Teratoma in Infants and Children

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Abstract

Background: Teratomas arise from three germ cell layers, the ectoderm, endoderm and mesoderm1, 2, and have several degrees of differentiation. We report our experience with teratomas at a tertiary pediatric surgery center.

Patients and Methods: The hospital records of all patients with the pathological diagnosis of teratoma treated during 10 years between 2004 and 2014 were reviewed and the following information was obtained: Sex, site of tumor, treatment and outcome.

Results: Sixty seven patients consisting of 49 girls (73%) and 18 boys (27%) were treated with teratomas at various sites of the body. These included: sacrococcygeal (SC) 32 patients (27 females & 5 males), ovarian 12 cases, cervical 4 patients (1 females & 3 males), retroperitoneal 9 (5 females & 4 males), Nasopharyngeal 2 patients both of which were females, mediastinal 2 cases (1 female & 1 male) and 5 testicular teratoma patients. All patients underwent surgery, and the most common procedure was total resection in 63(94%) of patients. Twenty eight (42%) received chemotherapy. In follow-up 52(77%) patients were in complete remission, 8(12%) had died, and 4 cases did not attend follow-up visits.

Conclusions: Teratomas are a group of tumors with similar histological picture but different behaviors. Sacrococcygeal teratomas are the commonest and the majorities are benign but the risk of malignant transformation increases with age. Management of teratomas is a combination of surgery and chemotherapy which may lead patients to a better prognosis.
Introduction

Teratomas arise from three germ cell layers, the ectoderm, endoderm and mesoderm, and have several degrees of differentiation and may be malignant, benign or something in between. One of the most common anatomic regions for teratoma in the pediatric population is the sacrococcyx (57%), along with other anatomic regions such as retroperitoneal, mediastinum, intracranial, cervical and gonads. Teratomas also can be seen at other parts of body including the GI tract, head & neck and spinal cord. The prognosis of this disease is very variable. Early diagnosis and treatment of this kind of tumor is of great importance since it could experience malignant transformation and invasion. We report our experience with teratomas at a tertiary pediatric surgery center.

Patients and Methods

The hospital records of all patients with the pathological diagnosis of teratoma treated during 10 years between 2004 and 2014 were reviewed and the following information was obtained: Sex, site of tumor, treatment and outcome.

Results

Sixty seven patients consisting of 49 girls (73%) and 18 boys (27%) were treated with teratomas at various sites of the body. These included: sacrococcygeal (SC) 32 patients (27 females & 5 males), ovarian 12 cases, cervical 4 patients (1 females & 3 males), retroperitoneal 9 (5 females & 4 males), Nasopharyngeal 2 patients that both were females, mediastina 2 cases (1 female & 1 male) and testicular 5 patients. All patients underwent surgery, and the most common procedure was total resection in 63 (94%) patients. Twenty eight (42%) received chemotherapy. In follow-up 52 (77%) patients were in complete remission, 8 (12%) had died, and 4 cases did not appear to follow-up visits.

(Fig 1,2,3,4) shows different types of sacrococcygeal teratomas in our patients. (Fig 5) is one case of ovarian teratomas and (Fig 6) is a patient with testicular teratomas.

Figure 1. Sacrococcygeal teratomas in one of our patients.

Figure 2. Imaging in one of our patients with sacrococcygeal teratomas.

Figure 3. Sacrococcygeal teratomas in one of our patients.

Figure 4. Sacrococcygeal teratomas in one of our patients.
Discussion

Teratomas are an interesting group of tumors with different behavior. They are rare tumors and commonly seen in the sacrococcygeal area, especially in newborns. The use of prenatal ultrasound has increased the number of SCT diagnosed in utero. Altman’s classification clearly states that, type IV SCT is completely intrapelvic and difficult to diagnose.

Gabra et al. presented 33 patients with SCT, in seven of which the tumor became symptomatic and was diagnosed after the newborn period and was associated with malignant histology and poorer outcome. The importance of this data needs to be emphasized. In our settings we had similar cases. Three of our patients who presented late and two of them were proven to be malignant. This can occur more commonly in Type III and IV of Altman’s classification. Total excision of SCT together with the coccyx is also important. Derikx et al. had 173 children with SCT and found that incomplete resection and immature /malignant histology are risk factors for recurrence and metastasis. Close follow-up of patients with SCT postoperatively is also very important. These patients have a significant risk of recurrence of about 11% within 3-years.

Cervical teratomas are rare tumors. Tapper and Lack reported only six (2.4%) cervical teratomas among their 354 cases. Four of our patients had cervical teratoma. Cervical teratomas are mainly benign, yet they can cause significant morbidity and mortality. Cervical teratomas tend to grow to a large size causing alteration of the normal anatomy with upper airway obstruction that is life-threatening. In one of our patients with cervical teratoma the anesthesiologist experienced difficulty in intubating and her oxygenation was delayed, leading to severe brain hypoxia. One way to overcome this problem is the EXIT procedure. Two of our patients with large cervical teratomas underwent staged resection and had good outcomes.

Retroperitoneal teratomas are rare (3.5%- 4% of all germ cell tumors in children). Mostly present with abdominal mass. Calcification on plain x-ray should raise the possibility of a teratoma. CT-scan is a valuable modality for planning and surgical excision, and complete surgical resection with careful attention to adjacent major blood vessels is mandatory. The majority of these tumors are benign. Only one of our patients was found to have malignant teratoma. He had complete excision followed by chemotherapy and was well 5 years postoperatively. The second common teratoma in the pediatric population is ovarian teratoma. We had 12 cases in our study. Eight of our patients had malignant teratomas, and others were benign. Five of our patients had testicular teratomas, and two had nasopharyngeal teratomas. One of our nasopharyngeal teratomas was 20-days old with alpha-fetoprotein level of 1403 mg/ml. The mass was excised surgically two days after admission, and he was left with a wide cleft palate after mass resection, which was repaired when he was one years old.

Conclusions

Teratomas are a group of tumors with similar histological picture but different behaviors. Sacrococcygeal teratomas are the commonest and the majorities are benign but the risk of malignant transformation increases with age. Management of teratomas is a combination of surgery and chemotherapy which may lead patients to a better prognosis.

References