Solid Pseudopapillary Neoplasm of the Diaphragm: A Case Report

Eden Gerszman MD[1,3], Vadim Sonkin MD PHD[2,3], Edmond Sabo MD[2,3], Natalia Radzishevsky MD[2,3], Riad Haddad MD[1,3], and Ahmad Mahamid MD[1,3]

[1]Department of General Surgery, Carmel Medical Center, Haifa, Israel
[2]Department of Pathology, Carmel Medical Center, Haifa, Israel
[3]Rappaport Faculty of Medicine, Technion–Israel Institute of Technology, Haifa, Israel

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Solid pseudopapillary neoplasms (SPNs) of the pancreas are rare tumors arising from pancreatic tissue, predominantly affecting young women and possessing low malignant potential. Extrapancreatic SPNs are exceedingly uncommon. According to data from the English literature, only 30 cases of extrapancreatic SPNs had been reported by 1990, accounting for less than 1% of all reported SPNs from 2004 to 2018. The testis, paratesticular region, and ovary are the more frequently documented sites of these tumors [1,2]. Notably, to the best of our knowledge, no cases of SPN originating in the diaphragm have been reported in the English literature to date. The prevailing theory suggests that SPNs behave similarly regardless of whether they originate in the pancreas or in extrapancreatic locations.

We present the case of a 79-year-old female with a history of lung and endometrial cancer, who was diagnosed with a liver lesion during a routine follow-up ^{18}F-fluorodeoxyglucose positron-emission tomography/computed tomography (^{18}F-FDG PET/CT). During surgery, the lesion was resected from the diaphragm and was confirmed to be consistent with the pathological findings of SPN.

PATIENT DESCRIPTION

A 79-year-old female patient with a history of lung and endometrial cancer underwent a CT scan as part of her routine oncological surveillance. The scan identified a lesion in the 8th segment of the liver [Figure 1A]. Although this lesion had been noted in a previous scan, it had increased in size, as indicated by the radiologist.

Subsequent imaging with magnetic resonance imaging (MRI) and ^{18}F-FDG PET/CT suggested a hemangioma and enhanced uptake in the 8th segment, respectively. The chest ^{18}F-FDG PET/CT was unremarkable, and tumor markers were within normal limits. After reviewing these results in a multidisciplinary tumor board meeting consisting of oncologists, radiologists, and surgeons, it was decided to perform an excisional biopsy, as the lesion was not amenable to percutaneous biopsy.

On 7 July 2019, the patient underwent exploratory hand-assisted laparoscopy. Contrary to initial predictions, the lesion was found on the right diaphragm [Figure 1B] and created a hollow space in the 8th segment. Intraoperative ultrasound (IOUS) confirmed that the liver was free of additional lesions. The diaphragmatic lesion was resected, and a perforation with air leak through the diaphragm was observed. This rupture, which led to pneumothorax, was repaired during the procedure.

Postoperatively, the patient's recovery involved managing the diaphragmatic rupture with a chest tube, which was eventually removed.

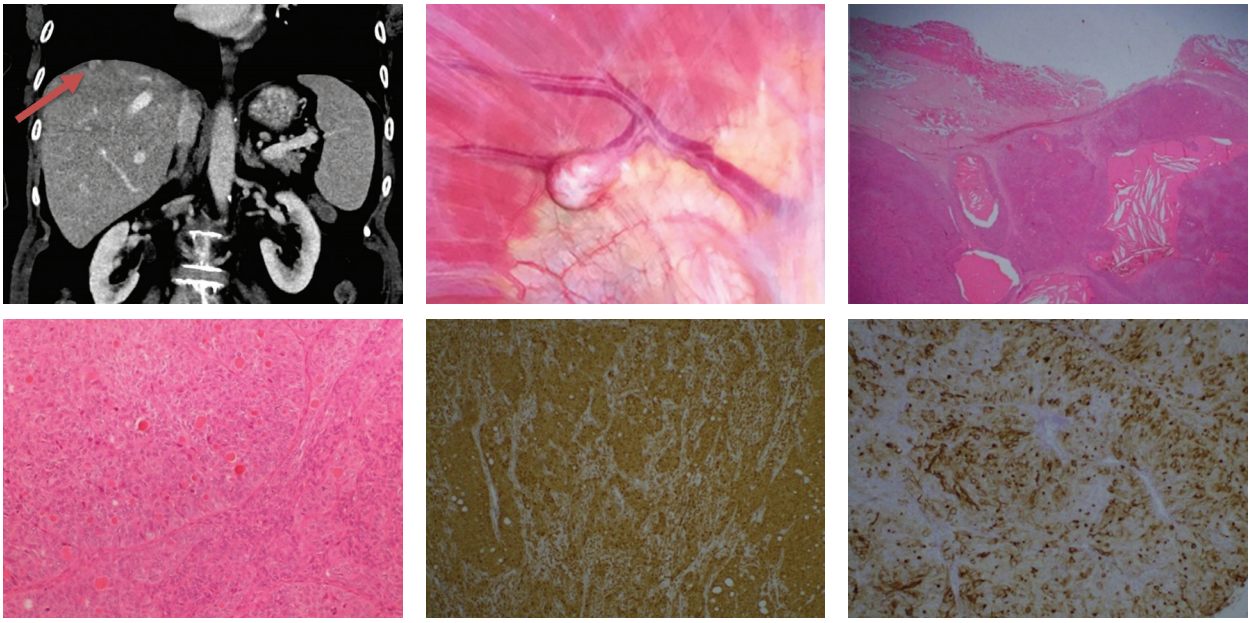
The resected specimen was a nodular tumor, tan to dark brown in color, and measured 2 cm at its largest dimension. Microscopic examination revealed a well-defined but partially encapsulated neoplasm surrounded by diaphragmatic muscle bundles [Figure 1C]. The tumor exhibited a pseudopapillary architectural pattern with fibrovascular septa, hemorrhages, and cholesterol clefts. Neoplas-

Figure 1: Macroscopic and microscopic views of the tumor

[A] Computed tomography scan image, arrow showing a lesion in the 8th segment of the liver

[B] Lesion location on the right diaphragm (during surgery, before resection)

[C] Histological stains: well-defined but partly encapsulated, completely excited neoplasm surrounded by diaphragmatic muscle bundles



[D] Prominent intracytoplasmatic hyaline globules

[E] Tumor cells positive to vimentin, β-catenin

[F] CD10

tic cells displayed mild cytological atypia, with a polygonal and some epithelioid appearance. The nuclei were round to oval, often with nucleoli and occasional grooves, while the cytoplasm varied from eosinophilic to clear and foamy. Prominent intracytoplasmic hyaline globules were noted in some areas [Figure 1D]. The tumor showed 11–12 mitoses per 10 high-power fields (HPF) in the hot spot, and necrosis was absent.

Immunohistochemical analysis revealed that tumor cells were diffusely and strongly positive for vimentin, β-catenin [Figure1E], CD10 [Figure 1F], pan-keratin, and synaptophysin, with focal positivity for chromogranin, P16, and progesterone. The Ki67 proliferative index was positive in 20–25% of the tumor cells, while P53 showed weak positivity in isolated cells. These morphological and immunophenotypic findings were consistent with an extrapancreatic (diaphragmatic) SPN. No pancreatic tissue was identified.

COMMENT

Pancreatic tumors encompass a broad spectrum of both solid and cystic neoplasms. Among the cystic neoplasms, four major categories are recognized: intraductal papillary-mucinous neoplasms, serous microcystic neoplasms, mucinous cystic neoplasms, and SPNs [3]. Among these, SPNs are the rarest.

SPN is an uncommon tumor, predominantly affecting young women (female-to-male ratio approximately 8:1), which lacks pathognomonic symptoms. It is classified as a low-grade malignancy with a favorable 5-year survival rate of approximately 96% [4]. Despite an increase in reported cases in recent years, primary SPNs of non-pancreatic origin remain rare.

Historic case reports have primarily documented SPNs originating from the testicular/paratesticular area, mesocolon, omentum, ovaries, liver, retroperitoneum, and mediastinum [1]. Our case represents the first documented example of an SPN located in the diaphragm, an anatomical site not previously described in the context of extrapancreatic SPN.

The etiology of SPNs remains unclear. One hypothesis suggests that it may arise from ectopic pancreatic tissue or cells related to the genital ridge/ovarian anlage, due to the proximity of these structures during

embryogenesis, which may explain the female predominance [1]. Ectopic pancreatic tissue was not observed in this case.

Radiological features can help distinguish SPNs from other pancreatic cystic lesions. Typically, SPNs present as an inhomogeneous mass with both solid and cystic components, influenced by hemorrhagic degeneration. The mass is usually encapsulated with progressive enhancement following contrast administration and gradual intralesional fill-in enhancement during portal and venous phases. MRIs and magnetic resonance cholangiopancreatography are valuable for demonstrating the tumor's relationship with the main pancreatic duct, its capsule, and intratumoral hemorrhage due to their superior resolution.

Histologically, SPNs exhibit distinct features with varying proportions of solid and pseudopapillary growth patterns, occasionally presenting with cystic elements. The immunohistochemical profile of SPNs is characterized by mandatory reactivity to β-catenin and CD10 markers [3].

Predicting the potential of aggressiveness of SPNs remains challenging. Key clinical and histological predictors include tumor size, patient age and sex, cellular atypia, mitotic rate, proliferative index (Ki67), lymphovascular and perineural invasion, and infiltration into surrounding organs [4]. Consensus among researchers suggests that the presence of lymphovascular invasion and a Ki67 index > 5% are significant indicators of tumor aggressiveness [5]. The overall 5-year survival rate following primary resection of SPN is over 95% in large-scale reviews, with a recurrence rate of up to 6.6%. The impact of extrapancreatic SPN location on survival and recurrence rates remains unclear due to the limited number of such cases. However, case reports suggest similar clinical courses and outcomes for pancreatic and extrapancreatic SPNs [5], although such extrapolation may be statistically unreliable.

The presence of prominent mitotic activity, a high Ki67, and focal strong positivity for P16 in our case may indicate an aggressive course. Long-term follow-up will ultimately provide insight into the tumor's behavior.

The only curative treatment for SPN is surgical resection. If an R0 resection is achieved, regular follow-up is typically sufficient. In rare examples of metastasis, chemotherapy (usually 5-fluorouracil and gemcitabine) may be required.

CONCLUSIONS

SPN of the pancreas is rare, with extrapancreatic occurrences being even less common. To the best of our knowledge, this is the first reported case of diaphragmatic SPN in the English literature. Further research is required to characterize these rare presentations of SPN and to identify patients at risk for such atypical manifestation. We encourage the medical community to report additional uncommon presentations of SPNs and advocate for the establishment of an international registry.

Correspondence

Dr. E. Gerszman
Dept. of General Surgery Carmel Medical Center, Haifa 3436212, Israel
Phone: (972-4) 825-0292
Fax: (972-4) 825-0927
Email: eden.gerszman@gmail.com

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**Life cannot be classified in terms of a simple neurological ladder, with human beings at the top;
it is more accurate to talk of different forms of intelligence, each with its strengths and weaknesses
This point was well demonstrated in the minutes before last December's tsunami [2004],
when tourists grabbed their digital cameras and ran after the ebbing surf, and all the 'dumb' animals made for the hills.**

Brian Reynolds Myers (born 1963), usually cited as B. R. Myers, an American professor of international studies at Dongseo University in Busan, South Korea, best known for his writings on North Korean propaganda.