Solid Pseudopapillary Neoplasm of Pancreas: A Case Report

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Abstract: Solid pseudopapillary neoplasm of pancreas is a rare tumor of the pancreas often detected initially on imaging. Of uncertain histogenesis, it has a low grade malignant potential with excellent post surgical curative rates and rare metastasis. On morphology alone other primary pancreatic tumors and metastatic tumors pose a diagnostic challenge. Recent advances in immunohistochemical characterization have made the histopathologic diagnosis more specific and in turn, shed light on the likely histogenesis of this rare tumor. We report a case of SPN of the pancreas that was suspected on radiology.

Keywords: Solid pseudopapillary neoplasm (SPN), Beta-catenin, E-cadherin, pancytokeratin, synaptophysin, vimentin

1. Introduction

Solid pseudopapillary neoplasm are being recognized with increasing frequency owing to increasingly widespread use of imaging techniques, along with a better knowledge and understanding of the disease (1,2). The first report of an SPN of the pancreas was described in 19 years old woman by Dr. Frantz in 1959. At that time he called this neoplasm a papillary tumor of pancreas, benign or malignant (3). In 2010 the World Health Organization (WHO) defined the tumor that Frantz described as a solid pseudopapillary neoplasm of the pancreas. These neoplasm account for 5% of cystic pancreatic tumor and 0.9% to 2.7% of exocrine pancreatic neoplasm (3). Most tumor are located in pancreatic body and tail and are often quite large (>10cm) before clinical detection (4,5,6).

2. Case Report

We report a case SPN arising in an otherwise healthy 19 years old cachectic female. The patient did not have any significant past medical history or family history. She originally presented with vomiting & pain in abdomen, which initially begin in epigastrium and subsequently localized to the patient left upper quadrant. At the time of presentation the patient had begun to have more posterior abdominal pain which increased in intensity with inspiration. The patient stated that she had been eating well but continued to lose weight. The patient reported no diarrhea, constipation. But felt that she has decreased energy. Further workup revealed a hypochromic microcytic anemia and a large abdominal mass by ultrasonography.

Before definitive resection of the tumor, numerous biopsy and FNAC were obtained and submitted for evaluation. Most biopsy sent for histopathological examination which diagnosed solid pseudopapillary neoplasm of pancreas.

Ultrasonography Report: There was round to oval shaped heterogenous hyperechoic with vascularity mass lesion in periampullary region, however, fat planes between mass and head of pancreas & pyloric part appear effaced which were suggestive of periampullary nodal mass.

Computer Tomography report: Well defined heterogenous enhancing solid mass lesion of size 7.5 x 6.3 x 6.0 cm with lobulated margins of head of pancreas, lesion causing widening of C loop of duodenum & compression on portal vein and superior mesenteric vein along its course, however no infiltration in adjacent organs which were suggestive of carcinoma of Head of pancreas.

3. Gross Examination

Specimen received as a whipples specimen of pancreatic mass, common bile duct, gall bladder and duodenum preserved in 10% formalin solution with clinically diagnosed as a pancreatic lymphoma. On gross examination tumor was 10 x 8 x 4 cm with solid cystic area found in head of pancreas. Multiple section studied of pancreatic mass, duodenum, & gall bladder evaluation for histopathological examination.

Histopathology Report: On microscopic examination of pancreatic mass revealed the tumor was composed of sheets of discohesive, large polygonal cells in a pseudopapillary pattern with clear to granular cytoplasm and hyalinized stroma with abundant small vessels and extensive necrosis reported as SPN but section from duodenum and gall bladder showed no specific pathology and chronic cholecystitis respectively and free from tumors. (Photomicrograph no. 1)

Immunohistochemistry Report: Immunohistochemistry stain performed and we found, PANCYTOKERATIN – weak positive , SYNAPTOPHYSIN – focal positive , PROGESTERON RECEPTOR – positive , CD-10 – positive , CD-56 – positive , VIMENTIN – positive.
The histopathological findings and immunohistochemistry support the final diagnosis of solid pseudopapillary neoplasm.

4. Discussion

Solid pseudopapillary neoplasm are a rare, typically benign neoplasm of unknown etiology that predominantly occurs in young females with a mean age of 28 years and display a female to male ratio of approximately 10:1. The tumor accounts for 0.95% to 2.7% of all exocrine pancreatic tumors. Large series & literature reviews have documented survival rates greater than 90%. However, SPN is still considered a tumor of low malignant potential, as local invasion & distant metastases are reported in 5-15% of cases. In our study age of patients was 19 years female, whereas various study showed variable age groups ranging from 17 – 44 years with female preponderance. In our study size of tumor was 10 x 8 x 4 cm. at head of pancreas (exocrine part). whereas various study showed metastasis in the liver & pancreas (exocrine part). whereas other author quoted in our study size of tumor was 10 x 8 x 4 cm. at head of pancreas. In study no metastasis observed but in study of few authors showed metastasis in the liver.

In our study Immunohistochemistry stain performed and we found, PAN CYTOKERATIN – weak positive, SYNAPTOPHYSIN – focal positive, PROGESTERON RECEPTOR – positive, CD-10 – positive, CD-56 – positive, vimentin – positive. Where as other authors showed positivity of IHC which compared with our study. Alpha – 1 antitrypsin, and Alpha – 1 antichymotrypsin are more sensitive markers but none of them performed to confirm the diagnosis of SPN. (Table No. 1)

5. Conclusion

Accurate diagnosis aids surgical management of SPN in which diagnosis was suspected on radiology and confirmed intra-operatively by cytology. Radiofrequency ablation offers a viable option for safe and effective treatment for unresectable liver metastasis.

Table 1: Comparison with other authors IHC Markers

<table>
<thead>
<tr>
<th>Present study in 2017</th>
<th>Progesterone receptor (+)</th>
<th>CD10 (+)</th>
<th>CD56 (+)</th>
<th>Vimentin (+)</th>
<th>Synaptophysin focal positive</th>
<th>Pancytokeratin weak positive</th>
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<tr>
<td>Jain et al in 2016¹¹</td>
<td>Progesterone receptor (-)</td>
<td>CD10 positive</td>
<td>CD56 positive</td>
<td>Vimentin Negative</td>
<td>Synaptophysin focal positive</td>
<td>Pancytokeratin weak positive</td>
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<tr>
<td></td>
<td>Cyclin D1 (+) Beta-Catenin (+)</td>
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<td></td>
<td>CD 99 (+) Non specific enolase (+)</td>
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<tr>
<td>Bailey et al in 2014⁴⁰</td>
<td>CD 9, Desmin,S100, K167, Beta-Catenin (+)</td>
<td>(+)</td>
<td>Strongly positive</td>
<td>Strongly positive</td>
<td>(+)</td>
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<tr>
<td>Lakhtakia study in 2013³</td>
<td>Beta – Catenin &amp; E – Cadherin (+)</td>
<td>(+)</td>
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References

Photo micrograph 1. Showing solid pseudopapillary neoplasm. The cells are adjacent to the tiny blood vessels are attached to the stroma and to one another (H & E, 100X).