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Original Research Article

HISTOLOGICAL AND EMBRYONIC OVERVIEW OF SACROCOCCYGEAL TERATOMA: A SIX YEAR RETROSPECTIVE STUDY

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ABSTRACT

Background: Sacrococcygeal teratomas are tumors that arise from sacrococcygeal region containing tissue from all the three germ layers. Sacrococcygeal teratoma (SCT) is a relatively uncommon tumor affecting neonates, infants and children with an incidence of 1 in 40,000 live births with malignant transformation with increasing age. It is four times more common in females than males.

Aim and Objectives: To evaluate histological features of congenital sacrococcygeal teratoma and to grade them. Add a note on embryological and clinical aspect of sacrococcygeal teratoma.

Material and Methods: The present study was retrospective, descriptive analysis of 10 patients presented with sacrococcygeal mass over the period of 6 years from December 2009 to December 2015 in Department of Anatomy in KIMSU, Karad and Hi-Tech diagnostic center, Dhule. The cases were reviewed histologically and grading of SCT was done and comparisons with other studies were made.

Results: We reviewed 10 patients of congenital SCTs. There were 07 girls and 03 boys with the age ranges from 2 months to 18 months. Majority of the cases were less than 3 months. There were 08 cases of mature and 02 cases of immature teratoma noted. The most common presentation was sacrococcygeal mass in both teratomas.

Conclusion: Majority of the SCTs are benign on histology, when diagnosed at birth with fully differentiated tissues. Histologically meticulous and careful search for immature or malignant components should be instituted as it will helps in therapeutic decisions. Additionally embryological studies and chromosomal analysis of SCTs in large series may be warranted to predict the further prognosis.

KEY WORDS: Sacrococcygeal Teratoma, Congenital, Histology, Mature, Immature.

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INTRODUCTION

Teratomas are formed when germ cell tumors arise from embryonic components. The name is derived from the Greek word "teratos" which literally means "monsters". The ending "oma" denotes a neoplasm [1]. The incidence of congenital tumors occurring in the fetal sacrococcygeal teratoma (SCT) are still rare, having an incidence calculated at 1 in 40,000 live births [2], having female preponderance with

80% of cases or a 4:1 female to male ratio[1].The first description of teratoma cases were documented as far back as 2000 BC [3].

Teratomas are germ the germ cell tumors composed of tissue elements foreign to the organ/ anatomic site of origin [4]. In 1863,the word teratoma coined by Virchow[5].The more recent and more appropriate definition is an encapsulated tumor with tissue/organ components that can be traced to derivatives of the three primordial germ layers; ectoderm, mesoderm and endoderm. The most common locations of teratomas in childhood are the sacrococcygeal, mediastinal, retroperitoneal and gonadal organs [4].

Broadly, SCT are classified as mature, immature and malignant. Most cases being benign (mature) reported upto 79% by some studies [6]. Mature or benign teratomas are chiefly composed of differentiated tissue. Immature are characterized by the presence of immature non-malignant tissue, but the incidence of immature are very low (18%) with malignant tumors as yolk sac tumor, choriocarcinoma and embryonal carcinoma etc [6]. In infants and children, the most common site of occurrence of teratomas is sacrococcygeal region. These tumors may grow posteriorly and presented as external protrusion or dissect anteriorly distorting the regional organs like rectum, vagina and bladder without invading them [6,7].

The purpose of the present study is to evaluate clinicohistological features of congenital SCTs, grade them and to compare with other studies in view of its rarity

MATERIALS AND METHODS

The present study was retrospective, descriptive analysis of 10 cases of sacrococcygeal mass operated in KIMSU, Karad and Hi-Tech diagnostic center, Dhule over a period of 6 years from December 2009 to December 2015. Detailed gross and microscopical examination of well formalin fixed specimen with histologically confirmed cases of SCT were included in the present study from retrospective review. Improperly fixed, autolysed specimens were excluded from the study. Total of 10 specimens were studied. Clinicohistological studies along with grading of SCT were made, data was analyzed and findings were compared with other studies.

RESULTS

In the present study , total of 10 cases of histologically proven SCTs were evaluated. These tumors frequently occurred in age group of 0-18 months with 06 out of 10 cases presenting in age group of < 2 months. There were 07 females and 03 males with female preponderance (M:F ratio 1:2.3) . Our findings were comparable with J Kouranloo et al[8], Hashish A et al[9],Sinha S et al [10] and Ramani M et al[11].(Table 1)

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Table 1: Comparison of different studies with present study.

Study by Parameters	J Kouranloo et al [8]	Hashish A et al [9]	Sinha S et al [10]	Ramani M et al [11]	Present study
Total no of cases	26	35	10	25	10
Period of study(years)	1986-2000(14)	1998-2008(10)	1998-2012(14)	2007- 2012(5)	2009- 2015(6)
Age incid.(months)	<36	<12	<01	<18	<02
Gender (M:Fratio)	1:3	1.7:4	2.3:1	1:4	1:2.3
Clinical manifestation	Pre-sacral mass	Sacrococcygeal mass	Sacrococcygeal mass	Sacrococcygeal mass	Sacrococcygeal mass
Tumor maturity(cases)	Mature-21, Immature-5	Mature-27, Immature-7	Mature-08, Immature-02	Mature-20, Immature-5	Mature-08, Immature-02

In our study, the most common clinical presentation of SCT was sacrococcygeal mass. All the cases of SCT presented with prominent mass at sacrococcyx (Fig 1a,1b). Grossly ,majority of the specimen showed mixed features with predominance of cystic areas(80%) in mature teratomas

(Fig 1c,1d and 1e).The immature teratomas showed predominance of solid (20%) appearance with tiny foci of cysts (Fig 1f).

Altman RP et al [2] proposed a classification based on tumor mass location and extension; however some researchers regard such classification as useful only for description, having no prognostic value [12-14].The classification currently used by the American Academy of Pediatrics Surgery Section (AAPSS) is based on the Altman classification system[15].

Type I SCT- is a completely external mass (most common) representing 85% of benign tumors.

Type II SCT-has both internal and external components.

Type III SCT-is mostly internal

Type IV SCT-is a completely internal mass.

The last group is least common and seems to be associated with higher rates of malignancy from 5-20% [15].

The distribution according to Altman classification[2] was discussed (Table 2).The most common type seen was Type I (7/10) followed by Type II (3/10).There were no type III or IV SCTs in our study. Altman RP et al[2] have classified the size of SCTs as follows. Small (2 to 5 cms diameter), moderate (5 to 10cms in diameter) and large (>10cms diameter).Going by this classification, we had majority of small (7/10) followed by moderate (3/10) size tumors. We don't have the large size tumor in our study. 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[12].

Table 2: Altman classification of SCT.

Altman classification	No of cases	Percentage (%)	Comparison with J Kouranloo et al [8]
I	07	70	15(57.6)
II	3	30	10(38.6)
III	--	--	1(3.8)
IV	--	--	--

Histological grading of SCTs (Gonzalez-Crussi F et al) from grade 0-3is based on presence or absence of immature neural elements and their quantities [16]. In the present series, majority of cases on histological evaluation were documented as mature teratoma constituting about 80%(8/10) followed by immature teratoma20% (2/10).We don't encounter any case of malignancy in our study (Table 3).

Table 3: Histological diagnosis and grade of tumor.

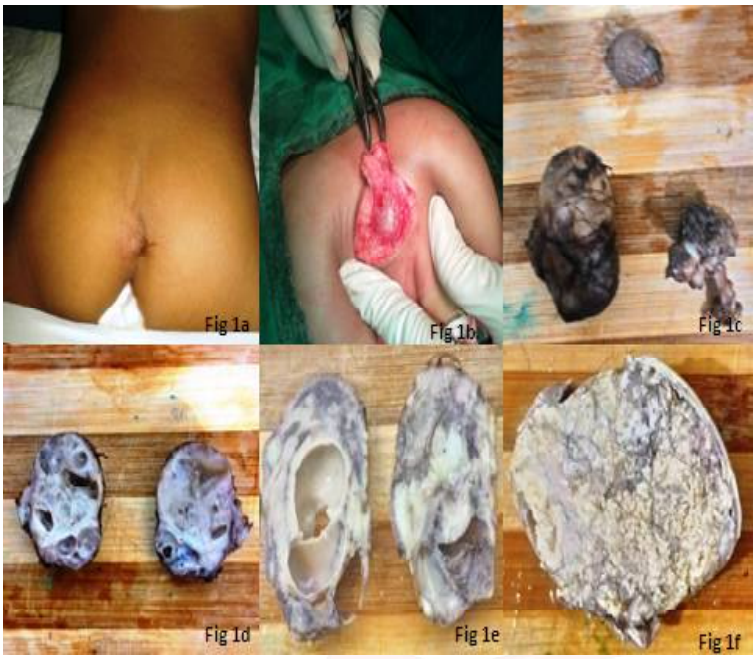
Tumor maturity	Tumor grade	No of cases (%)
Mature	**Grade 0** (Tumor contains only mature tissue)	**08(80)**
Immature	**Grade I** (Rare foci of immature tissue)	**02(20)**
	Grade II (Moderate quantities of immature tissue)	**----**
	Grade III (Large quantities of immature tissue like malignancy)	**----**
	Total	**10(100)**

Mature SCTs showed the components derived from all the three germ layers with completed differentiation. We observed predominance of ectodermal and endodermal derivatives (100%) followed by neuroectodermal (90%) and organoid structures (60%) (Table no.4). In ectodermal derivatives, we found skin, hair follicles and adnexal structures (Fig 2a) most commonly. Neuroectodermal (Fig 2e) with astrocytes, neuronal fibers was predominantly seen in all cases. In mesodermal components, fat, connective tissue (Fig 2d) noted predominantly. The varying sized cysts lined by epithelium, thyroid follicles (Fig 2b) and the gastrointestinal columnar epithelium, transitional epithelia (fig 2c) were the most common endodermal components observed in our study. The pancreas (Fig 2f) and the thyroid gland are the most common, organoid endodermal derivatives in present study. In immature teratoma, foci of immature nonmalignant neuropepithelium with rosette were evident.

Table 4: Components of mature SCT.

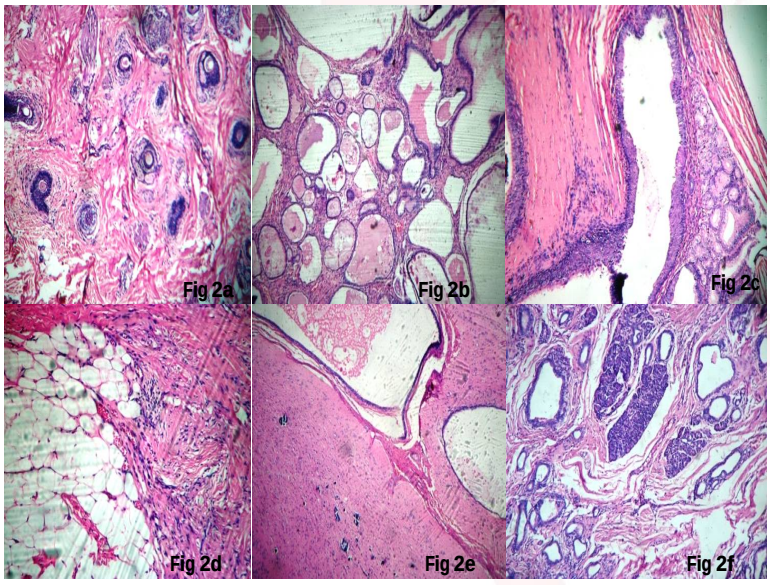
Sr. no	Components	Total cases	Percentage
1	Ectodermal derivatives	10	100
2	Endodermal derivatives	10	100
3	Mesodermal derivatives	9	90
4	Neuroepithelium	9	90
5	Organoid structures	6	60

Fig. 1: Gross description of mature SCT.



a) Gross photograph of a child presented with sacrococcygeal mass
b) Intra-operative photograph of resection of sacrococcygeal mass
c) Gross specimen of resected SCT with segment of coccyx
d) Cut section of SCT showing mixed pattern with predominance of cystic areas
e) C/S of other case showing predominance of multicystic areas
f) Tumor with predominant solid areas

Fig. 2: Histology of the mature SCT.



a) Photomicrograph showing mainly ectodermal derivatives as skin ,hair follicles and adnexal tissue (H&E,x400)
b) Multicystic area comprising many cysts lined by epithelium, thyroid follicles with colloid(H&E,x400)
c) Mesodermal tissues as GIT columnar mucosa, glands and transitional epithelium of bladder (H&E,x400)
d) Photomicrograph of connective tissue and fibroadipose tissue (H&E,x400)
e) Neuroepithelium and cyst lined by epithelium (H&E,x400)
f) Organoid structures mainly pancreatic acini noted (H&E,x400)

DISCUSSION

Sacrococcygeal teratoma is a relatively uncommon tumor in the neonatal period. When SCT is present at birth excisional surgery should be performed as early as possible to prevent malignant transformation, which may occur with advancing age of the patient [17].The surgical approach of the SCT consist of complete removal of the tumor through the sacral area or a combined abdominosacral approach with removal of coccyx to avoid recurrence rate[18].Majority of SCTs at birth presented as visible mass in the sacrococcygeal region. Most of the neonates don't have any symptom though some may require intensive care due to prematurity, high cardiac failure and rupture of tumor or bleeding within the tumor. Those neonates having lesions with intrapelvic components may present with urinary obstruction, constipation and abdominal mass [19].

Embryology: Sacrococcygeal teratomas have tissue derived from ectoderm, mesoderm and endoderm. Although ,their embryonic origin is still uncertain[7].They are believed to arise early in gestation (at around second/early third week) from the totipotent cells of Hensen's node(also called the primitive knot), a remnant of the

primitive streak in the coccygeal region [20-22].The primitive streak appears as a linear thickening in the ectoderm at the caudal edge of the bilaminar embryonic disc[7].It usually diminishes in size eventually disappearing after undergoing degenerative changes. As the mesoderm rapidly proliferates, the primitive streak comes to lie more and more caudally, where the remnant of Hensen's node descends to the tip or anterior surface of coccyx [20-22]. Growth of these primitive pluropotential cells escapes the control of embryonic inducers and organizers resulting in a teratoma [23]. Rearrangement within the proto-oncogene or in a regulatory sequence result in a molecular transformation of cells foreign to the anatomical site [23].

Thus, origin of SCT often occurs near the coccyx, where the greatest concentration of primitive cells exists for the longer period of time.SCT is formed from multiple neoplastic tissue that lack organ specificity, foreign to sacrococcygeal region, are derived from all three germ layers[23].

The female preponderance was noted by published series. This is also proven true for our case series, albeit the female to male ratio of 2.3:1.The most common age of presentation was below 3months of age. Our findings were consistent with available literature[8-11]. Regarding Altman distribution of SCT, most common type was Type I(70%) followed by Type II(30%) in our series.This was comparable with J Kournaloo et al. [8].

In our series, regarding gross features, mixed type with predominance of cystic areas (80%) was noted in mature teratomas whereas immature with solid (20%) appearance was evident. This findings were consistent with previous studies as by Ramani M et al. [11].On histology, majority (80%) of SCTs were mature followed by immature or grade I(20%).We don't found grade II and III tumors in our series. Our finding were collaborated with results of other studies [8-11].In our study, we found predominant components of mature teratoma from ectodermal and endodermal derivatives followed by mesodermal, neuroectodermal and organoid endodermal elements. Our findings were consistent with study by Ramani M et at. [11] studied in 2013.We diagnosed skin and hair follicles were most common components in ectodermal derivatives. In endodermal one, we found gastrointestinal epithelium, urothelium and respiratory tract epithelium respectively. Fat, connective tissue, cartilage, bone, and muscle with lymphoid tissue encountered in mesodermal elements. Pancreas mainly with parotid and salivary was diagnosed in organoid endodermal derivatives. Neuroepithelium was noted in all mature teratomas, whereas immature nonmalignant neuroepithelium was noted in immature teratomas in form of rosettes of primitive elements.

The differential diagnosis of SCTs include meningiocele, myelomeningiocele, and fetus in fitu (FIF)[17,24]. Histology is the gold standard for differentiation of above entities. The presence of axial skeleton with vertebral axis on gross and microscopy goes more in favor of FIT over teratomas [24].

CONCLUSION

Sacrococcygeal teratoma is a relatively uncommon tumor in the neonatal period. When SCT is present at birth, excisional surgery should be performed as early as possible, to prevent malignant transformation, which may occur with the advancing age. Histology is the gold standard for separation of mature/differentiated from immature or malignant components as it helps in therapeutic decisions.

Conflicts of Interests: **None**

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