Sacroccygeal Masses Other Than Meningomyelocele

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Abstract This series comprises of a variety of sacroccygeal masses other than meningomyelocele that presented to the department of pediatric surgery of Medical College Kolkata over last 10 years. In this series, 23 cases of sacroccygeal masses are included. Barring meningomyelocele, teratoma constitutes a major group of cases. It also includes few other interesting and atypical masses such as presacral dermoid, degenerated nerve fiber, fibrofatty tissue, rhabdomyosarcoma, etc. This is an endeavor to enlighten ourselves so that the diagnosis and management of unfamiliar sacroccygeal masses can be done.

Keywords Sacroccygeal masses

Introduction

Sacroccygeal masses are lesions present since birth. They comprise of both benign and malignant lesions present between the skin and sacrum. Congenital anomalies and embryonic rest cells are frequent causes of these lesions. Trauma and inflammation are other causes. Some of these etiologies are benign and some are overtly malignant.

Though a majority of series is made up of meningomyelocele, some other lesions have different tissues of origin.

The management of these entities depends on prompt diagnosis and appropriate treatment.

Materials

This series comprises of the pediatric patients presenting at the department of pediatric surgery of Medical College Kolkata in last 10 years. This series has 23 cases covering different etiologies and their management.

Methods

All the patients of sacroccygeal masses were first stabilized hemodynamically. Thorough clinical assessment was done including *per rectal examination*.

The patients having meningomyelocele were excluded from this series.

All the children underwent baseline hematological investigations. X ray of lumbosacral spine, USG of whole abdomen including pelvis was done. Signs of other anomalies were searched for.

The children having teratomas type III and IV were subjected to CT scan to know the abdominal extent. Two children having type II were subjected to CT scan as there was confusion in diagnosis. Children having solid tumors, degenerated tissue were subjected to CT to come to a definite diagnosis.

The children after resuscitation were subjected to standard operative procedures.

All children having teratomas type I, II, III and solid tumors were placed in prone knee chest position with hips flexed with head end low down with urinary catheter in place.

An inverted chevron incision was made & mass was gently separated out from gluteus muscles, ischioanal fats

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and rectum carefully. Hemostasis was achieved by diathermy coagulation. The entire mass was excised enbloc with last piece of coccyx (Photo 1).

Teratomas type IV were approached by abdomen and excised. Care was taken not to damage the presacral nerves.

Soft tissue masses were excised keeping a wide margin of clearance.

Diagnosis of case having degenerated tissue was not made till the last. The child did not have any neuro deficit. So excision was planned and no spinal connection was found during operation. Histopathology reports came out to be degenerated nerve tissue.

Post operatively all these cases were monitored in high dependency units and close check on proper temperature and glucose level was kept.

The entire specimens were subjected to histopathology examination. In the case of rhabdomyosarcoma, incision biopsy was done to confirm the diagnosis as the mass was huge at presentation.

Results

Out of 23 cases in this series 22 cases were operated. 21 cases recovered very well and were discharged in one week.

Average time of operation was 1 hours 30 min.

One child of 1 month having a large sacrococcygeal teratoma died on arrival due to hypothermia and hypoglycemia because of inadequate attention during transfer.

One case of type II Teratoma died during recovery due unknown reasons.

After the post op investigations the etiologies came out as follows:

Sacrococcygeal Teratoma:	17 cases
TYPE I	6
TYPE II	7
TYPE III	2
TYPE IV	2
Presacral Dermoid	1 case
Degenerated nerve tissue	1 case
Fibrofatty tissue	2 case
Rhabdomyosarcoma	1 case
Gluteal lipofibroma	1 case

Discussion

Sacrococcygeal masses in children can be of various etiologies (Table 1). Of the various presentations teratomas comprise of a major portion of sacrococcygeal masses.

Photo 1 Curarinos syndrome after excision till last piece of coccyx

Table 1 Different types of sacrococcygeal malformations

Benign
Congenital
1. Meningomyelocele
2. curarinos sundrome
3. anorectal malformation
4. infantile hamartoma
5. tail remnants
6. Dermoid
Inflammatory
1. rectal abscess
2. granuloma
3. osteomyelitis of sacrum
Neurogenic
1. Primary neurulation defect
2. Secondary neurulation defect
3. Caudal neural tube defect
4. neuroenteric cyst
5. degenerated nerve tissue
Malignant
1. teratoma and its variations
2. Rhabdomyosarcoma
3. basal cell carcinoma of the perineum
4. malignant basaloid (cloacogenic) carcinoma of the anal canal.
Miscellaneous
1. ulcerated lesions
2. trichoepitheliomas
3. Fibrofatty tissue
4. epidermoid cyst
5. pilonidal cyst

Photo 2 Large teratome where child is appendage of tumor

The term “Middledorpf tumors” has been used by some authors to describe all presacral and sacral tumors of embryonic rest origin [1, 2]. Apart of embryonic origin certain hamartomatous changes can also form masses in this region.

Teratomas are tumors arising from totipotent postmitotic but premeiotic cells. They are usually midline structures [3, 4]. They are derived from Greek word *teraton*, meaning monster. They are diagnosed commonly in the prenatal period, and may present with complication which may occur in utero, during or after birth and include polyhydramnios nonimmune hydrops fetalis etc. Atrioventricular shunting may lead to hydrops from highoutput cardiac failure. Due to large size of tumor and ill developed adapting capacity, children are at high risk of hypothermia (Photos 2 and 3). They may undergo ischemic necrosis and present as ulcerated lesion (Photo 4) Elevated serum alpha-fetoprotein (AFP) and beta-human chorionic gonadotropin (HCG) levels may be indic-

Photo 3 Larger teratoma

Photo 4 Atypical teratoma which had ulcerated

ative of malignancy. The treatment of mature teratomas is largely **surgical excision**. They may be complicated by malignancy which is usually adenocarcinoma having an incidence of 10.1% at birth and 91.7% when discovered after 2 months [5]. **Rhabdomyosarcoma** (RMS) is the most common soft tissue cancer of children arising from primitive muscle cells [6]. They commonly present at head and neck region and rarely in sacrococcygeal area (Photo 5).

It may be associated with some syndromes as Neurofibromatosis, Li-Fraumeni syndrome, Rubinstein-Taybi syndrome, Gorlin basal cell nevus syndrome, Beckwith-Wiedemann syndrome, Costello syndrome etc [7].

Imaging investigations include CT MRI & bone scan for metastasis to bone.

Cytogenetics, fluorescent in situ hybridization (FISH) is done to diagnose translocations & reverse transcriptase–polymerase chain reaction (RT-PCR) testing is done to assess characteristic translocations [8].

Photo 5 Sacrococcygeal rhabdomyosarcoma

Photo 6 Degenerated nerve tissue

Treatment in patients with RMS involves a combination of surgery, chemotherapy, and radiation therapy.

Sacral masses can be complicated with presacral mass, imperforated anus and sacral bone anomaly. This syndrome is known as **Currarino Triad**. It is an autosomal dominant genetic disorder [9–11]. It can be complicated with renal failure due to reflux. Fistula between colon and meninges can lead to meningitis. Because the incidence of Currarino triad is high when there is an anorectal stenosis (38% in the present series), the triad should be suspected in cases having anorectal stenosis. Magnetic resonance imaging is a preferred diagnostic method because it can diagnose tethered cord which has a high incidence of association with this triad. Treatment constitutes of repair of meningocele, followed by repair of anorectal malformation with excision of presacral masses. The anorectal malformation gives good results with Posterosagittal ano recto plasty after excision of last piece of coccyx (Photo 3).

Tail remnants are usually hamartomatous process or caudal vestige composed of Fibrofatty adipose tissues with smooth muscles, vascular structures, and cartilage (Photo 6) which occurs since birth in the perianal region. These are benign lesions not attached to the coccyx. Simple excision of the lesion is sufficient for the cure, though when associated with anal stenosis, anoplasty is required [12].

Degenerated nerve tissues in the sacrococcygeal region are unknown entity with no reference in literature (Photo 7). Probably, it is a malformation which might have a communication with spinal cord structures in embryonic period but development and differentiation was aborted later

Photo 7 Sacrococcygeal fibrofatty tissue

by complete and successful approximation of sclerotome and myotome components of intermediate cell mass.

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