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Original Article

An Epidemiological Review of Sacrococcygeal Teratoma over Five Years in a Tertiary Care Hospital

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Abstract

Background: Teratoma refers to neoplasm containing at least two germ cell layers derivatives foreign to the site of origin. Sacrococcygeal teratoma is most common congenital tumor commonly present as an exophytic mass of varying size at coccyx. **Aims and Objectives:** (1) The aim is to find age, sex, and clinical presentation of cases of sacrococcygeal teratoma. (2) Application of American Academy of Pediatrics' surgical section classification and histological grading in all cases. **Materials and Methods:** A retrospective, observational study was conducted in the Department of Pathology in collaboration with the Department of Pediatric Surgery from February 2009 to January 2014. A total 13 cases of sacrococcygeal teratoma were included in the present study. The records of these patients were reviewed, and clinical profiles were noted. An average 12 slides were examined in each case to evaluate histological type and grades. **Results:** A retrospective study was conducted including thirteen cases of histologically confirmed sacrococcygeal teratoma over 5 years period. Male to female ratio was 1:2. As per as age distribution is concerned, 3 neonates (23%) presented with sacrococcygeal mass. Associated congenital malformation was seen in 2 cases (15.4%). According to the American Academy of Pediatrics (Altman's) classification, 23% of cases were Type I, 31% of cases patients Type II, 31% of cases Type III and 15% were for Type IV. The significant presacral component was noted in all malignant tumors. **Conclusion:** Two clinical patterns were observed in sacrococcygeal teratoma related to the age of presentation. As sacrococcygeal teratoma has potential to become malignant, meticulous search for the malignant component is required for histopathological categorization.

Keywords: *Neuroepithelium, sacrococcygeal teratoma, Totipotential cell*

Introduction

Teratomas are composed of tissues derived from ectoderm, mesoderm, and endoderm. Sacrococcygeal teratomas are developed from the totipotential cells of primitive knot which is a remnant of the primitive streak in the coccygeal region.[1] With the incidence of 1/35000–1/40000 live birth, sacrococcygeal teratoma is considered as the most common germ cell tumor in the neonatal period and infancy.[2] Sacrococcygeal teratoma shows female preponderance with male to female ratio 1:4.[3]

Clinical presentation varies depending on the age of the patient and location of the tumor. In the neonatal period, tumor presents as exophytic mass at the sacral region with occasional surface ulceration and hemorrhage, but younger child presents with mass effect due to its enormous size and proximity to intra-abdominal organs.[4] Fetal tumors are diagnosed by prenatal ultrasound. Association with other congenital anomaly such as myelomeningocele and vertebral malformation were reported.[5]

As per as the histology is concerned, sacrococcygeal teratomas are classified into mature, immature and malignant category.[6] Biological behaviors are affected by age and sex. Mature teratomas are common in neonates.[7]

Clinical and pathological variables of cases of sacrococcygeal teratoma were evaluated in our study.

Materials and Methods

A retrospective, observational study was conducted in the Department of Pathology in collaboration with the Department of Pediatric Surgery from February 2009 to January 2014. A total 13 cases of sacrococcygeal teratoma were included in the study. The records of these patients

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with pediatric sacrococcygeal teratoma were reviewed for age, sex distribution, clinical presentation, preoperative investigation, mode of treatment, and follow-up. Pre-operative hematological and radiological evaluation were noted in all cases.

Depending on the radiological findings, the extent of the tumors was analyzed and categorized according to the American Academy of Pediatrics' surgical section classification which includes four categories:

1. Type I: Predominantly external with minimal presacral component;
2. Type II: Present externally but with significant intrapelvic extension
3. Type III: Apparent externally but predominantly a pelvic mass extending into the abdomen
4. Type IV: Presacral with no external presentation.[8]

Both size and appearance of the tumors were recorded thoroughly. Depending on the size, tumors were classified into small (2–5 cm diameter); moderate (5–10 cm diameter) and large (>10 cm diameter) tumors. Hematoxylin and eosin stained slides were reviewed in each case, and additional sections were from taken from paraffin block if needed. An average 12 slides were examined in each case. Depending on histological findings, tumors were classified broadly into mature, immature, and malignant tumors. Tumor showing differentiated tissues and tumor having immature neuroepithelium were considered to be mature and immature teratoma accordingly. Malignant tumors showed yolk sac tumor, choriocarcinoma, or embryonal carcinoma along with differentiated tissues.[9]

Histological grading was done:

i. Grade 0: Tumor shows only mature tissue
ii. Grade 1: Tumor shows rare foci of immature tissue <1 lpf/slide
iii. Grade 2: Tumor shows moderate quantities of immature tissues, 1–3 lpf/slide
iv. Grade 3: Tumor shows large quantities of immature tissue. >3 lpf/slide; with or without malignant yolk sac elements.

Result

In the present study, a total of 13 cases diagnosed as congenital sacrococcygeal teratoma were evaluated. The tumors presenting to us were mainly in the age group >2 months, with 10 out of 13 cases presenting in that age group. There were 9 female and 4 male children. Gender incidence showed female preponderance with male: female ratio of 1:2.

The most common clinical presentation of sacrococcygeal teratoma in our study was sacrococcygeal mass. All the cases of mature teratoma presented with a prominent mass at sacrococcyx. 2 cases showed associated malformations (anorectal malformations).

According to the American Academy of Pediatrics (Altman's) classification, 23% of cases were Type I, 31% of cases patients Type II, 31% of cases Type III and 15% were for Type IV. The significant presacral component was noted in all malignant tumors.

Majority of cases on histopathological evaluation were documented as mature teratoma, about 53.4% (7/13) followed by immature teratoma with 30.8% (4/13). Grossly, mature teratoma was predominantly cystic (75%), whereas the remaining 25% were solid and cystic. The immature teratoma was predominantly solid. Immature teratoma showed immature neuroepithelial tissue in the form of neuroepithelial rosette. Two cases of malignancy were reported in our study. Malignant yolk sac component was noted in both cases [Table 1].

Mature sacrococcygeal teratoma showed the components derived from all the three germ cell layers with complete differentiation. Ectodermal and endoderm derived tissues were seen in all the cases of mature teratoma. Neural elements, glial tissue, and choroid plexus were seen in 69.2%. Mesodermal elements such as adipose tissue, cartilage, smooth, and skeletal muscle were noted. Organoid elements such as pancreatic and salivary gland tissue in 15.4% [Table 2].

Discussion

Sacrococcygeal teratoma at birth usually presents as a visible mass in the sacrococcygeal region. Most of the neonates do not have any symptoms. Some may have cardiac failure, disseminated intravascular coagulation, and rupture or bleeding within the tumor. 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.[2] The present study mainly had neonates who presented with a

Table 1: Histopathological diagnosis and grades of sacrococcygeal teratomas

Tumor type	Tumor grade	Number of cases
Mature	0	7
Immature	1	1
	2	3
Malignant	3	2
Total		13

Table 2: Components of sacrococcygeal teratomas

Component	Total cases it is present (%)
Ectodermal elements	13 (100)
Endodermal elements	13 (100)
Neural and CNS elements	9 (69.2)
Mesodermal elements	8 (61.5)
Organoid elements	2 (15.4)

CNS – Central nervous system

Table 3: Comparison of different studies with the present study

Parameter	William *et al*.	Valdiserri *et al*.	Khanna *et al*.	Ramani *et al*.	Our study
Total cases	103	68	41	25	13
Age incidence (<2 month), (%)	79 (76)	54 (79)	23 (56)	18 (72)	9 (69)
Male:female	1:4	1:4	2:1	1:4	1:2
Clinical presentation	Sacrococcygeal mass	Sacrococcygeal mass	Sacrococcygeal mass	Sacrococcygeal mass	Sacrococcygeal mass
Tumour maturity (%)	Mature: 73 (70) Immature: 30 (29)	Mature: 51 (75) Immature: 8 (12) Malignant: 9 (13)	Mature: 31 (76) Immature: 5 (12) Malignant: 5 (12)	Mature: 20 (80) Immature: 5 (20)	Mature: 7 (53.4) Immature: 4 (30.8) Malignant: 2 (15.4)

mass at sacrococcyx. The older children presented with mass, pain, and constipation.

The grading of sacrococcygeal teratoma is done according to the presence of immature tissues. Grading of sacrococcygeal teratoma is done similar to the grading of an ovarian teratoma.[6] According to this grading, the cases are classified into Grade 0, 1, 2 and 3. Grading of sacrococcygeal teratoma does not correlate directly with prognosis, unlike that of ovarian teratoma where grading has direct correlation with prognosis.[6]

In our study, the most common age of presentation was above 2 months of age. The male: female ratio was 1:2, indicating female preponderance, as in other studies.[6,8,10-12]

Clinical manifestations of sacrococcygeal teratoma in our series included sacrococcygeal mass in the majority of cases. Immature teratoma presented with a visible to ill-defined mass at sacrococcyx. Our findings were similar to other studies.[6,8,13] In ours study, mass with congenital anomalies seen in 15%. This was 25% in a study by Billman *et al*. Children <2 month age have a better prognosis. Malignant risk rises with age (Herman *et al*., 2002). Rescorla *et al*. and Gabra *et al*. reported that mostly, children >1 year age had malignant tumors.[3] In our study, 16% were malignant and they all presented after 1 month. In our study, the incidence of Altman's types II and III were maximum. Amel Hashish *et al*. studied (Type I >Type II) in their study.[14]

A study by Bilik *et al*. reported that grossly 40% of the sacrococcygeal teratoma were solid, 20% were cystic, and 40% were mixed.[15] Another series by Khanna *et al*., reported that mature teratoma were predominantly cystic and immature teratoma was partly solid and partly cystic.[13] According to a review, majority of benign sacrococcygeal teratoma were cystic in nature.[15] In our study, majority of the mature teratoma were predominantly cystic while immature teratoma was mostly solid.

On histopathology, the majority of sacrococcygeal teratoma are mature followed by immature teratoma. In our study, Grade 0 or mature teratomas are most commonly seen (53%).

Our findings confirm the results by other studies.[6,11,16-20] According to Valdiserri and Yunis, 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] In the 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. Two cases of malignancy have been reported in our study where, yolk sac tumor is the malignant component. Some say size is independent of biological behavior [Table 3].

However, larger tumors are more likely to have immature histology and may have greater intraoperative complications.[16] Mature teratoma should not recur if complete surgical excision and coccygectomy are done. Persistently, elevated alpha fetoprotein levels may indicate a residual or recurrent tumor.

Conclusion

Sacrococcygeal teratoma is the most common congenital neoplasm presenting at birth. Sacrococcygeal teratomas when diagnosed at birth usually reveal fully differentiated tissues and are benign. Meticulous search for the 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 a better outcome. Continued follow-up (with alpha-fetoprotein and radiology) to rule out recurrence.

More effective antenatal screening, for early diagnosis and management, to reduce the risk of malignant transformation, hence better prognosis.

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Conflicts of interest

There are no conflicts of interest.

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