Clinical Features and Outcome of Solid Pseudopapillary Neoplasm

Differences Between Adults and Children

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Objectives: To delineate the clinical and pathological characteristics of solid pseudopapillary neoplasm (SPN), compare them between adults and children, and determine the predictive features suggesting malignant potential.

Design: Retrospective analysis of patients who underwent surgery for a pathologically confirmed SPN.

Setting: Tertiary care referral center.


Main Outcome Measures: Demographic information and clinical presentation, radiological details, surgical data, pathological characteristics, postoperative course, and long-term survival.

Results: Among 62 patients, 47 patients were adults (mean age, 36 years; range, 18-63 years) and 15 patients were children (mean age, 12 years; range, 8-13 years). A palpable mass was the most common presenting symptom in children (9 of 15; 60%) and an incidentally detected pancreatic mass, in adults (18 of 47; 38.3%) (P= .001). The mean tumor size in children was significantly larger than in adults (8.0 vs 6.0 cm; P < .03). In children, the tumor was located in the head of the pancreas (10 of 15; 66.7%) and in adults, in the body or tail (38 of 47; 80.9%) (P = .001). Nine patients (14.5%) had malignant SPN. There was no significant clinical factor suggesting malignant potential. Two patients had a tumor recurrence. They were still alive after debulking surgery. There were no tumor-related deaths.

Conclusion: Solid pseudopapillary neoplasm had different clinical features in adults and children. Because long-term survival can be achieved, even with the synchronous or metachronous metastatic lesions, SPN should be treated aggressively, with complete resection, even if this requires metastatectomy.

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A SOLID PSEUDOPAPILLARY neoplasm (SPN) is a rare primary neoplasm of the pancreas with low-grade malignant potential. Despite a recent increase in recognition, the pathogenesis and guidelines for SPN treatment remain unclear. Even in the event of metastases, extension into adjacent structures, or nodal involvement, radical surgical resection provides long-term survival.1 Solid pseudopapillary neoplasm in children is rare.4-7 Because of the rarity of this neoplasm, the reported information available is limited. To our knowledge, there is no prior report of a comparison of the clinical and pathological features in adults and children. In this study, we evaluated the clinical and pathological characteristics of SPN, the features suggesting malignant potential, and the long-term outcome, comparing them between adults and children, by examining a single institution's experience.

METHODS

Between January 1985 and December 2006, 62 consecutive patients who underwent surgery for a pathologically confirmed SPN at Seoul National University Hospital and Seoul National University Children's Hospital were retrospectively reviewed. Of the 62 patients, 47 patients were adults (mean age, 36 years; range, 18-63 years) and 15 were children (mean age, 12 years; range, 8-13 years). Demographic information and clinical presentation, radiological details, surgical data, pathological characteristics, postoperative course, and long-term survival were evaluated. We compared the outcomes of the clinical and pathological characteristics between females and males and between benign and malignant SPN in the adults and children. Pathologically, SPN was defined as malignant if it
demonstrated deep pancreatic invasion, perineural invasion, vascular invasion, or lymph node metastasis.

Statistical comparisons between the 2 groups were made using the $t$ test or the Fisher exact test for categorical parameters and the $t$ test for continuous variables. $P$ values $<.05$ were considered statistically significant.

## RESULTS

### COMPARISON OF CLINICAL AND PATHOLOGICAL FEATURES BETWEEN ADULTS AND CHILDREN

The clinical and pathological features of the 62 patients are summarized in Table 1. In the adult group, the diagnosis was usually made incidentally during screening by detection of a mass. By contrast, all of the children were symptomatic. This difference in presentation was significant ($P = .001$). Only 38 patients' (61.3%) SPN was diagnosed before surgery. The mean diameter of the tumors based on pathological examination was 6.0 cm (range, 1.5-14 cm) in adults and 8.0 cm (range, 3.5-14 cm) in children. The tumor size was significantly larger in children than in adults ($P = .03$). The most common location of the tumor in all patients was the pancreatic body or tail (43 of 62; 69.4%). In adults, the pancreatic body or tail was the most common location. However, in children, the pancreatic head was the most common site ($P = .001$).

### SURGICAL OUTCOMES

All patients underwent surgical resection for the tumor. Pancreaticoduodenectomy was performed in 18 pa-
tients; distal pancreatectomy, in 38 patients; central pancreatectomy, in 2 patients; and duodenum-preserving pancreatic head resection, in 1 patient (Table 1). Concurrent resection of other organs was performed in 4 patients (6.5%). Two patients underwent a transverse colon resection and anastomosis, and 1 patient underwent portal vein resection and anastomosis because tumor infiltration was suspected; pathology reports showed no tumor involvement. One child underwent a left lateral sectionectomy of the liver because of a synchronous liver metastasis. There was no surgery-related mortality. Postoperative complications were noted in 19 patients (30.6%), including pancreatic fistula in 6 patients, intra-abdominal fluid collection in 6 patients, and delayed gastric emptying in 3 patients. None of the complications required surgical intervention.

CHARACTERISTICS OF MALIGNANT SPN

The pathological findings suggesting malignancy are listed in Table 1. Nine patients (14.5%) had malignant SPN. Perineural invasion was the most common pathological finding. A 13-year-old girl had a synchronous liver metastasis; she underwent near total pancreatectomy with left lateral sectionectomy and was treated with chemotherapy with the OCCG321P2 (combination of cyclophosphamide, cisplatin, doxorubicin hydrochloride, and etoposide phosphate) regimen for a total of 13 cycles. At 3 years’ follow-up, this patient was living, with no evidence of recurrence. During the follow-up period that ranged from 5 months to 20 years (mean, 47.5 months), 2 patients had tumor recurrence. A 25-year-old patient underwent debulking surgery because of SPN in the tail of the pancreas and peritoneal seeding. This patient rejected adjuvant chemotherapy, and 13 years after the initial operation, the size of seeding nodules increased and debulking surgery was performed again. Sixteen years after the initial operation, liver metastasis was detected and a left lateral sectionectomy was performed. Twenty years after the initial operation, the patient was still living. The other patient was 8 years of age and she underwent spleen-preserving distal pancreatectomy because of a ruptured SPN. There were no pathological features suggesting a malignancy. Seven years after the initial operation, peritoneal seeding was detected. Debukling surgery was performed. There were no tumor-related deaths and all of the patients are still living.

PREDICTIVE FEATURES OF MALIGNANT SPN

We compared the clinical features of malignant and benign SPN. On univariate analyses, none of the preoperative features, including age, sex, tumor size, tumor location, elevated carcinoembryonic antigen levels, and elevated carbohydrate antigen 19-9 levels, were predictive of a malignant SPN. We compared the clinical features of malignant and benign SPN. On univariate analyses, none of the preoperative features, including age, sex, tumor size, tumor location, elevated carcinoembryonic antigen levels, and elevated carbohydrate antigen 19-9 levels, were predictive of a malignant SPN (Table 2).

Table 2. Predictive Factors of Malignant Potential

<table>
<thead>
<tr>
<th>Total (N=62)</th>
<th>Adults (n=47)</th>
<th>Children (n=15)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign (n=52)</td>
<td>Malignant (n=10)</td>
<td>P Value</td>
</tr>
<tr>
<td>Age, y, mean (SD)</td>
<td>31 (15)</td>
<td>25 (15)</td>
</tr>
<tr>
<td>Sex</td>
<td>F</td>
<td>47 10</td>
</tr>
<tr>
<td>Symptoms</td>
<td>Yes</td>
<td>33 9</td>
</tr>
<tr>
<td>Preoperative diagnosis</td>
<td>SPN</td>
<td>30 8</td>
</tr>
<tr>
<td>Size, cm, mean (SD)</td>
<td>6.4 (3.0)</td>
<td>6.0 (3.5)</td>
</tr>
<tr>
<td>Head of pancreas</td>
<td>16 3</td>
<td>.51</td>
</tr>
<tr>
<td>Calcification</td>
<td>15 3</td>
<td>.18</td>
</tr>
<tr>
<td>Elevated CEA level</td>
<td>3/34 1/7</td>
<td>.94</td>
</tr>
</tbody>
</table>

Abbreviations: CEA, carcinoembryonic antigen; CA, carbohydrate antigen; SPN, solid pseudopapillary neoplasm.

COMMENT

Recently, the number of cases of SPNs reported in the literature has been steadily increasing. However, there have been only a few large series reporting a single institution’s experience. Furthermore, to our knowledge, there is no prior report comparing the clinical and pathological features in adults and children. This study evaluated 47 adults and 15 children with pathologically proven SPN. The adults and children had different clinical features. A palpable mass was the most common symptom in children with SPN (9 of 15; 60%)...
and no child’s diagnosis was made incidentally. By contrast, an incidentally detected pancreatic mass was the most common presentation in adults (18 of 47; 38.3%) (P = .001). These results are different from the results of other reported studies. Abdominal pain was reported as the most common symptom. The incidental diagnosis in adults may be because of the improved generalized screening programs in Korea. The mean tumor size in children was significantly larger than in adults (6.0 vs 8.0 cm; P < .03). Because only a few cases of children have been reported and prior reports did not divide adult and childhood cases for comparison, we cannot confirm our findings by comparing the results with previous studies. In children, the tumor was more commonly located in the head of the pancreas (10 of 15; 66.7%), and in adults, location in the body or tail of the pancreas was more common (38 of 47; 80.9%) (P = .001). The tail of the pancreas has been the most commonly reported location of the tumor, except for 1 report from our institution. However, all prior reports analyzed the adults and children together, and therefore, potential differences could not be identified.

Although our study is one of the largest clinical and pathological studies to date, consistent with prior reports, there was no factor identified that predicted the malignant potential of an SPN. Malignant behavior (recurrence or metastasis) could not be completely excluded even in the absence of pathological features suggesting a malignant potential; therefore, aggressive tumor behavior was not predictable. In fact, 2 patients with recurrence in our study had no pathological features suggesting malignant potential. Therefore, regardless of the malignant potential, all patients with SPN must be observed closely. A few cases of SPN with peritoneal seeding have been reported who lived for several years after surgery. Solid pseudo-papillary neoplasm is considered to be a tumor that grows slowly and can have stable metastatic disease. For these reasons, despite the large size of these tumors and their ability to extend locally, a complete excision can provide benefits for most patients.

The role of adjuvant or neoadjuvant chemotherapy and radiotherapy for SPN is currently poorly defined because of the difficulty in obtaining data on adjuvant or neoadjuvant therapy. In our study, 2 patients were given adjuvant chemotherapy. To our knowledge, there are only 2 published case reports showing successful use of radiotherapy and neoadjuvant chemotherap yaw for locally advanced unresectable SPN. Further studies are needed to determine the radiosensitivity and chemosensitivity of SPN.

In summary, SPN demonstrated different clinical features in adults and children. Further study is required to elucidate the pathophysiology of these differences. Because long-term survival can be achieved even with the synchronous or metachronous metastatic lesions, SPN of the pancreas should be treated aggressively, with surgical resection, even if this requires metastasectomy.

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REFERENCES