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RESEARCH ARTICLE

SACRO-COCCYGEAL TERATOMA – CLINICAL ASPECTS AND HISTOLOGICAL EVALUATION IN PAEDIATRIC PATIENTS

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ABSTRACT

Background: Sacrococcygeal Teratomas (SCTs) are germ cell tumors arising from sacrococcygeal region containing derivatives of all three germ cell lines. It is predominantly affecting neonates, infants and children along with female preponderance. Classifying SCTs according to its location and histological grading in respect to presence of immature elements are helpful in early diagnosis and therapeutic decisions, providing better outcome.

Aims and objectives: Study of histological evaluation and grading of sacrococcygeal teratoma as well as correlation with clinical aspects in paediatric patients.

Material and method: A retrospective study of 22 patients presenting with sacrococcygeal mass at B.J.M.C, Civil Hospital, Ahmedabad over a period of three years from 2014 to 2016. All specimens of resected saccrococcygeal mass were received and overnight fixation in 10% formalin was done. Gross examination findings were noted followed by routine paraffin embedding and tissue sectioning. Slides were stained using H&E stain, examined microscopically and grading of SCTs was done along with clinical correlation followed by comparison with other studies.

Results: In present study, 22 cases which presented with sacrococcygeal mass were evaluated. There were 15 girls and 7 boys (male:female ratio of 1:2) with the age ranging from 1 day to 5 years. Majority of the cases (18(82%)) presented in infants below 2 months of age. One patient presented with recto-vaginal fistula and another with urinary retention. 19 cases were of mature teratoma (grade 0) and 3 cases were of immature teratoma (grade I) including one case of malignant transformation with yolk sac differentiation (grade II), which correlated with elevated serum AFP levels. On microscopic examination, mature teratoma showed ectodermal, endodermal and mesodermal differentiation while immature teratoma showed immature neuroepithelial elements.

Conclusion: Sacrococcygeal teratoma is the commonest congenital neoplasm presenting at birth as a sacrococcygeal mass with female preponderance. Many of them are presenting with associated anomalies or constipation and urinary retention due to mass effects. When diagnosed early at birth, majority of them are of benign mature type, so that meticulous search for immature or malignant component has to be undertaken. However early diagnosis is helpful in therapeutic decisions, providing better outcome.

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INTRODUCTION

Sacrococcygeal teratoma (SCT) is the most common congenital neoplasm (Kouranloo *et al*., 2006), occurring in 1 in 40,000 infants (Robbins and cotran 9th edition). Sacrococcygeal teratomas are tumors derived from more than one embryonic germ cell layer and are usually diagnosed in infancy (Bhosale *et al*., 2015). It has been hypothesized that they are derived from the pluripotent cells of hensen's node, a remnant of primitive streak located anterior to the coccyx. In 1937, it was the first time when sacrococcygeal teratomas were elaborately reviewed (Ramani *et al*., 2013). Sacrococcygeal teratoma was first described in 1951 in a study which included forty infants and children (Ramani *et al*., 2013). Majority of SCTs are solid-cystic on macroscopic evaluation (Kouranloo *et al*., 2006). Besides its clinical behavior, another important clinical aspect regarding sacrococcygeal teratoma is its anatomical location. According to location Altman classification divides SCTs into 4 types (Sacrococcygeal teratoma). Histologically, sacrococcygeal teratomas are classified as mature, immature and malignant (Ramani *et al*., 2013; Kostas *et al*., 2006; Robbins and Cotran 9th edition). Mature teratomas are chiefly composed of differentiated tissues and considered benign (Fig 2a- 2e). Immature teratoma is characterized by the presence of immature non-malignant tissue, predominantly neuroepithelial

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origin (Fig 2g). Teratomas with features of yolk sac tumor (Fig 2h), choriocarcinoma or embryonal carcinoma among differentiated tissues are regarded as malignant which can be correlated with the respective serum markers e.g. S.AFP, S. Beta HCG etc (Ramani *et al*., 2013).

Aims and Objectives

Study of histological evaluation and grading of sacrococcygeal teratoma as well as correlation with clinical aspects in a paediatric patients.

MATERIALS AND METHODS

A retrospective study of 22 patients presenting with sacrococcygeal mass at B.J.M.C., Civil Hospital, Ahmedabad over a period of three years from 2014 to 2016. All specimens of resected saccrococcygeal mass were received along with available clinical details and overnight fixation in 10% formalin was done. Gross examination findings were noted followed by routine paraffin embedding and tissue sectioning. Slides were stained using H&E stain, examined microscopically and grading of SCT was done along with clinical correlation followed by comparison with other studies.

RESULTS

In the present study, 22 cases presenting as sacrococcygeal mass were evaluated. These tumours frequently occurred in the infants less than 2 months of age (Rosai and Ackermann 10 edition). In the present study, 18(82%) of 22 cases presented in that age group. There were 15(68%) female and 7(32 %) male children. Gender incidence predominantly showed female preponderance with male: female ratio of 1: 2 (Chart 1). The most common clinical presentation of sacrococcygeal teratoma in our study was sacrococcygeal mass. One case showed associated rectovaginal fistula. Another case presented with urinary retention and constipation due to intrapelvic extension. On macroscopic examination, most (60%) of the cases were solid- cystic followed by cases with solid component (30%) and cystic component (10%) (Table 4). Some cases of mature teratoma showed completely differentiated organs e.g. limb like structures (Figure 1a), intestinal loops (Figure 1b) etc. In the present series, majority of cases on histopathological evaluation were documented as mature teratomas constituting about 86 % (19/22) followed by immature teratomas with 9 % (2/22). One case of malignancy (5%) with yolk sac differentiation was reported in our study which also correlated with high levels of serum AFP.

Table 1. Histopathological diagnosis and grades of Sacrococcygeal teratomas

Tumor maturity	Tumor grade	No. of cases
Mature	Grade 0	19(86%)
Immature	Grade 1	2(9%)
	Grade 2	1(5%)
	Grade 3	-
Total		22(100%)

Table 2. Classification and comparison according to Altman classification

Altman classification	kouranloo *et al*. (2006)	Present study
Type I	15(58%)	17(77%)
Type II	10(38%)	4(18%)
Type III	1(4%)	1(5%)
Type IV	0	0

Table 3. Multiparametric comparision of Sacrococcygeal teratoma with other studies

Parameters	William (1970) *et al*.	Valdiserri (1981) *et al*.	Khanna (1987) *et al*.	Present study
Total cases	103	68	41	22
Age incidence	< 2 month: 79 (76%)	< 2 month: 54 (79%)	< 2 month: 23(56%)	< 2 month: 18(82 %)
Male:female	1: 4	1: 4	2:1	1: 2
Clinical presentation	Sacrococcygeal mass	Sacrococcygeal mass	Sacrococcygeal mass	Sacrococcygeal mass
Tumor maturity	Mature: 73 (70%) immature: 30 (29 %)	Mature: 51 (75%) immature: 08 (12 %) malignant: 09 (13 %)	Mature: 31(76 %) immature: 05(12 %) malignant: 05(12 %)	Mature: 19 (86 %) immature: 3(14 %)

Table 4. Macroscopic Examination

Macroscopy	Present study	Ramani *et al*.(2013)
Solid- cystic	60%	40%
Solid	30%	40%
Cystic	10%	20%

Table 5. Percentage distribution of various microscopic components of SCT

S.No.	Components	Cases/total cases	Percentage
1	Ectodermal derivatives	22/22	100%
2	Endodermal derivatives	22/22	100%
3	Mesodermal derivatives	19/22	86%
4	Neuroepithelial component	18/22	82%

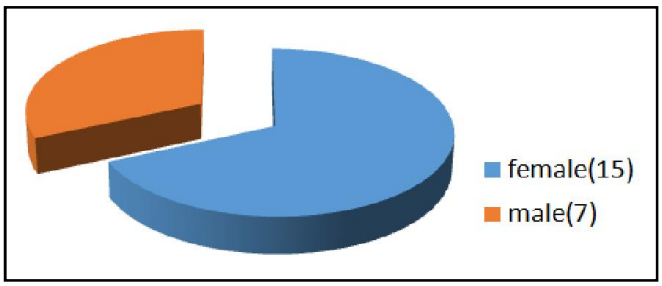


CHART 1: SEX incidence of the cases of SCT showing female preponderace

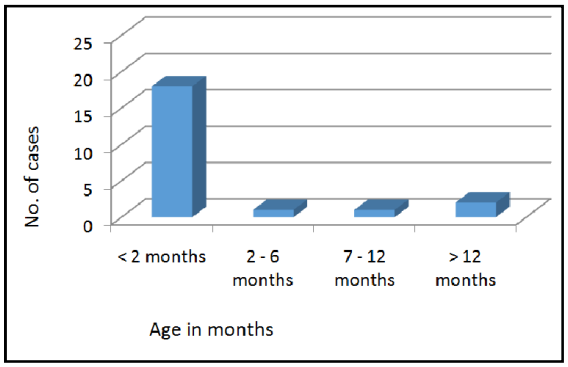


CHART 2: AGE incidence of the cases of SCTwith highest incidence in <2 months of age

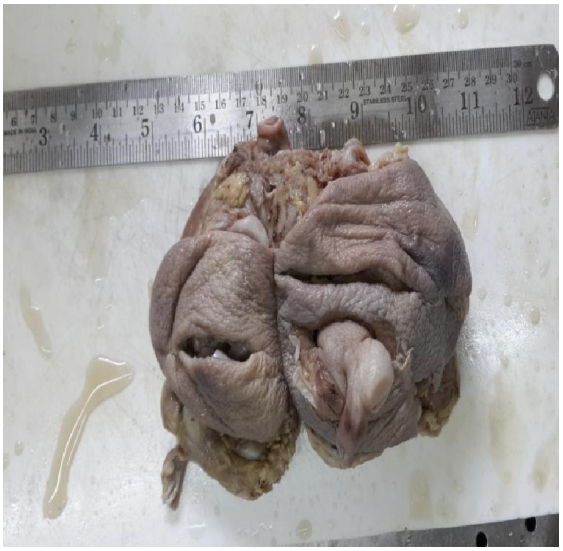


Fig 1a Gross photograph of resected sacrococcygeal mass

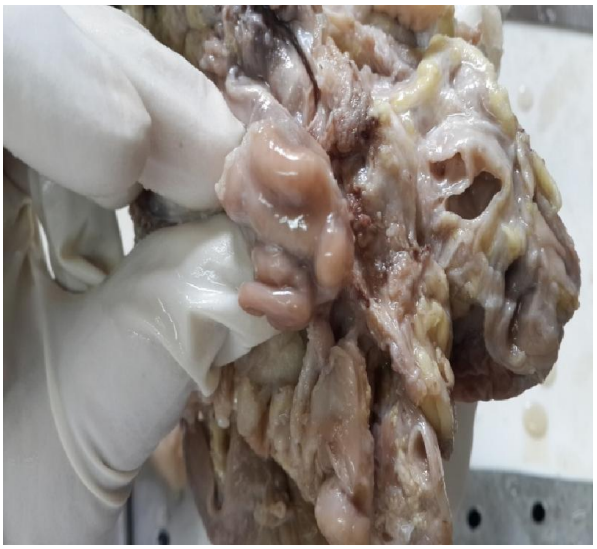


Fig 1b Gross photograph showing well differentiated intestinal loops

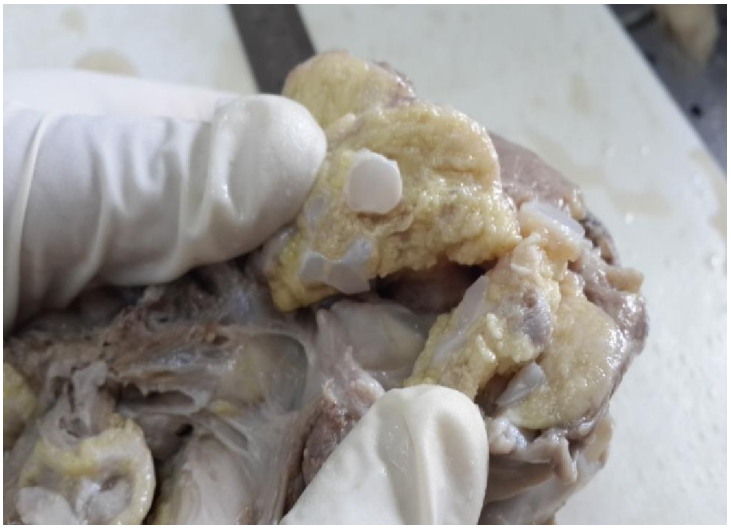


Fig 1c - Gross photograph showing cartilaginous differentiation (mesodermal derivation)

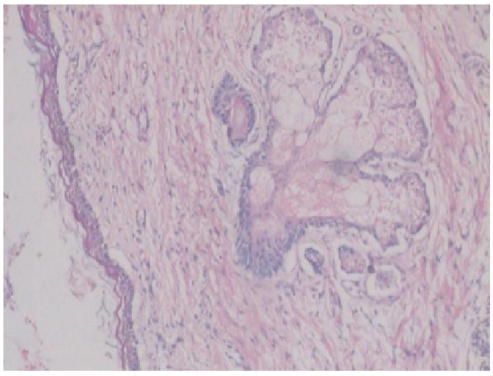


Fig 2a - Photomicrograph showing mainly ectodermal derivatives (squamous epithelium with pilosebaceous unit) (H&E,x20)

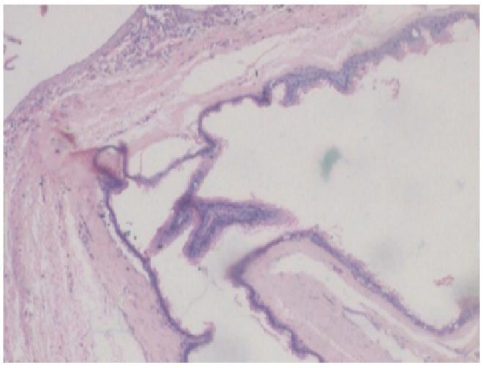
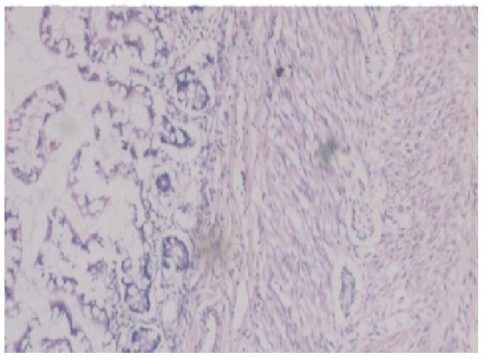


Fig 2b - Multicystic area comprising many cysts with papillae lined by secretory columnar epithelium (H&E,x20)



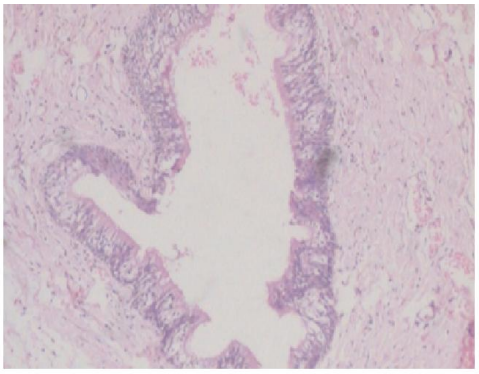


Fig 2c & 2d - Showing endodermal derivatives such as intestinal mucosa (H&E, X20)

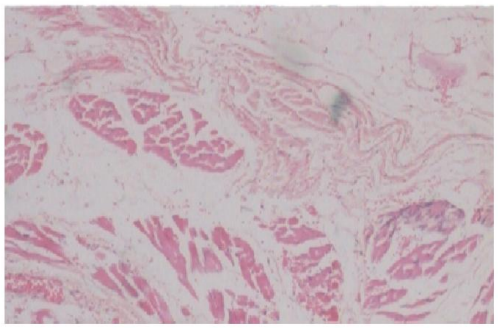


Fig 2e - Photograph of muscular and fibroadipose tissue (H&E,x20)

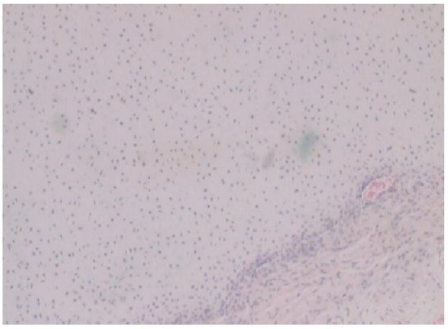


Fig 2f – Photograph of immature cartilaginous differentiation (H&E, x20)

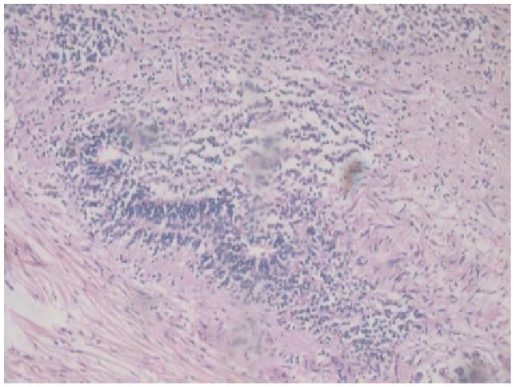


Fig 2g - Showing primitive neuroectodermal tissue in the form of rosettes. (H&E,x20)

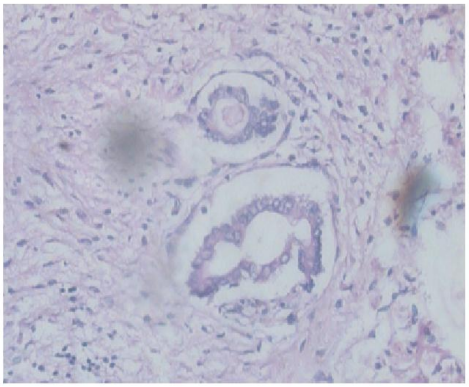


Fig 2h – Photograph showing reticular or microcystic areas formed by a loose meshwork lined by flat or cuboidal cells, pseudopapillary processes with central vessels (yolk sac diff.)

Mature sacrococcygeal teratomas showed the components derived from all the three germ cell layers with complete differentiation towards ectodermal, endodermal and mesodermal tissue (Fig 2a–2e). However immature teratomas showed immature non-malignant neuroepithelial tissue such as immature cartilage (Fig 2f) and neuroectodermal rosettes (Fig 2g). All the 19 cases of mature sacrococcygeal teratoma were benign and classified as grade 0. While 2 cases of immature teratoma were classified under grade 1 and one case with malignant yolk sac differentiation (Fig 2h) (Table 1). In present study, microscopy showed predominance of endodermal and ectodermal components (100%) followed by mesodermal(86%) and neuroepithelial components (82%) (Table 5).

DISCUSSION

A retrospective study of 22 patients presented with sacrococcygeal mass has been done, results were noted and comparison with other studies has been undertaken as follows.

Site

Majority of sacrococcygeal teratomas at birth present as a visible mass in the sacrococcygeal region (Robbins and Cotran 9th edition). Various sites of childhood teratoma includes sacrococcygeal region, testis, ovaries, and midline locations such as the mediastinum, retroperitoneum and head and neck (Robbins and Cotran 9th edition). Presentation of sacrococcygeal teratoma in present study was sacrococcygeal mass in most of cases. Almost all the cases of mature teratoma presented with mass at sacrococcygeal region, while immature teratomas presented with a visible to ill-defined mass at sacrococcyx. Our findings were similar to other studies (Valdiserri and Yunis, 1981; Williams *et al*., 1970; Khanna *et al*., 1987) (Table 3) Sacrococcygeal teratomas have been classified into four groups (Altman classification) depending upon the location of the tumor mass (Sacrococcygeal teratoma)

Type I - is completely external, evident at birth, and more easily resected or surgically removed. Type I does not typically spread.

Type II - has external and internal components. The internal portion is confined to the pelvic region. This type will spread in about 6% of cases.

Type III - also has external and internal components but the internal portion extends into the abdominal area. These types, II and III, are also evident at birth but the resection may be more difficult requiring access both from the back of the baby and from the front. Type III will spread in about 20% of cases.

Type IV - is completely internal. For this reason it may go undiagnosed for some time. Later, symptoms may develop that warrant an investigation and at this time the diagnosis is made. This type will spread in about 8 % of cases.

In present study, 77% of cases are of type I, 18% are of type II, 5% are of type III and type IV cases are nil compared with results of study done by Kouranloo *et al*. (Table 2). Altman RP *et al* have classified the size of SCTs as follows. Small (2 to 5 cms diameter), moderate (5 to 10cms in diameter) and large (>10cms diameter). Larger tumors are more likely to have immature histology and greater intraoperative blood loss, although some authors believe that the size of SCT is independent of its biological behavior (Keslar *et al*., 1994)

Signs and symptoms

Majority of sacrococcygeal teratomas at birth present as a visible mass in the sacrococcygeal region (Robbins and Cotran 9th edition). Most of the neonates do not have any symptoms though some may require intensive care because of mass effects on the surrounding organs e.g. abdominal pain, urinary retention and constipation (Ramani *et al*., 2013). In present study, one case presented with signs and symptoms of urinary retention and constipation.

Age/Sex

Congenital sacrococcygeal teratoma shows female preponderance (Rosai and Ackermann 10th edition). Age at the presentation is prognostically important as those who presented after age of 2 months are having increased chances of immature teratoma (Rosai and Ackermann 10th edition). In the present study, the most common age of presentation was below 2 months of age (chart 2). The male: female ratio was 1:2 indicating female preponderance (Chart 1). Our findings are consistent with available literature (Valdiserri and Yunis, 1981; Williams *et al*., 1970; Khanna *et al*., 1987) (Table 3)

Associated anomalies

Anorectal malformations are the most common anomalies associated with sacrococcygeal teratomas (Rosai and Ackermann 10th edition). In present study, one case is associated with recto-vaginal fistula.

Radiology

Radiological modalities such USG, CT scan and MRI imaging are reliable and helpful in diagnosis as well as clinical typing, which can add to the initial preoperative assessment in determining the anatomic relation of the tumor and the degree of trans-spinal tumor extension.

Clinical and radiological differential diagnosis

Various differential considerations include anterior sacral meningocele, rectal or anal duplication cyst, anal gland cyst,seroma, urinoma, tail gut cyst (retrorectal cystic hamartoma), chronic retrorectal abscess, pilonidal or dermoid cyst, soft tissue or bone tumours, osteomyelitis of the sacrum, chordoma, neurofibroma, fibrosarcoma, giant cell tumour or sacrum, postinjection granuloma and tuberculosis (Kostas *et al*., 2006). MRI should be clearly distinguishing meningomyelocele from SCT, which shows cystic lesion at a higher spinal level compared to lower sacral location of the SCT. In the present study, in a case of rectovaginal fistula, flouroscopy highlights the fistulous tract connecting rectum with the vagina as contrast promptly fills the vagina. Kouranloo *et al*. recommended that a preoperative CT is unnecessary in the neonate, but it is recommended in recurrent tumors and to rule out the presence of distant metastases (Kouranloo *et al*., 2006).

Serum markers

AFP and Beta-HCG are two most important markers of yolk sac differentiation and choriocarcinoma respectively (Rosai

and Ackermann 10th edition). Teratomas with yolk sac differentiation can be correlated with elevated levels of S. AFP which is a reliable indicator of tumor recurrence (Kouranloo *et al*., 2006). In present study, a single case with yolk sac differentiation (immature teratoma grade II) can be correlated with elevated (>1000ng/ml) S.AFP levels.

Antenatal diagnosis

Type I and II SCTs are commonly diagnosed by prenatal sonogram in the 24th – 34th weeks of gestation. The presence of a heterogeneous, well circumscribed exophytic mass at the caudal end of the fetus is pathognomonic. Even large type III and IV tumors can be diagnosed prenatally (Shalini Sinha *et al*., 2013). It was not carried out in the present study.

Macroscopic examination

On macroscopic examination, majority of mature teratomas were predominantly cystic, while the remaining cases were solid to cystic and immature teratomas were solid (Ramani *et al*., 2013). In present study, 60% cases were solid- cystic while remaining 30% were solid and 10% were cystic compared with study done by Ramani *et al*. (2013) (Table 4). Some cases of mature teratoma showed completely differentiated organs e.g. limb like structures (Figure 1a), intestinal loops (Figure 1b) etc.

Microscopy and histopathological grading

On microscopy, major components of mature teratoma were ectodermal and endodermal tissues followed by mesodermal elements. All the immature teratomas in our study were composed of neuroepithelial elements (Rosai and Ackermann 10th edition). In the present study, all cases of mature teratoma showed ectodermal (Fig 2a – 2b), endodermal (Fig 2c – 2d) and mesodermal (Fig 2e) differentiation. Cases of immature teratoma showed immature neuroepithelial elements (Fig 2g). The grading of sacrococcygeal teratoma is according to the presence of immature tissues (Roopali *et al*., 2016; Gonzalez Crussi *et al*., 1978). According to this grading, the cases are classified into grade 0, 1, 2 and 3.

Grade 0 tumours - contained mature tissues and were considered benign.
Grade 1 tumours - embryonal tissue was absent or present in one rare low magnification field within the tumour.
Grade 2 tumors - more than one, but less than four low-power foci of embryonal tissue in any one slide.
Grade 3 tumors - had individual sections with four or more low-magnification fields of immature tissue.

Grading of sacrococcygeal teratomas doesn't appear to correlate directly with prognosis (Valdiserri and Yunis, 1981). If the immature element is restricted to neuroectodermal components, the tendency is toward spontaneous differentiation. As a result, the behavior of this type of immature teratoma is usually benign, although occasional cases will recur or metastasize (Rosai and Ackermann 10th edition). In the present study, on histopathology, majority of sacrococcygeal teratomas are mature followed by immature teratomas. Our findings confirm the results by other studies (Valdiserri and Yunis, 1981; Williams *et al*., 1970; Khanna *et al*., 1987) (Table 3) In a study (Valdiserri and Yunis, 1981), 51(86%) cases of mature sacrococcygeal teratoma were classified as grade 0. Among 8 immature teratomas, 2(4%) were grade 2 and 6(10%) were grade 3. While in our study 19(86%) cases of mature sacrococcygeal teratoma were classified as grade 0 and among 3 cases of immature teratoma, 2(9%) were of grade 1 and 1(5%) was of grade II with yolk sac differentiation. Mature SCTs showed the components derived from all the three germ layers with completed differentiation. We observed predominance of ectodermal and endodermal derivatives followed by mesodermal structures. In ectodermal derivatives, we found skin, hair follicles and adnexal structures most commonly. The varying sized cysts lined by epithelium, the gastrointestinal columnar epithelium were the most common endodermal components observed in our study. Both ectodermal and endodermal components are present in 100% cases in the present study (Table 5). In mesodermal components, muscles, fat, connective tissuewere noted predominantly present in 86% cases. Neuroectodermal with astrocytes, neuronal fibers was predominantly seen in most of the cases. In immature teratoma, foci of immature nonmalignant neuroepithelium with rosette and yolk sac differentiation were evident. Neuroepithelial components were reported in 82% cases in the present study. The spectrum and percentage of various microscopic components of SCT in present study (Table 5) can be correlated with the statistics of a study done by Roopali d. Nikumbh *et al*. (2016).

Treatment

Total excision is curative treatment measure, the tip of the coccyx should be removed as part of the operation to prevent the reccurences (Rosai and Ackermann 10th edition). The surgical approach is to be through the sacral area or a combined abdominosacral approach. Another treatment measure can be surgery followed by chemotherapy/ radiotherapy which is reserved for malignant cases or inoperable cases (Kouranloo *et al*., 2006).

Disease recurrence

SCT, although histologically benign, has an alarming potential to recur either as a benign or malignant tumor during the first 3 years of life, therefore, a close follow up for at least 3 years (physical examination serum α-fetoprotein and diagnostic imaging) is recommended for all patients who have undergone excision of SCT (Kouranloo *et al*., 2006). None of the cases in the present study showed disease recurrence compared to Kouranloo *et al*. which stated that eight patients (30%) developed disease recurrence.

Conclusion

Sacrococcygeal teratoma is the most common congenital neoplasm presenting at birth as a sacrococcygeal mass with the female preponderance. Many of them are presenting with associated malformations or constipation and urinary retention due to mass effects. When diagnosed early at birth, majority of them are of benign mature type so that meticulous search for immature or malignant component has to be undertaken. However histological grading on the basis of presence of immature tissue predominantly neuroepithelial does not correlate directly with the prognosis but early diagnosis is helpful in therapeutic decisions, providing better outcome. Thus, histopathological evaluation of mass resected in cases of SCTs is of paramount importance in the treatment as well as prognosis of patients.

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