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Original Research Paper

Surgery

SPECTRUM OF SUBTYPES OF SACROCOCCYGEAL TERATOMA: A CASE SERIES STUDY IN A TERTIARY CARE HOSPITAL IN CENTRAL INDIA: A SURGEON'S AND A RADIOLOGIST'S PERSPECTIVE

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ABSTRACT

OBJECTIVE: 1.To demonstrate the clinical presentation, radiological features, management and histopathological features of sacrococcygeal tumors in neonates.

Material And Methods: Between 1st March 2020 and 1st December 2020, 5 cases(4 male, 1 female) of sacrococcygeal teratomas(SCT) were diagnosed in Government Medical College Nagpur using ultrasonography(USG) and Computed tomography(CT) scans. In each case, tumor size, its content, mass effect, and classification according to the Altman's criteria were determined and compared with other features. Then the patients underwent surgery and samples were sent for histopathological correlation.

Results: Among the 5 patients, 3 were of Type I type and 2 were of type II type. In histopathological reports, 4 were of mature type and 1 was of immature type.

Conclusion: USG and CT scans are effective in diagnosing and localizing the extent and involvement of SCT. SCT appear to be entirely benign during the neonatal period. Complete surgical excision remains the mainstay of treatment.

KEYWORDS : USG,CT scan,SCT, Mature and Immature type

INTRODUCTION

Of all congenital tumors, sacrococcygeal teratoma (SCT) is the most common, with a reported incidence of 1 : 35,000–40,000 [1,2] and a 3 : 1 female predominance [3]. Recent studies suggest that the incidence can be as high as 1 : 14,000 [3], and prenatally diagnosed tumors account for 50% of cases [4]. Fetal teratomas are believed to arise from an aggregation of totipotent cells in the primitive streak, also called the Hensen's node. Remnants of this area may persist and give rise to SCTs that contain cells from all 3 germ layers. While the majority of these tumors are benign, perinatal mortality rates are high and range from 13% to 16% for prenatally diagnosed cases [4]. These perinatal deaths are caused mostly by preterm delivery, cardiac failure, or damage to the tumor resulting in hemorrhage [5,6]. Large teratomas are highly vascular and contain significant amounts of blood which may lead to fetal anemia and high-output cardiac failure [3,5]. The clinical course of fetal SCTs is often unpredictable, with some tumors growing rapidly while others retain their initial size. Numerous predictive factors have been investigated, including tumor weight to fetal volume ratio, tumor structure, and vascularity [3,6-8].

MATERIALS AND METHODS

Between 1st March,2020 and 1st December 2020, 5 cases of SCTs were diagnosed in our center (4 males, and 1 female). The patients were clinically assessed and then sent for Ultrasonography. After that , the patients were subjected to Computed tomography scan. In every case, tumor size, its extent, and mass effect on nearby tissues were evaluated. Then the patients were treated surgically and the specimen were sent for histopathological analysis. HPE report was given as mature or immature teratoma depending on the histological features. Every tumor was classified according to the Altman's criteria (American Academy of Pediatrics Surgery Section Survey) (Table 1)

Equiments:

1. 256 Slice CT machine PHILIPS
2. GE LOQIQ S8 Ultrasound Machine: Transabdominal Probe (1-5 Hz); Linear Probe(4-6 Hz)

Table 1: Sacrococcygeal Teratoma Staging According To The American Academy Of Pediatrics Surgery Section Survey

TYPE	CHARACTERESTICS
TYPE I	Developing only outside the fetus (can have small pre-sacral component); accounts for the majority of cases, 47% [9]
TYPE II	Extra-fetal with intrapelvic presacral extension
TYPE III	Extra-fetal with extension through pelvis into abdomen
TYPE IV	Tumour developing entirely in the fetal pelvis

RESULTS

In 3 of the reviewed cases, type I SCT was diagnosed, and type II tumour was seen in 2 patients. 4 of the 5 cases were of mature type and 1 of the 5 case were of immature type.(Fig 1). The distribution of sex is shown in figure 2 in a tabular form.

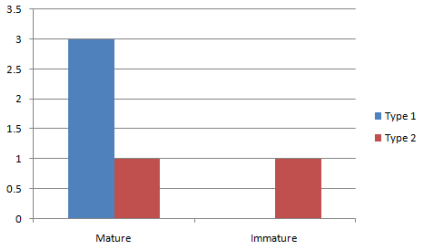


Figure 1: Reveals the distribution of mature and immature teratoma with respect to types of Sacrococcygeal teratoma.

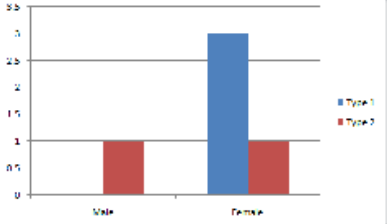


Figure 2: Reveals the distribution of sex with respect to types of sacrococcygeal teratoma.

Type I Sacrococcygeal Teratoma: (Figure 3)

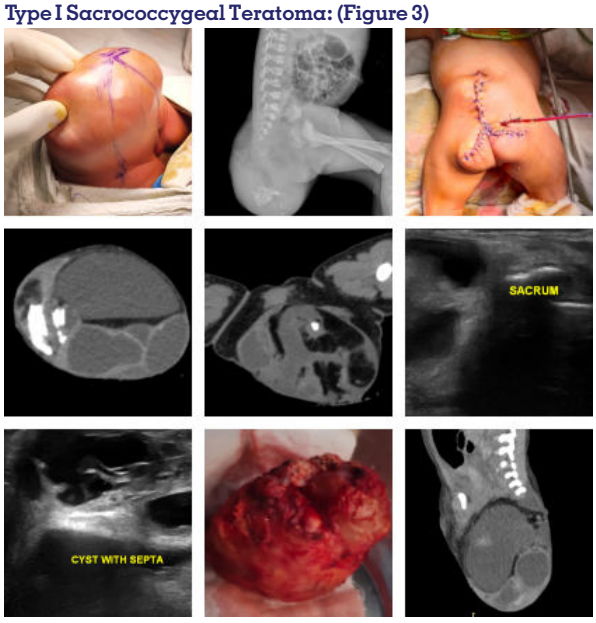


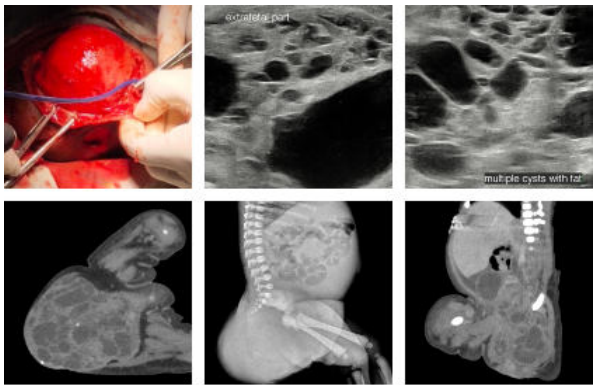
Figure 3: Shows clinical, Radiographic, CT and USG pictures of the 2 day old female who presented with sacrococcygeal mass.

A 2 day old female child presented to surgery casualty with presence of a large lobulated mass in sacrococcygeal region . On ultrasound, there is a well defined round to oval solid cystic lesion of size 6.2x7.9x6 cm in sacral region in subcutaneous and intramuscular plane with multiple cystic lesions and areas of calcifications. No extension into pelvic region was noted. On Contrast enhanced CT scan, there is a large relatively well defined lobulated heterogeneously enhancing lesion of size 8.3x6.2x9.3 cm arising from lateral aspect of L5 to S5 vertebra. Solid component measures 3.3x4.5x5 cm. No pelvic extension was noted. The lesion is showing intraspinal extension into the neural canal from the left side at S3 and S4 levels.

The patient was taken for surgical excision. Patient was kept in jack-knife position and taken under general anesthesia; inverting incision was given; incision deepened and dissection done in subcutaneous place; evidence of soft cystic mass approximately 8x8x9 cm posteriorly involving the rectum. Cyst was separated from rectum and excision was done. Coccygectomy was done along with excision of mass. Mass was sent for HPE analysis. Report came out to be mature sacrococcygeal teratomas.

Other 2 cases were of similar morphology measuring 5.1x4.2x8.3 cm and 5.7x4.8x9.3 cm. No intrapelvic extension was found in any of the cases.

Type 2 Sacrococcygeal Teratoma(Figure 4):



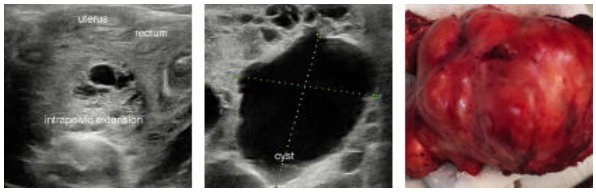


Figure 4: Shows clinical, Radiographic, CT and USG pictures of the 4 day old female who presented with sacrococcygeal mass.

A 4 day old female child presented to surgery casualty with presence of a large lobulated mass in sacrococcygeal region with complaints of bladder and bowel incontinenct. On ultrasound, there is a well defined round to oval solid cystic lesion of size 10x7x10 cm in sacral region in subcutaneous and intramuscular plane with multiple cystic lesions and areas of calcifications with possible extension into pelvic region was noted. On Contrast enhanced CT scan, there is a large relatively well defined lobulated heterogeneously enhancing lesion of size 7.6x8.10.1 cm arising from lateral aspect of L5 to S5 vertebra. Solid component measures 4.2x4.1x2.2 cm. Pelvic extension was noted to compress the bladder and rectum. It also compresses bilateral ureter and pelvicalyceal system to cause bilateral hydroureteronephrosis. Mutliple calcification and fat density foci noted.

The patient was taken for surgical excision. Patient was kept in jack-knife position and taken under general anesthesia; inverting incision was given; incision deepened and dissection done in subcutaneous place; evidence of soft cystic mass approximately 10.3x7.4 x 10.6 cm posteriorly involving the rectum with extension into the pelvis. Cyst was separated from rectum and excision was done. Coccygectomy was done along with excision of mass. Mass was sent for HPE analysis. Report came out to be mature sacrococcygeal teratomas.

Another child was of 3 day old presented with similar morphology and size(8.9x6.7x9.2 cm) with pelvic extension. He had complaint of neurodeficit also. HPE report of this tumour was immature teratoma. As it is of malignant nature, post-operative chemotherapy and radiotherapy was planned.

DISCUSSION

The newborn with sacrococcygeal teratoma (SCT) has an excellent prognosis depending on the timing of diagnosis, malignant potential of the tumour and the ease of surgical resection . Although prenatal diagnosis is possible in some cases, this was not possible in our cases since all the mothers did not attend antenatal care. SCT could be diagnosed from the second trimester of pregnancy when there is polyhydramnious and/or a uterus larger than the gestational age . Prenatal diagnosis is of significance since early prenatal presentation is associated with high fetal morbidity/mortality, while presentation after 30 weeks gestation is a relatively good prognostic indicator for fetal survival . In addition, such early diagnosis may predicate delivery by caesarian section in centers with good neonatal facilities where early surgical treatment can be offered to the baby. Similarly, fetal surgical interventional procedures could be undertaken when the diagnosis is made early in pregnancy . Since our patients' mother had no antenatal care, and delivery was not supervised by trained personnel, the chances of both maternal and child mortalities from obstetric complications were high. Because of the disposition of the tumour, this particular case could have led to dystocia, prolonged labour and damage to maternal birth canal. We presented four female and one male case. Although about 75% of cases are seen in females, the exact reason for female preponderance is not fully understood. A well-planned surgical excision was undertaken in our patients, including preoperative skin markings to enable flap reconstruction of the defect . Care was

also taken to ensure inclusion of the coccyx in the excision material in order to forestall possible recurrence. Apart from age at diagnosis and treatment, and the extent of resection, the prognosis is also determined by the histologic type and stage at the time of resection, and not the size of the tumour . Complete excision (including coccygectomy) as in our own cases, is the primary therapy for all SCT and it is adequate if the tumour is benign. Chemotherapy and radiotherapy are however indicated in malignant cases . Where the tumour is adjudged malignant, a recurrence as high as 37% has been reported if the coccyx is not removed in the primary surgery . Follow-up in patients with SCT is necessary especially during the first three years of treatment when recurrence is more likely . Extensive surgery in the pelvic and perineal region may involve disruption of nerves and muscles which supply the urinary/ano-rectal sphincters and provide maximum support in normal working respectively. In the 3-year period of follow-up of our patient, he has maintained good urinary and anorectal function, with no clinical, biochemical or radiological evidence of recurrence. Longer period of follow-up is however required, in order to make our claim more authentic.

Abbreviations
USG: Ultrasonography
CT: Computed tomography
SCT: Sacrococcygeal Teratoma
HPE: Histopathological Examination

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