University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2020, Vol. 6(3)

SOLID AND CYSTIC PSEUDOPAPILLARY TUMOUR OF PANCREAS - A RARE SURGICAL ENIGMA

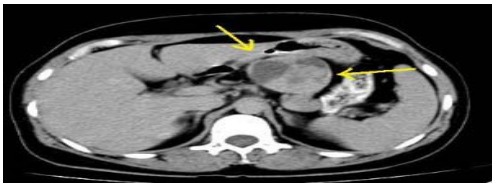
SANDIP T GEORGE
Department of General Surgery, MADURAI MEDICAL COLLEGE AND HOSPITAL

Abstract : Solid-pseudopapillary neoplasms (SPNs) of the pancreas are rare neoplasms, comprising only 1 to 2 of all pancreatic tumors. There is a strong female preponderance, and most SPNs present in the third and fourth decade of life. The first published description of an SPN was by Frantz in 1959. This report consisted of a pathologic description of three patients with SPN. Hamoudi and colleagues added an additional patient to the literature in 1970 and detailed the electron microscopic appearance of the tumor. The first report in the surgical literature of an SPN was by Sanfey and associates in 1983. SPNs are also called solid and papillary tumors, papillary cystic tumors, solid cystic tumors, Frantz tumors, and Hamoudi tumors. SPN is synonymous with the preceding names and is the preferred terminology. SPNs are defined by their gross and histologic appearance. They are composed of discohesive polygonal cells that surround delicate blood vessels and form a solid mass, with frequent cystic degeneration and intracystic hemorrhage. The neoplastic cells have uniform nuclei, finely stippled chromatin, and nuclear grooves. Eosinophilic globules, foam cells, and cholesterol clefts are often present. Symptoms of SPN are often nonspecific and include abdominal pain, dyspepsia, early satiety, and nausea and vomiting (41 to 64).SPNs are usually localized pancreatic neoplasms, although 10 to 15 of patients will develop metastases.These metastases are often amenable to resection, and complete extirpation is associated with long term survival. Reported clinical and histopathologic features predictive of recurrence or metastases include tumor size greater than 5 cm, venous invasion, nuclear grade, and prominent necrobiotic nests, but these features are not consistently reported in all large series.

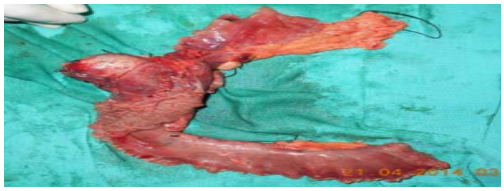
Keyword : pancreas, pseudopapillary, pancreaticoduodenectomy, frantz

CASE REPORT

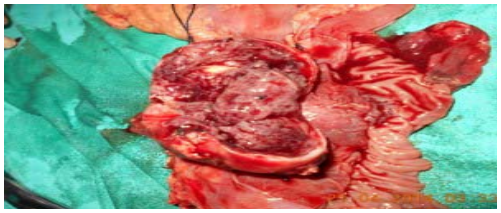
A 47year old woman, in good overall health sought medical attention for recurrent bouts of abdominal pain for 4 days and 4-5 episodes of vomiting and fever. There was no h/o weight loss/jaundice/diarrhea. Abdominal USG, revealed ill defined non homogenous solid lesion behind the head of pancreas of size 9*7.5cm, Common bile duct was dilated (9mm). Gall bladder was grossly distended with minimal fluid collection .CT scan suggested large heterogeneously enhancing soft tissue density at head of pancreas with compression of IVC - ?Ca head of pancreas USG guided needle aspirate from pancreas demonstrated features suggestive of either neuro endocrine/acinar cell carcinoma of pancreatic origin.



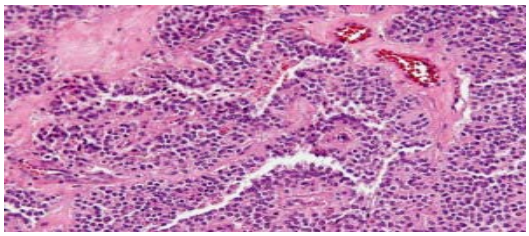
CT SCAN-PSEUDOPAPILLARY TUMOUR WITH CYSTIC DEGENERATION AT HEAD OF PANCREAS



MASS AT THE HEAD OF PANCREAS-POST OPERATIVE SPECIMEN



CUT SECTION



HISTOLOGICAL VIEW-PSEUDOPAPILLARY TUMOUR OF PANCREAS

IHC staining was inconclusive. Given the unusual appearance of the tumor ,Its growth and indeterminate pathology, as well as symptoms, laparotomy was performed for pancreaticoduodenectomy (whipples). The bulky mass (11.5*10*6.5cm)was seen at the head of the pancreas adhered onto the underlying IVC and duodenum .Surgery was performed without any complications. Post operatively, patient had uneventful 9 day period. Histological review of lesion revealed yellow tumour with solid and cystic areas. No associated lymph node metastasis was detected for malignancy. Microscopic analysis revealed, sheets of uniform polygonal cells with pseudopapillary appearance and gave the impression as-solid and cystic pseudo papillary tumour of pancreas. Surgical margins were also negative for malignancy. Patient was started on oral diet on 5th POD and was discharged on 10 th POD. Monthly post operative follow up also turned out to be symptom free with CT abdomen revealing normal results.

DISCUSSION

This case illustrates many of the salient features of this rare tumor, which makes up <1% of all pancreatic neoplasms. Many reports have emerged over the last 25 years describing nearly 300 cases. However, despite generally consistent pathologic and clinical characterizations, some controversies remain. The nomenclature of this pathologic entity is confusing and ranges from Frantz tumor to the recently favored papillary cystic neoplasm. We prefer the term solid pseudopapillary tumor of the pancreas for two reasons. First, cystic changes are not a ubiquitous feature, and instead usually occur in larger lesions secondary to longstanding tumor necrosis. Secondly, the papillary appearance of the tumor is a result of cellular clustering around the microvasculature with more discohesive cells in the periphery, and is not due to the presence of true papillary stalks. The precise cellular derivation of this tumor remains elusive, so routine immunohistochemical staining is not consistent in determining its phenotype. A variety of stain expressions have been described, representing neural, epithelial (ductal), and stromal (acinar) elements, but a general immunophenotype has emerged . These neoplastic cells regularly express vimentin, alpha-1-antitrypsin, and alpha-1-antichymotrypsin. Also, neuron specific enolase is usually faintly positive. The literature regarding the cellular differentiation of this tumor remains inconclusive. Arguments have been made that champion each of the three lineages described above. Given the inconsistent findings, an attractive hypothesis has been developed that these tumors originate from a ‘primordial’ pancreatic cell line. However, there are no conclusive data to support this line of reasoning. The biologic behavior of solid pseudopapillary tumor is less aggressive than that of many other pancreatic tumors, and its prognosis is better. Surgical extirpation of the tumor will result in almost total survival (>95%) for those patients with tumors confined to the pancreas at presentation. Despite its potential for local infiltration, recurrence is rare following complete excision. Likewise, it is rare for metastases to develop metachronously.

CONCLUSION

In conclusion, SPN is a rare neoplasm that is found primarily in young women. These lesions present with abdominal pain but not jaundice. Formal surgical resection may be performed safely and should be favored for patients with SPN. It is clear that although some SPNs behave aggressively, most do not, and we currently cannot predict outcomes for these patients

REFERENCES

1. Del Castillo CF, Reyes E, Muñhoz L, Campuzano M. Papillary cystic neoplasm of the pancreas. Two cases treated by surgery. Rev Invest Clin. 1990;42:217–21.
2. Bapat KC, Pandit AA, Joshi AS, Vora IM. Aspiration cytol ogy of papillary cystic neoplasm of the pancreas. A case report. Ind J Cancer. 1992;29:100–3.
3. Ky A, Shilyansky J, Gerstle J, et al. Experience with papillary and solid epithelial neoplasms of the pancreas in children. J Pediatr Surg. 1998;33:42–4
4. Frantz VK.. Tumors of the pancreas, Atlas of Tumor Pathology VII. In:. Fascicles 27 and 28. Washington: Armed Forces Institute of Pathology, 1959 : 32–3.
5. Kingsnorth AN, Galloway SW, Lewis Jones H, Nash JR, Smith PA. Papillary cystic neoplasm of the pancreas: presentation and natural history in two cases. Gut. 1992;33:421–3.
6. Matsunou H, Konichi F, Yamamichi N, Takayanagi N, Mukai M. Solid, infiltrating variety of papillary cystic neoplasm of the pancreas. Cancer. 1990;65:2747–57.
7. Jayaram G, Chaturvedi KU, Jindal RK, Venugopal S, Kapoor R. Papillary cystic neoplasm of the pancreas. Report of a case diagnosed by fine needle aspiration cytology. Acta Cytol. 1990; 34:429–33.
8. Cubilla AL, , Fitzgerald PJ.. Tumors of the exocrine pancreas, Atlas of Tumor Pathology. In:, 2nd series, Fascicle 19. Washington: Armed Forces Institute of Pathology, 1984 : 201–7.
9. Compagno J, Oertel JE, Kremzar M. Solid and papillary epithelial neoplasm of the pancreas, probably of small duct origin: a clinicopathologic study of 52 cases. Lab Invest. 1979;40 :248–9.
10. Pezzi CM, Schuerch C, Erlandson RA, Dietrick J. Papillary-cystic neoplasm of the pancreas. J Surg Oncol.1988;37:278–85.
11. Crinnion JN, Williamson RCN. Garden OJ. WB Saunders; London: 1997. Pancreatic neoplasia in hepatobiliary and pancreatic surgery, Hepatobiliary and Pancreatic Surgery; pp. 321–51.
12. Bombí JA, Milla A, Badal JM, Piulachs J, Estape J, Cardesa A. Papillary-cystic neoplasm of the pancreas. Report of two cases and review of the literature. Cancer. 1984;54:780–4.
13. Yamaguchi K, Hirakata R, Kitamura K. Papillary cystic neoplasm of the pancreas: radiological and pathological characteristics in 11 cases. Br J Surg. 1990;77:1000–3.
14. Wang KS, Albanese C, Dada F, Skarsgard ED. Papillary cystic neoplasm of the pancreas: a report of three pediatric cases and literature review. J Pediatr Surg. 1998;33:842–5.
15. Ladanyi M, Mulay S, Arseneau J, Bettez P. Estrogen and progesterone receptor determination in the papillary cystic neoplasm of the pancreas with immunohistochemical and ultrastructural observations. Cancer.1987;60:1604–11.
16. Komorn JH, Zirkin RM, Nathan LE., Jr Papillary cystic neoplasm of the pancreas: report of two cases of a surgically curable tumor. Surgery. 1986;99:110–13
17. Horisawa M, Niinomi N, Sato T, et al. Frantz's tumor (solid and cystic tumor of the pancreas) with liver metastasis: successful treatment and long-term follow-up. J Pediatr Surg. 1995;30:724

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities