumor may grow internally from its presacrococcygeal site of origin. In these cases, rectum or bladder obstruction is common and often indicates cancer. Donnellan noted that only five percent of benign teratomas produce obstructive symptoms, whereas 80 percent of malignant tumors produce symptoms of partial bowel or urinary tract obstruction. Although a sacrococcygeal teratoma may mimic a hemangioma (Fig. 2), an infected pilonidal cyst or ischiorectal abscess, a careful rectal examination will demonstrate a tumor anterior to the coccyx, so that the sharp tip of this bone cannot be felt. The mass may feel solid, cystic or a combination of both. Other findings, such as vascular or lymphatic obstruction to the extremity are rarely seen, and almost always indicate cancer. In the absence of an associated myelocoele, neurological changes in bladder or anal sphincters or in the extremities also indicate cancer and are usually seen in recurrent tumors.

X-Ray Findings
A plain lateral film of the pelvis will usually show a soft tissue mass in the region of the coccyx; if there is significant presacral extension the mass may displace the rectum forward. In approximately half the cases, the plain film will show varying degrees of calcification; this finding is not a reliable index of benignancy. Barium enema is useful to determine the degree of presacral and intra-abdominal extension and shows more accurately any anterior displacement of the rectum. Intravenous pyelogram may be helpful if there is a large intra-abdominal component. However, it will rarely show evidence of obstruct

Fig. 2. Teratoma in left buttock mimicking a hemangioma.

Differential Diagnosis
Any tumor in the sacrococcygeal or presacral region or in the buttocks must be considered as a possible sacrococcygeal teratoma. Failure to recognize and
treat this lesion at an early stage may result in malignant change, and change the prognosis from excellent to hopeless.

Before making a definitive diagnosis, however, rule out other lesions which may be mistaken for teratoma including:

**Meningocele:** These lesions are entirely cystic and often associated with a sacral bone abnormality. Neurological defects in the sphincters of extremities are common. The meningocele will become tense when the baby cries and pressure over the lesion will be transmitted to the fontanelles. Occasionally, a meningocele and teratoma may be present in the same patient.

**Hemangioma:** A large hemangioma of the buttocks may mimic a teratoma and the consistency of the two lesions may be deceiving. However, a hemangioma will usually not have a presacral extension and the tip of the coccyx will be easily felt.

**Lipoma:** This tumor is occasionally seen and may also mimic a teratoma. Differential diagnosis requires excision.

**Rectal Duplication:** These can be located in the presacral region and may not be demonstrable by barium enema. Again, excision is necessary for diagnosis.

**Infected Pilonidal Cyst and Ischiorectal Abscess:** Occasionally, a teratoma in this region may become infected and differentiation from these lesions may be difficult. For draining abscesses in this location, tissue should always be obtained for histological diagnosis.

**Surgical Management**

Stanley reported the first successful removal of a sacrococcygeal teratoma performed by Blizard in 1841. Studies of individual cases and small series followed, but it was not until the classic papers by Gross and Ravitch that the true nature of this disease was understood. Modern surgical management of this interesting tumor is based largely on their work.

As soon as the diagnosis is made, complete excision of a sacrococcygeal teratoma is the only guarantee of cure. Excision should include the entire tumor...
and the coccyx, the site of origin.

In most instances, the tumor can be removed entirely from below through a perineal incision. In the rare patient where massive intra-abdominal extension is present, a laparotomy incision may be performed either at the same operation or a few days later. The child is placed face down and an inverted V or transverse elliptical incision is made across the middle of the buttocks. (Fig. 3.) The whole tumor is freed up from below, including the presacral extension. With the tumor mobilized, the coccygeal attachment will be apparent. (Fig. 4.) At this point the coccyx is removed from the sacrum in continuity with the tumor mass. (Fig. 5.) Attenuated levator ani muscles should then be approximated, and dead space obliterated by suturing levator muscles to the presacral fascia. Penrose drains are usually adequate to prevent accumulation of fluid deep in the wound. (Fig. 6.) These can be removed on the fourth postoperative day. The child should be nursed face down in the postoperative period, until the drains are out and the wound sealed. This ensures minimal soiling of the wound with urine or feces.

Results

If the tumor is benign and the entire coccyx is removed with the tumor, a cure rate in excess of 90 percent can be achieved. If the coccyx is not removed, an approximately 33 percent recurrence rate can be expected with a high degree of malignant change in the recurrent tumors. If the teratoma is malignant from the onset, usually the case in older children, the cure rate approaches zero. However, even in patients with malignant sacrococcygeal teratomas, total excision should be carried out, followed by irradiation and chemotherapy. Although patients with recurrent malignant sacrococcygeal teratomas may be palliated by irradiation therapy and chemotherapy, no cures have been reported at this time. Most of these children will expire within two years of the diagnosis.
Summary

1. Sacrococcygeal teratomas are seen most commonly in infancy and early childhood, a large portion being present at birth.

2. The tumor is seen predominantly in girls with a ratio of four females to one male.

3. Teratomas occur more frequently in twins or in families where there is a history of twins.

4. A tumor diagnosed before the age of four months is likely to be benign; a tumor diagnosed after the age of four months is likely to be malignant.

5. The cure rate of benign sacrococcygeal teratoma is over 90 percent; the cure rate of malignant teratoma is near zero.

6. All sacrococcygeal masses should be excised before the child is four months of age, if possible. If the tumor proves to be a teratoma, the whole tumor mass and entire coccyx must be removed.

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