

A RARE CASE OF MALIGNANT SOLITARY FIBROUS TUMOR OF THE PANCREAS

Deka M K* and Talukdar A

Pathology Department, Cachar Cancer Hospital & Research Centre (CCHRC),
Meherpur, Silchar-788015, Assam

ARTICLE INFO

Article History:

Received 22nd May, 2018

Received in revised form 5th
June, 2018

Accepted 16th July, 2018

Published online 28th August, 2018

Key words:

Solitary Fibrous Tumor, Pancreas,
Cytologically malignant.

ABSTRACT

Solitary Fibrous Tumor (SFT) of the Pancreas is a rare tumor with a recent literature review tallying only 18 cases. Only 2 cases of cytologically malignant SFT of the pancreas have been reported till date. A SFT histology shows a patternless architecture, characteristic hypercellular and hypocellular areas separated by collagen bands and branching thin walled vessels. IHC plays an important role in separating tumors with SFT like features. CD34 is positive in 90-95% of SFT although the most emerging sensitive and specific marker is STAT6. Complete surgical excision is the treatment of choice. We are reporting the third malignant SFT of pancreas and the first instance where a similar tumor recurred.

Copyright © 2018 Deka M K and Talukdar A. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

INTRODUCTION

SFT of the Pancreas is a rare tumor and only 18 case reports have been found. Most have benign histology and only 2 cases have been reported as malignant SFT. A SFT histology shows a patternless architecture, characteristic hypercellular and hypocellular areas separated by collagen bands and branching thin walled vessels. IHC plays an important role in separating tumors with SFT like features. CD34 is positive in 90-95% of SFT although the most emerging sensitive and specific marker is STAT6.

MATERIALS AND METHODS

The present study on a rare case of malignant solitary fibrous tumor of the pancreas was undertaken in Cachar Cancer Hospital & Research Centre (CCHRC), Meherpur, Silchar, India.

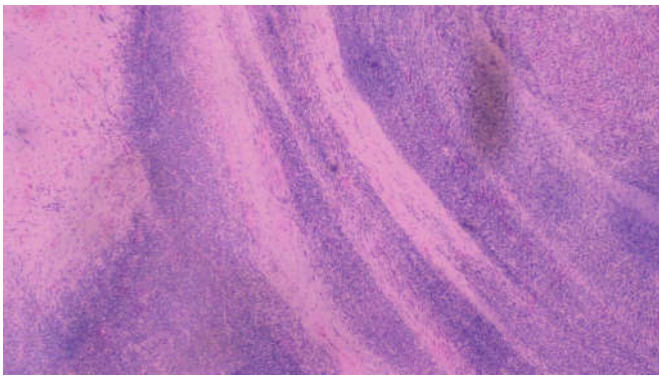
Case Report

A 28year old male from Silchar was admitted in the Surgical Department with abdominal pain on and off for last 3 months. USG abdomen revealed a hypoechoic mass in the pancreatic region. CECT confirmed a large necrotic tumor involving the body and tail of pancreas. No significant abdominal nodes or metastatic disease found. Abnormal laboratory finding include an elevated amylase level. USG guided FNAC was non diagnostic. The Surgical removal of the mass was considered necessary and the patient underwent distal pancreatectomy. Segmental resection of the colon was also done because of presence of adherence. The mass was well circumscribed and

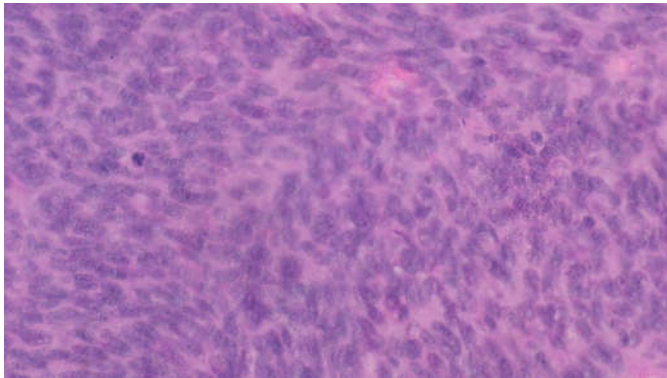
was contained entirely within the pancreas on intraoperative examination. The resected mass measured 6.5cm in greatest diameter. Histologically showed a cellular tumor composed of spindle shaped tumor cells with elongated nuclei in a patternless architecture. There was alternating hypercellular and hypocellular areas, and some areas showed necrosis as well as hyalinization. Nuclear atypia and mitosis (>4/10HPF) including atypical forms were prominent. Stromal blood vessels were thin walled, dilated or staghorn shaped. There was no infiltration into the pancreatic tissue. Colon was free from tumor infiltration. Immunohistochemically, the tumor cells were positive for CD34 and MIC2 was patchy weak positive. KI index was 20%. The tumor cells were negative for smooth muscle actin, Desmin, S100, and BCL2. The final diagnosis was malignant pancreatic solitary fibrous tumor. One year and 8months later he came with another swelling in the anterior abdominal wall. CT Scan revealed a large heterogenous mass with central necrotic areas seen in left rectus abdominus muscle. Resection of the abdominal wall mass was done. The final diagnosis was malignant solitary fibrous tumor. In the 12 month follow up period the patient is alive and well.

*Corresponding author: Deka M K

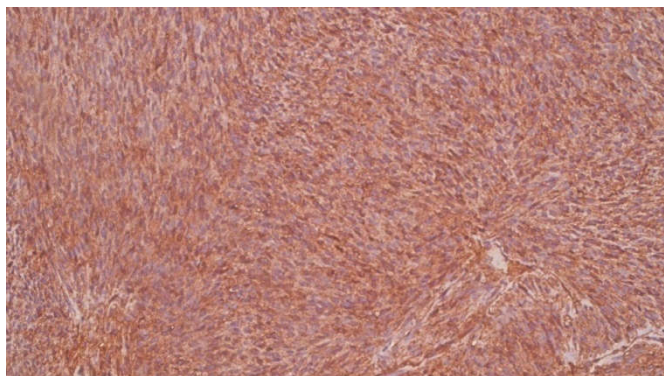
Pathology Department, Cachar Cancer Hospital & Research Centre (CCHRC), Meherpur, Silchar-788015, Assam



Photomicrograph 1 shows a patternless architecture of Solitary Fibrous Tumor.



Photomicrograph 2 shows cytologically malignant SFT



Photomicrograph 3 shows tumor cells positive for CD34.

DISCUSSION

Pancreatic SFT is an exceedingly rare occurrence, with a recent literature review tallying only 18 cases [1,2,3]. Only 2 cases of cytologically malignant SFT of the pancreas have been reported till date [3,4]. 2013 WHO classification defines SFT as an intermediate (locally aggressive) tumor of fibroblastic/myofibroblastic differentiation [5]. A tumour primarily of the pleura it can also occur in extrapleural locations most commonly subcutaneous and deep tissues of the extremities and head and neck area [6].

Paramythiotis *et al* in his review of 16 case reports of Pancreatic SFT found a female predominance, age range of 24 to 78 years and a median tumor size of 5.83 cm [2]. Two more case reports after that showed a similar statistics [1,3]. The 2 cases of malignant SFT occurred in a 52 years old woman in the pancreatic head with a size of 15cm and the other occurred in a 39 years old woman in the pancreatic head with a size of only 2.2cm [1,4]. No single case report has shown any recurrence or metastatic disease.

Our case was the first instance where similar tumor recurred in the anterior abdominal wall. Histologically SFT shows a patternless architecture, well characterized alternating hypocellular and hypercellular areas separated by thick collagen bands and branching thin walled vessels [7,8].

The malignant SFT exhibits hypercellularity, mitosis $>4/10\text{HPF}$, necrosis, prominent cytologic atypia and infiltrative margin [7].

Our case was cytologically malignant SFT except there was no infiltration into the surrounding pancreatic tissue.

The differential diagnosis of cytologically malignant SFT include MPNST, Spindle cell synovial sarcoma both showing SFT like features, GIST, spindle cell rhabdomyosarcoma and leiomyosarcoma [6].

90-95% of SFT are positive for CD34. SFT can also be positive for EMA, SMA, CD99 (MIC) and BCL2. A high KI67 proliferation index is indicative of malignancy. The most sensitive and specific marker of SFT is STAT6. The molecular pathogenesis is NAB2-STAT6 fusion resulting in overexpression of STAT6 [6,9].

In our case MPNST (S100-ve), Spindle cell synovial sarcoma (CD34+ve & BCL2-ve), GIST (CD117-ve), spindle cell rhabdomyosarcoma & leiomyosarcoma (SMA & Desmin -ve) could be conveniently excluded from IHC analysis.

Complete surgical excision is the treatment of choice [10]. Baxter *et al* in analysis of 9 case reports of pancreatic SFT (all benign histology) showed no recurrence and the average length of follow up was 19.3 months, highest being 3 years post-op. In one of the two case reports of malignant pancreatic SFT there was no evidence of recurrence or metastasis at 40 months after resection [10].

CONCLUSION

We are reporting the third malignant SFT of pancreas and the first instance where a similar tumor recurred. Both histology and IHC plays an important role in diagnosis. Long term surveillance should be followed.

References

1. Clare French, Russell Dorer, Flavio G Rocha Malignant Solitary Fibrous Tumor of the Pancreas JOP. J Pancreas (Online) 2017 Mar 30; 18(2):159-162.
2. Daniel Paramythiotis, KonstantinaKofina, Petros Bangeas, FaniTsiompanou, Georgia Karayannopoulou, and George Basdanis Solitary fibrous tumor of the pancreas: Case report and review of the literature World J Gastrointest Surg. 2016 Jun 27; 8(6): 461-466.
3. Liljana Spasevska, Vesna Janevska, Vlado Janevski, Biljana Noveska, JulijaZhivadnovik Solitary Fibrous Tumor of the Pancreas. A case report and review of literature. 10.1515/prilozi-2016-0024. *Contributions Sec. of Med. Sci.*, XXXVII 2-3, 2016 MASA.
4. Estrella JS1, Wang H, Bhosale PR, Evans HL, Abraham SC. Malignant Solitary Fibrous Tumor of the Pancreas. *Pancreas*. 2015 Aug; 44(6):988-94.
5. Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F, eds. WHO classification of Tumors of Soft Tissue and Bone. 4th ed. Lyon, France: IARC Press; 2013. World Health Organization of Classification of Tumors; vol 5.
6. Shi Wei, Evita Henderson-Jackson, XiaohuaQian, Marilyn M Bui Soft Tissue Tumor

- Immunohistochemistry Update. Illustrative examples of Diagnostic Pearls to avoid Pitfalls. *Arch Pathol Lab Med*-Vol 141, August 2017.
7. Peter A. Humphrey, Louis P. Dehner, John D. Pfeifer The Washington Manual of Surgical Pathology 2nd Edition.
 8. Sugawara Y, Sakai S, Aono S, Takahashi T, Inoue T, Ohta K, Tanada M, Teramoto N. Solitary fibrous tumor of the pancreas. *Jpn J Radiol.* 2010;28:479-482.
 9. Fisher C Immunohistochemistry in diagnosis of soft tissue tumors. *Histopathology.* 2011 Jun; 58(7):1001-12.
 10. Andrew R. Baxter, ElliotNewman, and Cristina H. Hajdu Solitary fibrous tumor of the pancreas. *J Surg Case Rep.* 2015 Dec; 2015(12): rjv144.

How to cite this article:

Deka M K and Talukdar A (2018) 'A Rare Case of Malignant Solitary Fibrous Tumor of the Pancreas', *International Journal of Current Medical And Pharmaceutical Research*, 04(8), pp. 3569-3571.
