UNRAVELING THE ENIGMA OF PANCREATIC SOLID PSEUDOPAPILLARY NEOPLASMS: UPDATED INSIGHTS OF 416 PATIENTS FROM THE SURVEILLANCE, EPIDEMIOLOGY, AND END RESULTS (SEER) DATABASE

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Background:

- Pancreatic solid pseudopapillary neoplasia (SPNs) or Frantz tumor, first discovered in 1950, makes up 1-2% of pancreatic tumors. Usually, SPNs are slow growing, but metastatic cases have been reported. SPNs mainly affect young women in their 20s or 30s, with the reason for this trend still under investigation.
- Symptoms are often vague, and the tumor is discovered unexpectedly on imaging for other diagnoses. While its origins are unclear, some genetic mutations do exist. Surgery remains the treatment of choice, offering the highest survival rates.
- The current study describes the largest cohort of patients with SPNs from the Surveillance, Epidemiology, and End Results (SEER) database to better identify the demographic, clinical, and treatment modalities that impact clinical outcomes..

Methods:

- **Data Source:** Demographic and clinical data were abstracted on 416 patients with SPNs from the SEER research plus database (2000 – 2018). SEER*stat software (version 8.4.0) was used to extract data using the International Classification of Diseases version 3 (ICD-O-3). Patients diagnosed by autopsy, death certificate only, and/or non-microscopically confirmed methods were excluded from the study.
- **Outcomes and Variables:** Patient demographics, including age, sex, and race, were collected at the time of SPNs diagnosis. Tumor characteristics, such as tumor size, grade, stage and metastasis to four organs, liver, lungs, bone and brain were also extracted.
- **Statistical Analysis:** Data were exported to the Statistical Analysis System (SAS) v9.4 for analysis. Descriptive statistics were used to summarize the data. Categorical variables were reported as frequencies and percentages and compared using chi-square tests. Survival analysis was performed using log-rank analysis and Kaplan-Meier survival plots to compare survival among patients with different characteristics. Univariate and multivariate regression analysis was performed to identify variables significantly associated with mortality. A p-value of ≤ 0.05 was considered statistically significant. Hazard ratios (HR) and 95% confidence intervals (CIs) were calculated.

Results:

- A total of 416 cases of SPNs were identified, comprising 353 (84.9%) female and 63 (15.1%) male patients. Most of the patients were of Caucasian decent 283 (68.0%), and the mean age of diagnosis for the female was 35.2 years and 45.1 years for male.
- When information regarding tumor staging 338 (81.3%), grading 89 (21.4%), metastasis 318 (76.4%), and tumor size 204 (49.0%) was available, most of the tumors were localized 218 (64.5%), well differentiated 62 (69.7%), showed no metastasis 307 (96.5%), and were more >4cm in size 131 (64.2%).
- Also, where the treatment information was available, most of the patient received surgery alone 347 (83.4%). The overall 5-year survival was 93.3% (89.2-95.9) and highest in those who received surgery 98.0% (94.9-99.2), while addition of chemotherapy decreased the overall survival rate 72.7% (46.3-87.6).
- Multivariable analysis through cox survival regression analysis identified age >40 hazard ratio (H.R) 9.279 (0.001) and distant stage H.R 4.267 (0.015) as factors that were associated with decreased mortality, p-value <0.05.

Conclusion:

- SPNs preferably affects Caucasian female in the 3rd decade of life and show low metastatic potential. Surgical resection remains a treatment choice as it significantly increases 5-year survival.
- Although chemotherapy alone decreases survival, more extensive studies are needed to investigate the proper role of chemotherapy for SPNs, given the limited number of cases who received chemotherapy in the current study.