ISSN 1019-8555
The Journal of
Teachers Association
RMC, Rajshahi

Case Report

Sacrococcygeal Teratoma in a Teen—a Case Report

Md. K Rahman[1], S M M Haq[2], A K Sarker[3], S M A Shahid[4], M B Islam[5],

Abstract

Sacrococcygeal teratoma is a common congenital neoplasm. This tumor contains derivatives of more than one of the three embryonic germ cell layers e.g. ectoderm, mesoderm & endoderm and usually arises as a mass in the sacrococcygeal region. Here we are reporting a case of huge sacrococcygeal teratoma presenting as a lower abdominal and perineal mass in a thirteen-year-old schoolgirl, which is very rare.

TAJ 2003; 16(2): 76-78

Introduction

The word 'Teratoma' is derived from the Greek *teratos* which means "monster". [1] This tumor, derived from the three germinal layers is found frequently in the sacroccocygeal region. In fact, this is the most frequent site of teratomas of the fetus.[2]

About 67% of sacroccocygeal teratomas are diagnosed by the age of one year. The incidence is 1 in 35,000 to 1 in 40,000 live births. Females are four times more likely to be affected than males, but malignant change is seen more frequently in males. In childhood, they normally occur as extragonadal masses located along the midline, about 40-50% occurs in the sacroccocygeal region. Gonadal teratomas occur more frequently after puberty.[3]

The etiology is unknown, but has been suggested that it is derived from totipotential cells in Hensen's node. A theory of "twinning accident" with incomplete separation during embryogenesis has also been proposed. Some cases of sacroccocygeal teratoma are familial, with autosomal dominant inheritance. Sacrococcygeal teratoma has been reported in an infant born to a mother treated with acetazolamide until the 19th week of pregnancy. Most sacrococcygeal teratomas appear solid or varie-gated in nature, containing randomly arranged, irregularly shaped cysts. [6]

Development of teratoma in the precoccygeal region explained by the fact that this area is the site of the 'Primitive Knot', a group of totipotent cells that retain their totipotentiality longer than any others save the sex analage[1].

Possibly there are two types of tumors. One arises from the distal portion of the sacrococcygeal region, clinically obvious from birth and nearly always benign.

Another type arises proximally in the retrorectal or adjacent retroperitoneum, is noticed after birth, usually gonadal origin and malignant from the start.[5]

[1] Professor, Department of Surgery, Rajshahi Medical College, Rajshahi.
[2] Indoor Medical Officer, Surgery Unit – 2, Rajshahi Medical College Hospital, Rajshahi
[3] Assistant Professor, Department of Surgery, Rajshahi Medical College, Rajshahi.
[4] Assistant Registrar, Surgery Unit – 2, Rajshahi Medical College Hospital, Rajshahi.
[5] Registrar, Surgery Unit –1, Rajshahi Medical College Hospital, Rajshahi.

Most teratomas in infancy and childhood are benign; there is a tendency toward development of malignant teratomas with increasing age.

Case History

A thirteen year old school girl was admitted in a surgery unit of Rajshahi Medical College Hospital with a huge lobulated swelling in her sacro-perineal region and in the lower abdomen. According to the statement of her mother the swelling in her back is present since birth. The patient also noticed a lump in her lower abdomen from the last one year.

On abdominal examination, a mass was found to occupy her pelvic region and lower abdominal cavity corresponding to 22 wks of pregnancy. It was painless and slowly growing in nature. The mass was irregular in shape and size but the skin was free. The mass was occupying the entire lower abdomen and was little mobile. The mass also bulged through the perineum just behind the anus. She had increased frequency of micturition with intermittent retention of urine and was constipated for the last nine months. Her menstruation started at 12 years of age and was regular.

Per Rectal examination revealed that her anal orifice was pushed forwards towards the symphysis and the rectum was pushed anteriorly as the mass was palpable posteriorly through the rectum.

Per Vaginal examination showed that her vagina admits only one finger and approach to the fornices were difficult.

Ultra sonography of the abdomen found an enormous sized predominantly cystic mass occupying most of the lower abdominal cavity with multiple thick internal septations and solid tissue component. The mass was located postero-inferior to the urinary bladder, suggestive of intra pelvic pre sacral cystic mass which is likely to be a sacrococcygeal teratoma.

Aspirated fluid was cloudy in appearance but bio-chemically it was transudative. Cytological smears showed some well differentiated squamous epithelial cells and abundant squamous of Keratin. The features were very much consistent with Dermoid cyst.

IVU with cystogram found slight dilated calyces in left kidney. Both the ureters were distorted and deviated laterally. Urinary bladder was over distended and quite high in position.

The patient was operated under general anesthesia through abdomino-perineal approach by two team of surgeons. The teratoma was removed in toto with great difficulty.

Macroscopic features revealed that the cyst was filled with thick fluid which was dirty white in colour; a gruel like substances with small deposits and tissues of different germ layers e.g. hair follicle, teeth, epithelial tissue, muscle, bones etc were found (Fig-1). Both the ovary and the uterus were found to be normal.

The lower abdominal and perennial incisions were then closed in layers keeping drains. Her postoperative period was uneventful. The patient was discharged on fourteenth postoperative day and advised for regular monthly follow-up.

Histopathology of the resected lump showed the features of mature cystic teratoma.

Fig. 1: Divided operated specimen for histopathology

Discussion

About 67% of sacroccocygeal teratomas are diagnosed by the age of one year. The incidence of teratoma is 1 in 35,000 to 1 in 40,000 live births. Females are four times more likely to be

affected than males, but malignant change is seen more frequently in males. In childhood, they normally occur as extragonadal masses located along the midline, about 40-50% occurs in the sacrococcygeal region. Gonadal teratomas occur more frequently after puberty.[3]

In this case the presentation was late, which was probably due to lack of awareness and lack of proper diagnostic facilities.

The treatment of choice for sacrococcygeal teratomas is surgical excision. Most teratomas in adults can be excised by a posterior parasacrococcygeal approach. However, teratomas that extend proximally in the pelvic cavity have to be removed by a combined abdominal and perineal approach. As it was huge in size abdomino-perineal approach was more suitable. Her pelvic floor became weak during dissection. So she was on non-residual diet and catheterized for fourteen days to give rest to the pelvic floor.

In patients with teratomas, the coccyx often contains nests of totipotential cells and therefore, it should be removed en block with the tumor. Failure to remove the coccyx has been associated with a high risk of recurrence. For mature and immature teratomas, the prognosis is good after surgical excision alone, but malignant teratomas have a tendency to recur and metastasize. Therefore, teratoma should be removed meticulously if any part remains; regular follow up can detect early recurrence.

References

1. Russel RCG :Bailey & Love's Short Practice of Surgery, 23rd Edition' 2000; 66; 1121.
2. Garcia AM. *In utero decompression of a cystic grade IV sacrococcygeal teratoma.* Feial Diagonosis & Therapy. 1998; 13 (5): 305-8.
3. Graf JL. *A surprising histological evolution of preterm sacrococcygeal teratoa.* Journal of Pediatric Surgery. 1998; 33 (2): 177-9.
4. Holterman AX. *The natural history of sacrococcygeal teratoma diagnosed through routine obstetric sonogram: a single institution experience,* Journal of Pediatric Surgery. 1998; 33 (6): 899-903.
5. Kawamura DM. *Abdomen and Superficial Structures,* 2nd ed. Lippincott-Raven Publishers. 1997; 15: 532-33.
6. Putman CE. *Textbook of Diagnostic Imaging,* 2nd ed. W.B. Saunders Company. 1994; 26: 316-318
7. Rubin E. *Pathology.* 3rd ed. Lippincott-Raven Publishers. 1999; 277.

All correspondence to:
S M Mahmudul Haq
Indoor Medical Officer, Surgery Unit - 2,
Rajshahi Medical College Hospital,
Rajshahi.