Presacral tumors are rare and heterogeneous group of tumors with the majority of published literature addressing adults. In 1985, there were approximately 1:40 000 to 1:63 000 admissions with the diagnosis of a presacral tumor yielding an incidence between 1.4-6.3 patients/year (1). The majority of reports are retrospective small case series and those with more than 10 years of follow-up have fewer than 30 patients (2-5). Reports of presacral tumors in the pediatric literature are even less common and have focused mainly on congenital tumors (6, 7), or have grouped presacral tumors together with general comprehensive cancer reports in children (2, 6-9). To date, there are no reports comparing the pediatric and adult population with presacral tumors. Differences in presacral tumor incidence, presentation, diagnosis, radiographic evaluation, and surgical treatment are not well-defined and may exist between these two groups.

Therefore, the aim of this study was to compare the differences in presacral tumors symptoms, surgical treatment, pathological diagnosis, complications, and outcomes between pediatric and adult patients who have been managed at our tertiary institution.

MATERIAL AND METHODS

An IRB-approved chart review was conducted to identify patients who were treated for presacral tumors at our institution between 1981 and 2011. All patients were identified from a prospectively maintained pathology database using a combination of a natural language search and SNOWMED codes. Search terms for common tumor names included: congenital cysts (developmental cysts, epidermoid cysts, dermoid cysts), tailgut cysts (teratomas, chondomas, anterior meningocele, rectal duplica-
tion, adrenal rest tumors), inflammatory cysts (granulomas, perineal abscess, pelviectical abscess, fistula), neurogenic cysts, (neurofibroma, neurolemmoma, ependymoma, ganglioneuroma, neurofibrosarcoma), osseous (osteoma, osteogenic sarcoma, sacral bone cyst, Ewing’s tumor, giant cell tumor, chordromyxosarcoma), and miscellaneous (metastatic disease, lymphangioma, desmoid tumor, leiomyoma, fibrosarcoma, endothelioma).

We queried SNOWMED codes 49203 (largest tumor ≤ five cm diameter), 49204 (largest tumor 5.1 to 10 cm diameter) and 49205 (largest tumor > 10 cm diameter) to obtain data related to tumor size. Within each size range, we further queried tumor location (peritoneal, mesenteric, or retroperitoneal primary or secondary tumor) and excision method (excised mass or destruction of intra-abdominal tumors, cysts or endometriomas). Patients associated with code 49215 (excision of presacral or sacrococcygeal tumor) were identified within each data set obtained by these queries.

Following chart review and diagnosis verification, patients were divided into a children/pediatric group (< 18) and an adult group (≥ 18 years old). We defined the pediatric group based on The United Nations Convention on the Rights of the Child (10). During the chart review, other collected data included symptoms, physical exam, surgical details, and follow-up. Presacral tumors were then grouped according to the classification published by Uhlig and Johnson (11).

Local recurrence was defined as the presence of a tumor in the presacral space following complete surgical resection. Patients who underwent a complete resection at an outside institution and were later found to have a mass in the same location were included and deemed a local recurrence.

### Statistical analysis

Non-normally distributed quantitative variables were reported as median and range. Differences in categorical variables between groups were analyzed using Chi-square test and associations with quantitative variables were compared using the Mann-Whitney U test. Statistical significance was defined as p<0.05.

### RESULTS

During the study period, 173 patients were treated for the diagnosis of a presacral tumor. Of these, 87 adults and 14 children had a complete record available for review. We randomly selected 50 adults to compare to our pediatric population, resulting in 64 patients in the study group. The median age of the adult group was 48 years (22-83, range) and 1.2 years (1 day to 16 years, range) in the pediatric group. There were more females in both the pediatric (9/14, 64%) and adult (38/50, 76%) groups. Basic patient demographics and characteristics are listed in tab. 1.

### Presenting symptoms and diagnostic imaging

Table 2 lists the patient reported symptoms and imaging modality used in each group. The majority of patients in both the pediatric and adult groups reported symptoms at diagnosis, 9/14 (64%) and 45/50 (90%) respectively, p=0.1. Pain was the most common symptom in 24/45 (48%) adults, while only 2/9 (22%) children had pain, p=0.09. A sacrococcygeal mass was noted in 3/14 (21%) children, while only 1/50 (2%) adults were noted to have a mass, p=0.03. Urinary symptoms were present in 4/50 (9%) adult women. Twelve of forty-five (27%) of adults reported more than one symptom at diagnosis, while only 2/9 (22%) of children had more than one symptom. Additional symptoms seen in both groups included diarrhea, rectal bleeding/blood in stool, constipation, and one tumor was discovered in an adult female following dystocia during her second childbirth. The diagnosis of a presacral tumor in the pediatric group was often made at the initial exam after birth. Perineal anomalies including a tail were found in 5/14 (36%) children in the
Presacral tumors: how do they compare in pediatric and adult patients?

Table 2

<table>
<thead>
<tr>
<th>Presenting symptoms, exam findings, and diagnostic imaging</th>
<th>Pediatric</th>
<th>Adult</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptoms reported</td>
<td>9/14 (64%)</td>
<td>45/50 (90%)</td>
</tr>
<tr>
<td>pain or pressure</td>
<td>2/9 (22%)</td>
<td>24/50 (48%)</td>
</tr>
<tr>
<td>abscess, fistula, or sinus</td>
<td>1/9 (11%)</td>
<td>4/45 (9%)</td>
</tr>
<tr>
<td>gastrointestinal</td>
<td>3/9 (33%)</td>
<td>7/45 (14%)</td>
</tr>
<tr>
<td>anal stenosis</td>
<td>1/9 (7%)</td>
<td>1/45 (2%)</td>
</tr>
<tr>
<td>urinary complaints</td>
<td>0</td>
<td>4/45 (9%)</td>
</tr>
<tr>
<td>Mass detected on physical exam</td>
<td>3/14 (21%)</td>
<td>1/50 (2%)</td>
</tr>
<tr>
<td>Mass detected on digital rectal exam</td>
<td>6/14 (43%)</td>
<td>32/50 (64%)</td>
</tr>
<tr>
<td>Imaging studies (some had more than one)</td>
<td>11/14 (79%)</td>
<td>48/50 (96%)</td>
</tr>
<tr>
<td>CT scan</td>
<td>7/14 (50%)</td>
<td>42/50 (84%)</td>
</tr>
<tr>
<td>MRI</td>
<td>3/14 (21%)</td>
<td>20/50 (40%)</td>
</tr>
<tr>
<td>TRUS</td>
<td>1/14 (7%)</td>
<td>8/50 (16%)</td>
</tr>
</tbody>
</table>

*p=0.03, **p=0.01

pediatric group; there were none in the adult group. Tumors were palpable upon digital rectal exam (DRE) in 6/14 (43%) children and 32/50 (64%) adults, p=0.1.

Diagnostic imaging was obtained less frequently in 11/14 (79%) pediatric patients compared to 48/50 (96%) adults, p=0.07. A CT scan of the abdomen and pelvis was the most commonly obtained radiological study in both groups, but significantly more in adults, 42/50 (84%) compared to 7/14 (50%) pediatric patients, p=0.01. MRI tended to be used more frequently in adults, 20/50 (40%) compared to pediatric patients, 3/14 (21%), p=0.2. Transrectal ultrasound (TRUS) was the least frequently utilized test: 8/50 (16%) adults versus 1/14 (7%) pediatric patients. Presacral tumors were discovered incidentally in 6/50 (12%) adults: four were discovered during a CT scan performed for unrelated reasons, one was discovered during an exam to evaluate a rectal cancer, and one was noted to have an abnormal indentation of the rectum during a screening colonoscopy. In the pediatric group, 2/14 (14%) presacral tumors were discovered incidentally: one during a prenatal ultrasound and the other following birth at a routine exam.

Pre-operative biopsy, treatment, surgical approach, and complications

For the entire group, a total of 20/64 (34%) patients underwent preoperative biopsies, 16/50 (32%) adults and 4/14 (29%) pediatrics. Of the 20 biopsies obtained, malignant pathology was identified in 11 (55%) patients (nine adults; two pediatric). Benign tumor pathology was seen in the remaining seven adults and two pediatric patients. Following tissue diagnosis, the treatment plan was changed in one pediatric patient from surgery to palliative medical treatment due to a diagnosis of metastatic, extra skeletal Ewing’s tumor. Additionally, 5/50 (10%) adults with four hematologic malignancies and one malignant spindle-cell tumor underwent non-surgical management following their biopsy results.

The remaining 58/64 (91%) (45 adult, 13 pediatric) patients were managed surgically. In adults, 9/45 (20%) patients underwent an anterior-only approach, 10/45 (22%) a combined anteroposterior approach, and 27/45 (60%) a posterior-only approach. The adult presacral tumors were located strictly above S4 in 12/45 (27%), both above and below in 12/45 (27%), and strictly below S4 in the remaining 21/45 (47%). In pediatric patients, 3/13 (23%) underwent resection using an anterior-only approach, no combined anteroposterior approach was used, and 10/13 (77%) were resected using a posterior-only approach. Resected pediatric presacral tumors were located strictly above S4 in 1/13 patients, both above and below in 3/13 patients, and strictly below S4 in the remaining 9/13 surgically managed patients. The surgical approaches, tumor size, and location are listed in tab. 3.

Post-operative complications were documented in 2/13 (15%) children. A newborn female developed hemiplegia, rectal prolapse, and decreased anal sphincter tone following perineal resection of a presacral teratoma. The other pediatric patient was a 16 year-old female who...
underwent a laparoscopic resection of a 9.6 cm presacral hamartoma. Postoperatively she was noted to have a rectovaginal fistula that required a stoma and rectal advancement flap for eventual closure. At last follow-up (46 months), there was no recurrence of tumor or fistula, but she continues to self-catheterize her bladder due to urinary retention.

There were 7/45 (16%) adults who developed post-operative complications. Five patients (three male) had postoperative hemorrhage. One 30 year-old male underwent resection of a schwannoma via an anterior approach and suffered a common iliac vein injury, and his additional postoperative complications included *Clostridium difficile* colitis requiring diverting colostomy (later reversed), a superficial surgical site infection, and a cerebral spinal fluid leak. Two other male patients developed hemorrhage and were controlled with pelvic packing for 48 hours; however both developed pelvic abscesses that were drained, treated with antibiotics, and neither experienced tumor recurrence. The fourth case of hemorrhage occurred in a female patient who underwent a schwannoma resection via a posterior approach that resolved spontaneously with conservative management. The fifth case of hemorrhage occurred in a female patient who underwent a schwannoma resection via a posterior approach that resolved spontaneously with conservative management. The fifth case of post-operative hemorrhage was in a 1 year-old female with a teratocarcinoma resected via a combined anteroposterior approach for a 14.5 cm tumor. The bleeding resolved with packing, and although she required surgical resection for multiple local and distant recurrences, she remains alive at the age of 31.

Presacral tumor histology

The distribution of presacral tumors according to Uhlig and Johnson is listed in tab. 4 (11). Congenital tumors were most common in both groups; however the most common congenital tumor type in pediatric patients was a teratoma 6/14 (43%, all females), while in adults it was a hamartoma 17/50 (34%). There were no presacral tumors of neurogenic origin in the pediatric group, but there were 7/50 (14%) neurogenic presacral tumors in adults; six schwannomas and one ganglieneuroma. There were a variety of malignant tumors in both pediatric and adult groups; however malignant tumors tended to occur more frequently in adults, 15/50 (30%), compared to children, 2/14 (14%), p=0.2. One female pediatric patient had a presacral tumor with two histologic components (teratoma and carcinoid), and there were a total of six different malignant presacral tumor types that developed in adults with hematologic presacral malignancies all B-Cell Lymphomas being most common, followed by three chordomas, and two carcinoids. There were no hematologic presacral malignancies in children.

Tumor recurrence

A total of 3/13 (23%) pediatric presacral tumors recurred. One patient developed a local recurrence of a dermoid cyst (original resection at a previous hospital), which was re-excised (repeat posterior-only approach) and has not subsequently recurred. Two additional pediatric patients developed metastases, one from a malignant teratoma (age 1.4 years) and another from a malignant large-cell neoplasm (age 15 years); both patients subsequently died secondary to complications of metastatic disease. Nine of forty-five (20%) adult presacral tumors recurred. Of those adults, four had benign pathology: two hamartomas (one recurred at 19 months and the other at four months), one teratoma, and one schwannoma. Of the seven malignant tumors, two had congenital origins: a neuroblastoma that recurred at 9 months, and a malignant teratoma that recurred at 11 months; two had neurogenic origins: a schwannoma that recurred at 24 months and a neuroblastoma that recurred at 6 months; and three had hematologic origins: a malignant teratoma that recurred at 3 months, a malignant large-cell neoplasm that recurred at 6 months, and a sarcoma that recurred at 9 months. Of these adults, two died secondary to complications of metastatic disease.
Presacral tumors: how do they compare in pediatric and adult patients?

years) and two teratomas (one recurred at 12 months and the other at 10 years). Five adults with malignant tumors developed recurrence disease: two chordomas, one with femur metastasis at six years and the second with local recurrence at 25 months, a fibrous histiosarcoma with local recurrence at ten years who also developed lung metastases at 14 years, a teratoma with a carcinoid that had multiple local recurrences over a 14-year span before a definitive resection, and schwannoma with pulmonary metastasis at 11 months.

DISCUSSION

While presacral tumors are rare, clinicians are likely to encounter at least one during their career (12). Therefore, the subtle yet important differences between pediatric and adult patients may be important for the treating surgeon to recognize. This study highlights differences in presacral tumors that develop in pediatric and adult populations.

Symptoms at presentation

Although previous reports state that the majority of patients with presacral tumors are asymptomatic (26-57%) (13, 14), many patients present with vague, non-specific complaints that may or may not be secondary to their presacral tumors (4, 15-17). In adults, these symptoms include chronic perineal pain, back pain, lower extremity weakness, dimpling of the skin, constipation, and gynecologic complaints (3, 4). Overall, pain has been the most commonly reported symptom in adults with presacral tumors (2, 4, 11, 18) and we also found 53% of our adult patients reported pain as a presenting symptom. Pediatric patients with presacral tumors appear to have fewer symptoms at presentation, but defining pain in a very young patient may be difficult to assess. Pediatric patients commonly present with a posterior sacral mass, persistent embryonic tail, or dimple noted during the initial exam after birth (3, 8, 9). Additional changes in physical appearance or a question of a behavior that seems unusual may be reported by the family, which in turn could alert the physician to a problem. Although external physical exam findings may be absent in adults, digital rectal exam (DRE) has been reported to be very sensitive and discover up to 96% of presacral tumors (3). The sensitivity of DRE for determining a presacral mass in children has been previously reported at 35% (18), though we report a higher detection rate of presacral tumors on DRE
in 43%. Though the data is very limited, benign presacral tumors in children are largely asymptomatic at diagnosis (1). Our data support that claim as only 33% of pediatric patients with benign presacral tumors had symptoms, mostly non-specific abdominal and back pain, and gastrointestinal symptoms including constipation and bowel obstruction.

Diagnostic imaging

Radiographic evaluation of presacral tumors is often obtained prior to surgical intervention. CT, MRI, and TRUS technology remain extremely important to define the tumor anatomy in anticipation of surgical resection (16, 19, 20). It has been reported that CT and MRI are the best individual imaging modalities for the assessment of retrorectal masses with 100% sensitivity, while TRUS has been reported to display 100% sensitivity, but only when combined with proctoscopy (18). Although these techniques were performed frequently in both groups of our study, diagnostic imaging was more frequently obtained in adult compared to pediatric patients, p=0.07 with CT scans the most frequently obtained radiologic study (adult group at 84% compared to children at 50%, p=0.01). The less frequent utilization of CT imaging in children compared to adults may simply be due to recognized concerns over risks of radiation exposure in children. Also during the years of this study, the advantages of CT and MRI and the ease of obtaining them have improved, thus these issues may have decreased the use of these tests in the earlier years. Despite their frequent use even in the adult population, there has not been a clear, documented advantage of CT over MRI in preoperative management of presacral tumors (3, 16, 20-26).

Surgical approach

Aggressive surgical management with complete resection of the mass is typically the treatment of choice for presacral tumors (1, 3). The surgical approach to these tumors is often determined by the relationship of the tumor within the pelvis to the 4th sacral vertebrae (S4), and potential approaches include an anterior-only (abdominal), posterior-only (trans-perineal) or Kraske’s type approach, or a combination of the two (anteroposterior) (1, 3-5, 27, 28). Laparoscopic approaches have recently also been described (29, 30). We found that in both pediatric and adult patients, the majority of presacral tumors were located below S4 and a posterior approach was used most frequently (69% and 47%, respectively). Buchs et al, demonstrated that the posterior only approach is safe and acceptable for low-lying presacral tumors, reporting 15/16 (94%) presacral tumors resected using a posterior-only approach (27). It is noted that this rate of posterior-only approaches is much higher than our study.

Interestingly, a combined procedure was required for 20% of adults, but the combined approach was never utilized in pediatric patients. Additionally, one pediatric tumor was approached laparoscopically, however, it was converted to open due to tumor size and inability to work around the tumor.

Tumor histology

In review of the literature, it would appear that tumor heterogeneity has increased at least in adult patients with presacral tumors as evident by an increase in the “miscellaneous” group of tumors according to Uhlig and Johnson (11); however the overall incidence of presacral tumors has remained the same. Since the incidence of these tumors remains very low, the differences in reporting likely represents improved histopathologic diagnostic techniques and expanded tumor definitions, less likely the development of new sentinel presacral tumors. Between both groups, there were 17 different presacral tumor types identified in this study with seven types in pediatric and 15 types in adult patients. Our distribution of presacral tumors is consistent with prior reports (3, 6, 8, 9, 11, 12, 18, 22, 31). Of all tumors in both pediatric and adult groups, benign, congenital cysts (as a group) occurred most frequently (n=38/64, 59%). However, of all congenital cysts, children developed benign teratomas (43%) most frequently, whereas in adults, hamartomas (34%) occurred most frequently. Of the children with benign teratomas, 4/5 developed in females, and it remains unclear as to why, but this gender predisposition is consistent with previous reports (18, 32-34). We report a great-
er frequency of malignant tumors in adults (30%) compared to children (14%). This may be explained by a low number of children in this study, but our results are within the range of previous reports (8.7-50%) (22, 35). While congenital cystic lesions are the most frequent benign presacral tumor (1, 2, 13), lymphomas occurred as the most frequent malignant presacral tumor overall (resected and unresected), though chordomas were the most commonly resected malignant tumor.

Pre-operative biopsy, tumor recurrence, and survival data

Preoperative tumor biopsies were obtained in 34% of patients with near equal percentages in both children (29%) and adults (32%). Although concerns for complications and increased local recurrence rates in patients undergoing preoperative biopsies are debatable (2, 16, 22, 36, 37), it has been shown that both are comparable to patients who do not undergo biopsy (2, 3). In our study, we report no complication or local recurrence in both pediatric or adult patients who underwent preoperative biopsies, regardless of tumor pathology. Despite this, we report an overall recurrence rate of 21% (12/57) patients and 41% (7/17) patients with malignant tumors, similar to prior reports (2, 3). Five patients who had their overall management changed as a result of their tissue diagnosis from surgical to medical management. Together, this data suggests that a preoperative biopsy may not increase local recurrence rates, but add potentially vital information that may change overall management strategies. One additional adult patient developed a recurrent hamartoma, and is most likely explained by an incomplete initial tumor resection. This is assumed as our Pathology department does not routinely attest margin status in benign tumors.

Overall survival of patients with presacral tumors is favorable; however, it remains worse in patients with malignant disease (2, 6, 7, 13). We report a 95% overall survival for both pediatric and adult patients at 79% at 48 months (1.8-171, range) and 98% at 34 months (1-309, range), respectively. It is well-recognized that malignant presacral teratomas have had an unfavorable prognosis, yet, advances in non-surgical adjunctive therapy have improved overall patient survival (38, 39). Recently, Bittmann et al, reported an overall survival of 94% after 5-year follow-up for pediatric patients with a presacral teratoma (40). We report only three deaths in our study, two children at 12 and 16 months from metastatic presacral tumors, and one adult at 52 months from aspiration pneumonia.

The retrospective nature of this study introduces an inherent bias. Our institution is a tertiary referral center and is likely to report a higher incidence of presacral tumors than most hospitals; however our reported incidence is similar to that of prior publications (2-4, 41). Additionally, we selected 50 adult patients for comparison. While the selection was indeed random, there could be some element of bias due to not including all adults. We also relied in some instances, on outside records provided by referring doctors, which may not have been as complete as we would normally record. Regardless, important clinical information can be gleaned from this study as this comparison has never been reported before. We recognize that interpretation of the results must be performed cautiously given the low numbers of pediatric and adult patients.

CONCLUSIONS

Although presacral tumors in children and adults are similar in most respects, important tumor and clinical characteristics do exist when further investigated. Pediatric presacral tumors are more likely to present as an asymptomatic mass or embryologic remnant, whereas adult tumors are more likely to present with non-specific symptoms of generalized pain or pressure. Preoperative biopsy of these tumors remains controversial; however, we have shown that this may be performed without increased concern for local recurrence in both children and adults, and that histologic diagnosis from tissue biopsy has potential to augment treatment strategies without adding the morbidities of surgery if unnecessary. Moreover, as hematologic malignancies (lymphomas) can occur within the presacral space (42, 43), tissue diagnosis would be paramount as these tumors are primarily treated non-surgically. Though recurrence rate is higher for malignant disease, which is more commonly detected due to more stringent follow-up, be-
nign disease can also recur locally emphasizing complete resection at the time of initial surgery and this principle is applicable for both children and adults. Although outcomes may be similar in pediatric and adult patients, clinicians evaluating patients with presacral tumors should be alert to important differences as they may influence management.

REFERENCES

Presacral tumors: how do they compare in pediatric and adult patients?


Received: 8.03.2013 r.
Adress correspondence: Cleveland Clinic, 9500 Euclid Ave., A30, Cleveland, Ohio 44195
e-mail: messicc@ccf.org