Case Report

Two-stage excision of type III sacrococcygeal teratoma with massive abdominal extension in a 3-month-old girl: A case report with brief literature review

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ABSTRACT

Sacrococcygeal teratomas (SCTs) are the most common germ cell tumor in neonates occurring in approximately 1 in 30,000 to 40,000 births. Here in reporting a 3-month-old girl presented with rapid distension of the abdomen and a tumor-like growth at her right gluteal area, since birth. She also had features suggestive of intestinal obstruction. Clinical examination revealed marked distension of the abdomen, although it was soft and features suggestive of ascites. Visible bowel loops were also detected, suggesting intestinal obstruction. Her perineal/right gluteal examination revealed a soft-to-firm tumor. Her clinical and radiological investigations (ultrasonography and magnetic resonance imaging scan of the abdomen, pelvis, and perineal area) were suggestive of SCT. Abdominal exploration was carried out on an emergency basis for intestinal obstruction, and the cystic tumor was partially excised. After a few days, the remaining pelvic and perineal parts of the tumor were completely excised through posterior, sagittal incision. Her postoperative recovery was uneventful, and she was discharged in good condition. Her resected specimens were subjected to histology and were reported as mature teratoma, without any evidence of malignancy. She was doing well, without a recurrence, in a follow-up done 2 year after the surgical procedures.

Key words: Abdominal lump, Children, Germ cell tumor, Sacrococcygeal teratoma, Surgery

CASE REPORT

History and clinical presentation

A 3-month-old girl presented to the author’s department of pediatric surgery with progressive distension of her abdomen and a tumor-like growth at the right gluteal area since birth. She also had features suggestive of intestinal obstruction. Her perineal and right gluteal examination revealed a soft-to-firm tumor. Her clinical and radiological investigations (ultrasonography and magnetic resonance imaging scan of the abdomen, pelvis, and perineal area) were suggestive of SCT. Abdominal exploration was carried out on an emergency basis for intestinal obstruction, and the cystic tumor was partially excised. After a few days, the remaining pelvic and perineal parts of the tumor were completely excised through posterior, sagittal incision. Her postoperative recovery was uneventful, and she was discharged in good condition. Her resected specimens were subjected to histology and were reported as mature teratoma, without any evidence of malignancy. She was doing well, without a recurrence, in a follow-up done 2 year after the surgical procedures.

Clinical examination

Clinical examination revealed marked distension of her abdomen, and bowel loops were also visible (Fig. 1a), although it was soft and features suggestive of ascites. Her perineal and right gluteal examination revealed a firm, non-tender tumor (Fig. 1b). Perirectal examination allowed only a tip of a little finger, and more details could not be ascertained.

Diagnostic workup

Her biochemical and other hematological investigations were normal. Serum alpha-fetoprotein (AFP) was obtained before the
operative procedures and reported as 160.8 units/mL. USG of the abdomen and pelvis was reported as a large abdominopelvic cystic lesion of 14×10×13 cm, and an approximate volume of 1000 mL. Moderate left-sided hydroureteronephrosis was also reported. She was further investigated with an MRI of the abdomen and pelvis. Her MRI of the abdomen reported a large abdominopelvic cystic lesion that occupied the entire abdominal cavity. The cystic tumor also extended toward the pelvis, anorectal area, perineum, and perineal soft tissues, and into the midline natal cleft (Fig. 2). Multiple septations were also noted within the cystic tumor. MRI of the abdomen further reported that the bowel loops were displaced toward the left side. Bilateral hydroureteronephrosis, more on the left side, was also reported. The conclusion of the MRI findings was suggestive of the abdominal cystic lymphatic malformation. Based on her clinical history and findings, radiological investigations, and raised AFP level, a clinical diagnosis of SCT with intestinal obstruction was made.

Surgical procedures

Abdominal exploration was carried out on an emergency basis due to the findings of intestinal obstruction. Her abdomen was explored through an infra-umbilical midline incision. A large cystic tumor occupying the entire abdominal and pelvic cavity was detected (Fig. 3a). Approximately 1000 mL of clear fluid was aspirated out from the tumor (Fig. 3b). Further pelvic examination revealed that her uterus and fallopian tube were normal. The entire abdominal and most of the pelvic part of the cystic tumor was excised. The distalmost margin of the cystic tumor was closed with a non-absorbable suture. Looking at her poor general condition, a decision was taken to remove the remaining tumor/perineal part of the tumor, in a second operation. After a few days, the remaining pelvic and perineal part of the tumor was completely excised through the posterior sagittal incision (Fig. 4a and b).

Post-operative outcome

She responded well to the surgical procedures, made an uneventful recovery, and was discharged home in good general condition. Her resected specimens were subjected to histology and were reported as mature teratoma without any evidence of malignancy.

Follow-up

She was doing well without a recurrence in a follow-up done 12 months after the surgical procedures. A telephonic conversation done with the father of the girl, 2 years after the surgical procedures, confirmed that she is well, gaining weight, and doing well without any urological or gastrointestinal problems.

DISCUSSION

SCTs are the most common form of extragonadal GCT that occurs during infancy and accounts for up to 70% of all childhood
Ghritlaharey Two-stage excision of Altman’s type III sacrococcygeal teratoma

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<tbody>
<tr>
<td>Sex</td>
<td>Females (%)</td>
<td></td>
<td>74</td>
<td>63</td>
<td>76</td>
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<td>23</td>
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<tr>
<td>Type I</td>
<td>Tumors predominantly external with minimal presacral component (%)</td>
<td>46.7</td>
<td>15.7</td>
<td>39</td>
<td>41</td>
<td>22</td>
<td>22.35</td>
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<td>Type II</td>
<td>Tumors presenting externally but with a significant intrapelvic extension (%)</td>
<td>34.6</td>
<td>29</td>
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<td>48</td>
<td>30</td>
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<td>Type III</td>
<td>Tumors are apparent externally but the predominant mass is in the pelvic with extension to the abdomen (%)</td>
<td>8.7</td>
<td>15.7</td>
<td>17.1</td>
<td>19</td>
<td>16</td>
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<td>Type IV</td>
<td>Presacral tumors with no external presentation (%)</td>
<td>9.7</td>
<td>39.4</td>
<td>19.5</td>
<td>6</td>
<td>12</td>
<td>15</td>
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<td>Histology</td>
<td>Mature (benign) (%)</td>
<td>82</td>
<td>66</td>
<td>68.3</td>
<td>82</td>
<td>66.6</td>
<td>75</td>
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<td>Immature (%)</td>
<td>21</td>
<td>12.2</td>
<td>10.6</td>
<td>10.6</td>
<td>9</td>
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<td></td>
<td>Malignant (%)</td>
<td>17</td>
<td>13</td>
<td>19.5</td>
<td>7.4</td>
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Table 1: Summary of the literature review (number of cases, sex, Altman’s type, etc.)

Teratoma can occur at any part of the body or organ but is most frequent in the sacrococcygeal, gonads (ovary and testes), mediastinal, retroperitoneal, cervicofacial, and intracranial areas [1,2].

A survey conducted by Altman et al. in 1973 included n=405 SCT cases and found that three-fourths (74%) of the cases were females [3]. His results also showed that more than 50% of the cases were detected on the 1st day of life [3]. Predominantly abdominal/pelvic SCTs may present late, and the diagnosis is delayed, in such cases due to the non-specific symptoms [2,3,6]. SCTs not only occur in infants and children but have also been reported in adults, but are rare [8].

Clinically, SCT cases predominantly present with a large tumor located at the sacral area and detected just after birth, or may reported during early infancy or later in childhood [2,3,6,9,10]. SCTs also occur in the pelvis with or without abdominal extension, and, in such cases, the diagnosis is delayed. Such cases may present with abdominal/gastrointestinal symptoms, or urological symptoms due to the compression effect over the urinary bladder or rectum/colon [2,9,10].

SCTs are most frequently detected/discovered before birth by a routine antenatal ultrasound examination [2,4,5,11]. Clinically, most of Altman’s type I and II cases are clinically possible to suspect due to the predominantly sacral mass/tumor present since birth [9,10]. In general, the cases of SCTs are radiologically investigated by doing USG, CT scans, and or MRI scans of the swelling and the abdomen [2,6,9,10,12]. Serum AFP levels are usually obtained before the therapy is initiated/carried out for the cases of SCTs [2,6,9,10,13].

In general, one of the best options for therapy is the complete surgical resection of the SCTs at the earliest. The surgical excision of the SCTs mostly depends on the Altman’s type [2]. Altman’s type I and type II teratomas can easily be excised and approached through the prone position [2,6,9,10]. The removal of the coccyx is the essential step of surgical therapy. For cases with large tumors/abdominal tumors or unresectable SCTs, the best approach is the confirmation of the diagnosis by biopsy, followed by adjuvant chemotherapy and surgical resection of the tumor [2]. SCTs type III and type IV can be approached through a combined abdominoperineal route [9,10].

Depending on the histological/microscopic findings, SCTs are divided/classified as benign (mature), immature, and malignant. Valdiserri and Yunis in a review of n=68 cases of SCTs found that 75% of their cases were benign subtypes [16]. Parvez et al., in a histopathological review of n=66 SCT cases, found that 66.6% of their cases were benign (mature) varieties [14].

SCT cases are also graded on histological findings as follows:

- Grade 0 - Tumor contains only mature tissue (benign tumors)
- Grade 1 – Tumor also contains rare foci of immature tissues
- Grade 2 – Tumor contains moderate quantities of immature tissues, and
- Grade 3 – Tumor contains large quantities of immature tissue with or without malignant yolk sac elements. A brief summary of the literature review is provided in Table 1.

Recurrence after the surgical resection of SCTs has been reported in 10–20% of the cases, and it was more after the surgical excision of initially malignant tumors than the benign SCTs [2,9,10]. Recurrence is mostly due to the incomplete surgical resection of the original malignant tumor, the transformation of the benign tumor into the malignant tumor, or failure of adjuvant therapy [2].

Regular follow-up is a must for the cases treated for the SCTs. During the follow-up visit, a complete clinical examination, including a per-rectal examination, must be done. Ultrasonographic evaluation of the abdomen and pelvis is periodically also required [2]. Serum AFP not only has a prognostic value but is also one of the important tools for detecting the recurrence of SCTs [13,17]. The sensitivity of the AFP levels to detect the recurrence of SCTs is more than 95% [13]. Long-term follow-up of the neonates and older children treated for SCTs is suggested and required. On the long-term follow-up, urinary and or bowel...
problems have been reported in the long-term survivors in as many as one-fourth to 40% of the cases [15].

Overall, the prognosis of SCT cases depends upon the age of patients, Altman’s type, presence of metastasis at the diagnosis, response to the adjuvant therapy, if required, and the potential for recurrence and recurrence [2]. Higher Altman stage, unfavorable tumor biology, and requirement of reoperations are factors associated with poor functional outcomes [15].

CONCLUSION

Although SCT Altman’s type III is common, and is mostly treated by the abdominoperineal approach as a single-stage procedure. Stage-surgical excision of the type III SCT may be a better option for the selected/unusually presented cases, such as the present case report.

REFERENCES


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