Case Report

Pancreatic Schwannoma - A Rare Entity

Rashi Garg[1], N Kakkar[2], R.K.Vashisht[2]

[1]Department of Cytology, Postgraduate Institute of Medical Education and Research, Chandigarh, India.
[2]Department of Histopathology, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Keywords: *Pancreas, Schwannoma*

ABSTRACT

Pancreatic schwannomas are rare neoplasms. These tumors vary in size and two thirds are partially cystic which grossly mimic pancreatic cystic lesions. Computed tomography and magnetic resonance imaging are the primary imaging modalities. Definitive diagnosis is made on histopathologic examination. Surgical resection is the mainstay of treatment.

***Corresponding author:**
Dr. N .Kakkar, Professor, Department of Histopathology, Postgraduate Institute of Medical Education and Research, Chandigarh, India.
E-mail: nandita_kakkar@yahoo.com

Introduction

Pancreatic Schwannomas are exceedingly uncommon neoplasms. According to recent studies less than 50 cases of pancreatic schwannomas have been described in English literature over past 30 years.[1,2] Here we report a rare case of schwannoma in the uncinate process of pancreas with preoperative diagnosis of cystic tumor of pancreas.

Case Report

A 62-year-old male presented with pain abdomen in the left upper quadrant. Physical examination and laboratory data including the tumor markers were within normal limits. The contrast enhanced computed tomography (CECT) scan of abdomen demonstrated a large heterogeneous mass lesion measuring 3.5x5x7cm in precaval and peripancreatic region which was suggestive of a lymphnode mass or an exophytic mass in pancreas. Endoscopic ultrasonography showed 5.5x3.5cm lesion with well defined region, central hypoechoiec and anechoiec (0.2x0.2cm) areas. Patient was taken up for Whipple procedure. The laparotomy disclosed a mass in the uncinate process of the pancreas, measuring 6.5x6.5cm with central necrosis (Fig 1). In addition to the mass lesion in the pancreas a duodenal polyp measuring 1x1cm in D2(away from papilla) was also identified. On microscopic examination, the tumor was well encapsulated. It was composed of spindle cells, arranged in fascicles showing focal nuclear palisading. Hypo/and hypercellular areas were seen and many foamy macrophages were also noted indicating degenerative changes (Fig2,3). Tumor cells were positive for S-100 (nuclear positivity)(Fig 3inset) and negative for smooth muscle actin. The tumor was therefore histologically diagnosed as a benign schwannoma. The polyp in the duodenum was a submucosal lipoma.

Discussion

In 1910, Verocay reported a schwannoma as a true neoplasm which originated from Schwann cells, and which did not contain neuroganglion cells.[3] Since then, schwannomas have become well known as benign spindle cell tumors derived from Schwann cells. Schwannomas usually occur in the extremities, but can also be found in the trunk, head and neck, retroperitoneum, mediastinum, pelvis and rectum.[4-6] Pancreatic schwannomas are rare neoplasms that arise from either autonomic sympathetic or parasympathetic fibers, both of which course through the pancreas as branches of the vagus nerve.[4-6] Moriya et al. reviewed 47 cases from literature and found that vast majority of these tumors are located in the pancreatic head (40%), followed by body (21%),body and tail (15%),uncinate process (13%),head and body (6%) and tail (4%). Patients are in age group of 18-87 years and 45% are males. Size of the tumors ranged from 1-20cm. Majority of the tumors were cystic (60%) and benign (87%). [1]Table 1 shows 6 case reports of different years including the present one. All the 6 cases are benign with size ranging from 1-16cm.

Cystic pancreatic schwannomas can mimic the whole spectrum of cystic pancreatic lesions including: intraductal mucinous-papillary neoplasms, mucinous cystic neoplasms, serous cystic neoplasms, solid and pseudo-papillary neoplasms, lymphangiomas, and pancreatic pseudocysts. Microscopically, a typical schwannoma is composed of two areas, namely Antoni A and Antoni B areas. The Antoni A area is hypercellular and characterized by closely packed spindle cells with occasional nuclear palisading and Verocay bodies, whereas the Antoni B area is hypocellular and is occupied by loosely arranged tumor cells.[9] Degenerative or cystic changes such as calcification or hemorrhage are often recognized in the Antoni B area. These changes result from vascular thrombosis and subsequent necrosis.[9]

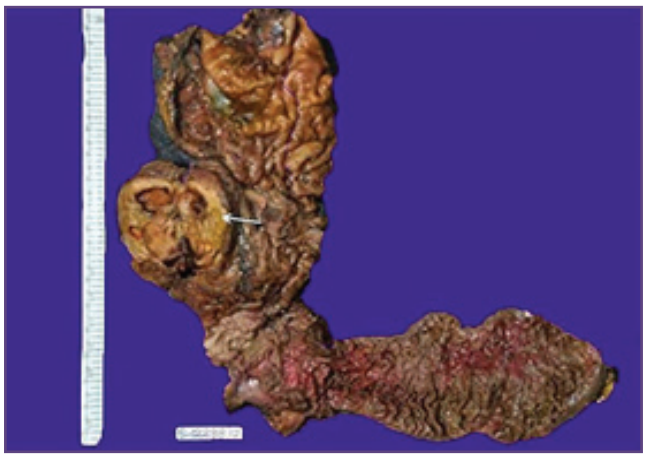

Fig. 1: Well circumscribed tumor with central areas of necrosis.

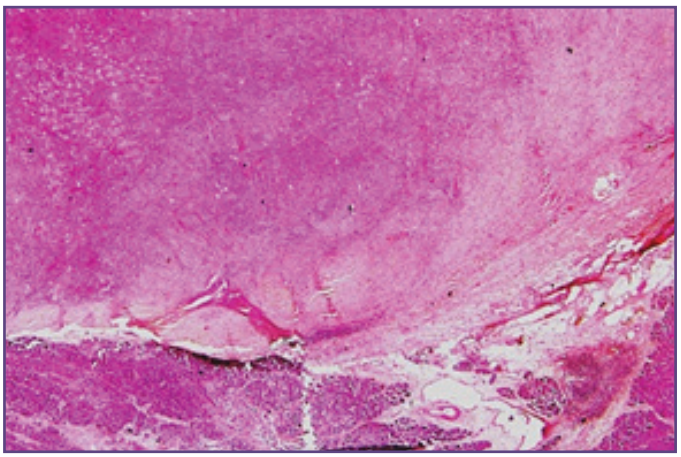

Fig. 2: Tumor with surrounding normal pancreatic tissue.

Table 1: Case reports with Pancreatic Schwannoma

S. No.	Author	Year	Sex	Age	Size in cm	Histology
01	Present study	2014	M	62	7cm	Benign
02	Devi Jetal2	2014	F	63	1cm	Benign
03	Moriya T etal1	2012	F	64	4cm	Benign
04	Lis etal7	2009	M	37	16cm	Benign
05	Hirabayashi K etal8	2008	M	51	6cn	Benign
06	Paranjape C etal5	2004	F	77	3.5cm	Benign

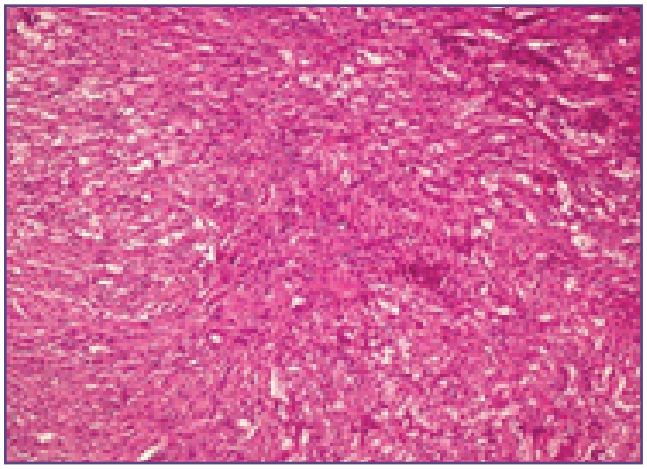

Fig. 3: Closely packed spindle cells with nuclear palisading and interspersed foamy macrophages in between the tumor cells.

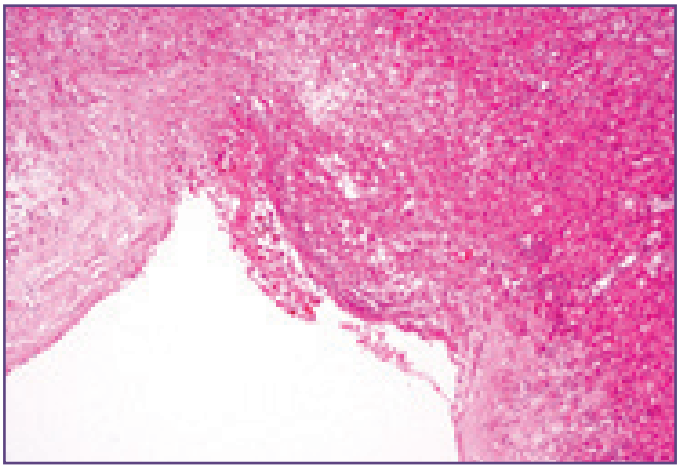

Fig. 4: Cystic areas with pigment laden macrophages.

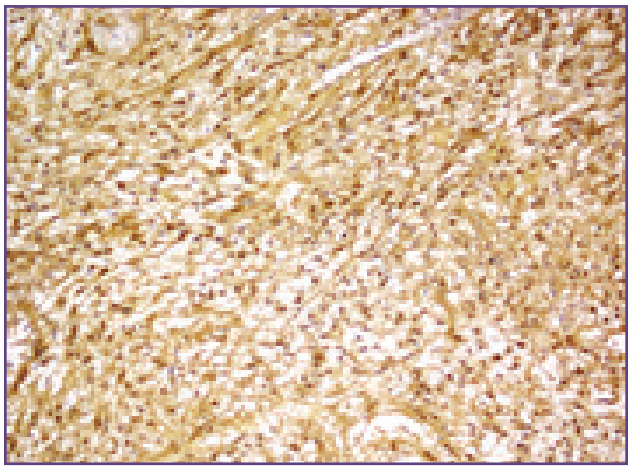

Fig. 5: Nuclear staining of the tumour cells with S-100 on immunohistochemistry.

Immunohistochemically, schwannomas are strongly positive for S-100 protein, vimentin and CD 56, while negative for other tumor markers including cytokeratin AE1/AE3, desmin, smooth muscle myosin, CD 34 and CD 117.[9]The symptoms from the reported cases of pancreatic schwannoma are variable. Majority (70%) of patients are symptomatic. Abdominal pain is the most common reported symptom (57%). Symptoms such as back pain (6%), nausea and/ vomiting (4%), weight loss (13%), melena (4%) and jaundice (4%) have been also reported. Thirty percent of patients are asymptomatic and the lesions are incidentally discovered on radiological examination scans performed for other reasons.[1] The preoperative diagnosis of pancreatic schwannoma is very difficult, especially in cystic schwannomas. The most characteristic feature on CT scan was the presence of an area of low density and/or cystic images reflecting the Antoni B component or degenerative cystic areas of the schwannoma.[6] Other pancreatic tumors often share imaging features with pancreatic schwannoma, and differential diagnoses should always be considered. Ultrasound-guided Fine Needle Aspiration (EUS-FNA) biopsy has been used increasingly at many institutions. This procedure may be useful for accurate preoperative diagnosis.[7,8].Since malignant transformation of pancreatic schwannomas is uncommon, simple enucleation is usually sufficient. A review of the treatment showed that the most common resection was pancreatico-duodenectomy (32%), followed by distal pancreatectomy (21%) and enucleation (15%). An intraoperative frozen section

should be performed, as it helpsto establish the diagnosis of a benign schwannoma and avoid more radical resection. Larger tumors, tumors involving portal vein, ampulla, or splenic hilum, may require a more radical resection than simple enucleation. Benign Schwannomas with cystic degeneration tend to be of larger size. Because of their larger size and resemblance with other cystic neoplasms, preoperative diagnosis may be challenging. [1]

Conclusion

Accurately diagnosing Pancreatic Schwannomas with cystic changes preoperatively is of paramount importance to avoid extended resection.

Funding

None

Competing Interest

None declared

Funding source

Nil

Presentation at Meeting

Nil

Conflict of Interest

There is no conflict of interest in this case report.

Pancreatic Schwannoma a rare entity.

References

1. Moriya T, Kimura W, Hirai I, Takeshita A, Tezuka K, Watanabe T etal. Pancreatic schwannoma: Case report and anupdated 30-year review of the literature yielding 47 cases. WorldJ Gastroenterol 2012; 18: 1538-44.
2. Devi J, Sathyalakshmi R, Chandramouleeswari K, Nalli R. Sumitra Devi. Pancreatic Schwannoma - A Rare Case Report. J Clin Diagn Res. 2014; 8: FD15–FD16.
3. Verocay J. Zur Kenntnis der “Neurofibrome”. Beitr Pathol Anat Allg Pathol 1910; 48: 1-69.
4. Almo KM, Traverso LW. Pancreatic schwannoma: an uncommon but important entity. J Gastrointest Surg 2001; 5:359-363.
5. David S, Barkin JS. Pancreatic schwannoma. Pancreas 1993; 8: 274-276.
6. Paranjape C, Johnson SR, Khwaja K, Goldman H, Kruskal JB, Hanto DW. Clinical characteristics, treatment, and outcome of pancreatic Schwannomas. J Gastrointest Surg 2004; 8:706-712.
7. Li S, Ai SZ, Owens C, Kulesza P. Intrapancreatic schwannoma diagnosed by endoscopic ultrasound-guided fine-needle aspiration cytology. Diagn Cytopathol 2009; 37: 132-135.
8. Hirabayashi K, Yasuda M, Umemura S, Itoh H, Itoh J, Yazawa N et al. Cytological features of the cystic fluid of pancreatic schwannoma with cystic degeneration. A case report. JOP 2008; 9: 203-208
9. Weiss SW, Langloss JM, Enzinger FM. Value of S-100 protein in the diagnosis of soft tissue tumors with particular reference to benign and malignant Schwann cell tumors. LabInvest 1983; 49: 299-3082.
10. Suzuki S, Kaji S, Koike N, Harada N, Hayashi T, Suzuki M et al. Pancreatic schwannoma: a case report and literature review with special reference to imaging features. JOP 2010; 11: 31-35.