CONGENITAL SACROCOCCYGEAL TERATOMAS IN CHILDREN - A PATHOLOGISTS OVERVIEW
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ABSTRACT: BACKGROUND: Sacrococcygeal teratoma is the most common congenital neoplasm and accounts for 35% to 60% of all the teratomas. It has an incidence of 1 per 30,000 to 40,000 live births with an increased prevalence of 3 to 4 times in female children. Sacrococcygeal teratomas are derived from all the three germ cell layers. Majority of the cases present at birth as a visible mass in the sacrococcygeal region. Age is an important predictor of malignancy in sacrococcygeal teratoma. Early diagnosis influences clinical decision and management, providing better outcome. AIMS AND OBJECTIVES: To evaluate clinicopathological features of Congenital Sacrococcygeal Teratomas and grade them. MATERIAL AND METHODS: We analysed 25 patients with Congenital Sacrococcygeal Teratomas presenting at Department of Pathology, Paediatric referral hospital from 2006 to 2012. Cases were reviewed and graded in a manner similar to grading of Ovarian Teratomas. RESULTS: We reviewed 25 patients with congenital sacrococcygeal teratoma. There were 20 girls and 5 boys with the age ranges from 1 day to 6 years. Majority of the cases (72%) were seen in the age group below 1 month of age. There were 20 cases of mature and 5 cases of immature teratoma. The most common clinical presentation was sacrococcygeal mass in both and mature and immature teratomas. CONCLUSION: Sacrococcygeal teratomas when diagnosed at birth reveal fully differentiated tissues and are benign on histopathology. Meticulous search for immature or malignant component should be instituted as it helps in therapeutic decisions. Histopathological grading of immature tissue in sacrococcygeal teratomas does not correlate directly with prognosis. KEYWORDS: Congenital Sacrococcygeal Teratoma, Mature teratoma, Immature teratoma.

INTRODUCTION: Sacrococcygeal teratoma is the most common congenital neoplasm and accounts for 35% to 60% of all the teratomas [¹]. It has an incidence of 1 per 30,000 to 40,000 live births with an increased prevalence of 3 to 4 times in female children [²].

Teratomas are considered to be among the oldest known tumours [³]. The first record of a sacrococcygeal teratoma was in the form of an inscription on a Babylonian cuneiform tablet which dated back to 600 B.C. Sacrococcygeal teratoma was first described in 1951 in a study which included forty infants and children [⁴]. In 1937, it was the first time when sacrococcygeal teratomas were elaborately reviewed [⁵].

Sacrococcygeal teratomas are derived from all the three germ cell layers. The uncertainty about the origin of sacrococcygeal teratoma still exists, though it is hypothesized that it arises from the totipotential cells of Hensen’s node, a remnant of primitive streak in the coccygeal region [⁶].

Broadly, sacrococcygeal teratomas are classified as mature, immature and malignant [⁶]. Mature teratomas are chiefly composed of differentiated tissues and considered benign. Immature
teratoma is characterized by the presence of immature non-malignant tissue. Teratomas with features of yolk sac tumour, choriocarcinoma or embryonal carcinoma among differentiated tissues are regarded as malignant [7].

Age is an important predictor of malignancy in sacrococcygeal teratoma. At the time of birth, the risk of malignancy is less than 10% while the risk rises to greater than 75% after the age of 1 year [1]. In infants and children the most common site of occurrence of teratomas is sacrococcygeal region. These tumours may grow posteriorly and present as external protrusion or dissect anteriorly distorting the regional organs like rectum, vagina and bladder without invading them [4].

In the present study we have attempted to evaluate the clinicopathological features of congenital sacrococcygeal teratomas and grade them.

MATERIAL AND METHODS: The present retrospective study was undertaken at Department of Pathology, Paediatric Referral Hospital for duration of 5 years from June 2007 to May 2012. During this period, a total 25 cases were diagnosed as congenital sacrococcygeal teratomas. Detailed history, physical examination, routine laboratory and radiological investigations which included ultrasound examination and computed tomography scan were done in all cases. Detailed gross and microscopic examination of all the specimens were done. Grading was done for all the cases of sacrococcygeal teratoma in method similar to grading of Ovarian Teratomas [6].

RESULTS: In the present study, a total of 25 cases diagnosed as congenital sacrococcygeal teratomas were evaluated. These tumours frequently occurred in the age group of 0 -1 month with 18 of 25 cases presenting in that age group (Figure 10). There were 20 girls and 5 boys. Gender incidence predominantly showed female preponderance with Male: Female ratio of 1: 4.

The most common clinical presentation of sacrococcygeal teratoma in our study was sacrococcygeal mass. All the cases of mature teratoma presented with prominent mass at Sacrococcyx (Figure 1A, 1B). 4 cases showed associated malformations, which included 3 cases with anorectal malformations and one case with genital malformation. Oozing from the tumour and surface ulceration was seen in 3 cases and 2 cases, respectively (Chart 2). Whereas cases with immature teratoma presented with visible to ill-defined mass at Sacrococcyx. One case presented with urinary retention and constipation due to intrapelvic extension. Genital malformation was associated with one case (Chart 3).

Grossly majority of mature teratomas were predominantly cystic (85%), while the remaining 15% were solid to cystic. An example of a mature cystic teratoma can be seen in the Figure 3A and 3B. All the immature teratomas were predominantly solid. Example of immature teratoma seen in the Figure 2A and 2B.

In the present series, majority of cases on histopathological evaluation were documented as mature teratomas (MT) constituting about 80 % (20/25) followed by immature teratomas (IMT) with 20 % (5/25). There were no cases of malignancy reported in our study. (Table 1) (Chart 1)

Mature sacrococcygeal teratomas showed the components derived from all the three germ cell layers with complete differentiation (Table 2). Ectodermal and endoderm derived tissues were seen in all the cases of mature teratoma(Figure 4A, 4B, 7A, 7B). Neural elements and Central nervous system elements like glial tissue (Figure 5A) and choroid plexus (Figure 5B) were documented in 95% of the cases. Mesodermal elements like adipose tissue, cartilage, smooth and skeletal muscle
bundles were appreciated in 90% of the cases (Figure 6A, 6B), while organoid elements like pancreatic (Figure 8A) and salivary gland tissue (Figure 8B) in 15%. However immature teratomas showed immature non-malignant neuroepithelial tissue in the form of neuroepithelial rosettes in all the cases (Figure 9A, 9B).

All the cases were graded in a manner similar to grading of Ovarian Teratomas. All the 20 cases of mature sacrococcygeal teratoma were benign and classified as Grade 0. While all the 5 cases of immature teratoma were classified under Grade 2. (Table 1)

**DISCUSSION:** Way back in 1841, a typical case of sacrococcygeal teratoma was described in detail under the name of congenital pelvic tumour. However in 1863, the term teratoma was introduced to designate a monstrous and malformed tumour[3].

Majority of sacrococcygeal teratomas at birth present as a visible mass in the sacrococcygeal region. Most of the neonates do not have any symptoms though some may require intensive care because of prematurity, high cardiac failure, disseminated intravascular coagulation and rupture of tumour or bleeding within the tumour. Those neonates having lesions with an intrapelvic component may present with urinary obstruction. Children present with constipation, urinary retention, an abdominal mass or symptoms of malignancy, like failure to thrive[4].

The grading of sacrococcygeal teratoma is according to the presence of immature tissues. Grading of sacrococcygeal teratoma is done in method similar to grading of Ovarian Teratomas[6]. According to this grading, the cases are classified into Grade 0, 1, 2 and 3.

Grade 0 tumours contain mature tissues and are considered benign. In Grade 1 tumours embryonal tissue was absent or present in one rare low magnification field within the tumour. Grade 2 if they had more than one, but less than four low-power foci of embryonal tissue in any one slide. Grade 3 teratomas 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, unlike that of ovarian teratomas where grading has direct correlation with prognosis[6].

In the present study, the most common age of presentation was below 2 months of age. The male: female ratio was 1:4 clearly indicating female preponderance. Our findings are consistent with available literature[6, 8, 9, 10, 11, 12]. (Table 4)

Clinical manifestations of sacrococcygeal teratoma in our series included sacrococcygeal mass in majority 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[6, 8, 13]. (Table 4)

A study reported that, grossly 40% of the sacrococcygeal teratomas were solid, 20% were cystic and 40% were mixed[14]. Whereas another series reported that mature teratomas were predominantly cystic and immature teratomas were partly solid and partly cystic[13]. According to a review, majority of benign sacrococcygeal teratomas were cystic in nature[15]. In our study majority of the mature teratomas were predominantly cystic while immature teratomas were mostly solid.

On histopathology, majority of sacrococcygeal teratomas were mature followed by immature teratomas. Our findings confirm the results by other studies[6, 14, 16, 17, 18, 19, 20]. (Table 4)

In this present study the major components of mature teratoma were ectodermal and endodermal tissues followed by mesodermal and organoid elements. All the immature teratomas in our study were composed of neuroepithelial elements. Another study showed similar findings in
mature teratoma except for the endodermal tissues which were third commonest. All cases of immature teratoma showed predominantly neuro-epithelial elements\(^{[13]}\).

In a study, 51 cases of mature sacrococcygeal teratoma were classified as Grade 0. Among 8 immature teratomas 2 were Grade 2 and 6 were Grade 3\(^{[6]}\). While in our study all the cases of mature sacrococcygeal teratoma were classified as Grade 0 and all the cases of immature teratoma were Grade 2.

**CONCLUSION:** Sacrococcygeal teratoma remains the most common congenital neoplasm presenting at birth they. When these tumours are diagnosed at birth they reveal fully differentiated tissues and are benign on histopathology. Meticulous search for immature or malignant component should be instituted as it helps in therapeutic decisions. Immature tissue in sacrococcygeal teratoma is predominantly neuroepithelial. Histopathological grading of immature tissue in sacrococcygeal teratomas does not correlate directly with prognosis. Early diagnosis influences clinical decision and management, providing better outcome.

**REFERENCES**


**Table 1: Histopathological diagnosis and grades of Sacrococcygeal teratomas.**

<table>
<thead>
<tr>
<th>TUMOUR MATURITY</th>
<th>TUMOUR GRADE</th>
<th>NO. OF CASES</th>
</tr>
</thead>
<tbody>
<tr>
<td>MATURE</td>
<td>GRADE 0</td>
<td>20</td>
</tr>
<tr>
<td>IMMATURE</td>
<td>GRADE 1</td>
<td>-</td>
</tr>
<tr>
<td></td>
<td>GRADE 2</td>
<td>05</td>
</tr>
<tr>
<td></td>
<td>GRADE 3</td>
<td>-</td>
</tr>
<tr>
<td>TOTAL</td>
<td></td>
<td>25</td>
</tr>
</tbody>
</table>

**Table 2: Components of Mature Sacrococcygeal teratomas.**

<table>
<thead>
<tr>
<th>COMPONENT</th>
<th>TOTAL CASES IT IS PRESENT</th>
<th>PERCENTAGE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ectodermal elements</td>
<td>20</td>
<td>100</td>
</tr>
<tr>
<td>Endodermal elements</td>
<td>20</td>
<td>100</td>
</tr>
<tr>
<td>Neural and CNS elements</td>
<td>19</td>
<td>95</td>
</tr>
<tr>
<td>Mesodermal elements</td>
<td>18</td>
<td>90</td>
</tr>
<tr>
<td>Organoid elements</td>
<td>03</td>
<td>15</td>
</tr>
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</table>
Table 3: Comparison of different studies with the present study.

<table>
<thead>
<tr>
<th></th>
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</thead>
<tbody>
<tr>
<td>TOTAL CASES</td>
<td>103</td>
<td>68</td>
<td>41</td>
<td>25</td>
</tr>
<tr>
<td>AGE INCIDENCE</td>
<td>&lt; 2 Month: 79 (76%)</td>
<td>&lt; 2 Month: 54 (79%)</td>
<td>&lt; 2 Month: 23 (56%)</td>
<td>&lt; 2 Month: 18 (72%)</td>
</tr>
<tr>
<td>MALE: FEMALE</td>
<td>1:4</td>
<td>1:4</td>
<td>2:1</td>
<td>1:4</td>
</tr>
<tr>
<td>CLINICAL</td>
<td>Sacrococcygeal mass</td>
<td>Sacrococcygeal mass</td>
<td>Sacrococcygeal mass</td>
<td>Sacrococcygeal mass</td>
</tr>
<tr>
<td>PRESENTATION</td>
<td>Mature: 73 (70%), Immature: 30 (29%)</td>
<td>Mature: 51 (75%), Immature: 08 (12%), Malignant: 09 (13%)</td>
<td>Mature: 31 (76%), Immature: 05 (12%), Malignant: 05 (12%)</td>
<td>Mature: 20 (80%), Immature: 05 (20%)</td>
</tr>
<tr>
<td>TUMOUR MATURITY</td>
<td>Mature: 73 (70%), Immature: 30 (29%)</td>
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<td>Mature: 20 (80%), Immature: 05 (20%)</td>
</tr>
</tbody>
</table>

FIGURES

Figure 1: (A) Clinical Picture of a case of Sacrococcygeal teratoma. (B) Another case of Sacrococcygeal teratoma.

Figure 2: Morphology of Sacrococcygeal teratoma (A) Gross specimen of Sacrococcygeal teratoma. (B) Cut section shows mainly solid and few cystic areas.
Figure 3: Morphology of Sacrococcygeal Teratoma (A) Gross specimen of Sacrococcygeal teratoma. (B) Cut section showing predominantly cystic areas.

Figure 4: (A) Photomicrograph showing Mature teratoma with cystic spaces lined by epithelium. (Haematoxylin and Eosin, 10X) (B) Photomicrograph showing Mature teratoma with ectodermal elements (Haematoxylin and Eosin, 10x)

Figure 5: (A) Photomicrograph of Mature teratoma showing glial tissue. (Haematoxylin and Eosin, 40X) (B)Photomicrograph of Mature teratoma showing papillary structures resembling choroid plexus of CNS (Haematoxylin and Eosin, 10x)
Figure 6: (A) Photomicrograph of Mature teratoma showing mesodermal elements. (Haematoxylin and Eosin, 10X) (B) Photomicrograph of Mature teratoma showing cartilage (Haematoxylin and Eosin, 10x)

Figure 7: (A) Photomicrograph of Mature teratoma showing cystic space lined by columnar epithelium and goblet cells. (Haematoxylin and Eosin, 40X) (B) Photomicrograph of Mature teratoma showing cystic space lined by respiratory epithelium (Haematoxylin and Eosin, 40x)

Figure 8: (A) Photomicrograph of Mature teratoma showing Pancreatoid tissue. (Haematoxylin and Eosin, 10X) (B) Photomicrograph of Mature teratoma showing salivary gland tissue. (Haematoxylin and Eosin, 40x).
Figure 9: (A) Photomicrograph of Immature teratoma. (Haematoxylin and Eosin, 4X) (B) Photomicrograph of Immature teratoma showing neuroepithelial rosettes. (Haematoxylin and Eosin, 10x).

CHARTS:

Chart 1: Age incidence of the cases of Sacrococcygeal teratoma.
Chart 2: Clinical manifestations of the cases of Mature Sacrococcygeal teratoma.

Chart 3: Clinical manifestations of the cases of Immature Sacrococcygeal teratoma.
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