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CASE REPORT

A case of primary pancreatic schwannoma diagnosed by endoscopic ultrasound-fine needle aspiration

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Abstract
Pancreatic schwannoma is difficult to diagnose preoperatively. A 79-year-old man was found to have a 9-mm pancreatic mass on abdominal ultrasonography. On EUS, there was a 9-mm, clearly demarcated, round, solid, hypo-echoic mass in the pancreatic body. The differential diagnosis included a pancreatic neuroendocrine tumor, a solid-pseudopapillary neoplasm, and an atypical pancreatic cancer. EUS-FNA was performed with a 22G needle. On pathology examination, spindle-shaped tumor cells were seen proliferating in bundles. On immunostaining, the lesion was negative for c-kit, CD34, and α-SMA but positive for S-100 protein. The MIB-1 index was < 2%. Based on the above findings, the lesion was diagnosed as a benign pancreatic schwannoma. We, therefore, decided to follow the patient with careful observation rather than resecting the lesion surgically. The tumor has not changed significantly after 3 years of follow-up. EUS-FNA is useful for the diagnosis of pancreatic schwannoma. If the tumor can be determined to be benign preoperatively, unnecessary surgery can be avoided. EUS-FNA should be actively implemented for pancreatic tumors that are difficult to diagnose definitively on imaging.

Keywords Pancreatic schwannoma · EUS-FNA

Introduction

Primary pancreatic schwannoma is a very rare tumor, with about 70 cases previously reported in the English literature [1–4]. It is usually a benign tumor that can be followed conservatively if diagnosed before surgery is undertaken, although most previous cases were diagnosed by surgical resection. However, there are a few reports of diagnosis by endoscopic ultrasound-fine needle aspiration (EUS-FNA). Here, we report a patient diagnosed with a benign pancreatic schwannoma by EUS-FNA, allowing us to avoid unnecessary surgery.

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Case presentation

A 79-year-old man had no remarkable past medical history other than a branch duct-type intraductal papillary mucinous neoplasm (IPMN) of the pancreatic tail that had been observed for 6 years. A routine abdominal ultrasound revealed a 9-mm, clearly demarcated, hypo-echoic mass in the pancreatic body (Fig. 1). On physical examination, there was no abdominal tenderness or palpable mass. Blood tests showed no increase in the pancreatic enzyme levels (amylase 125 U/l, lipase 56 U/l), and the CA19-9 and CEA levels were in the normal range. Abdominal contrast-enhanced computed tomography (CT) showed a 9-mm hypovascular tumor in the arterial phase and the lesion was slightly enhanced in the portal phase (Fig. 2). Magnetic resonance cholangiopancreatography (MRCP) showed a branch duct-type IPMN (17 mm) in the tail of the pancreas with no evidence of an increase in size from previous images. Magnetic resonance imaging (MRI) T2-weighted image showed a 9-mm high-intensity mass in the pancreas body. Diffusion-weighted images (DWI) showed slightly high intensity at the same site (Fig. 3). On EUS, there was a 9-mm, clearly demarcated, round, solid, hypo-echoic mass in the pancreatic body

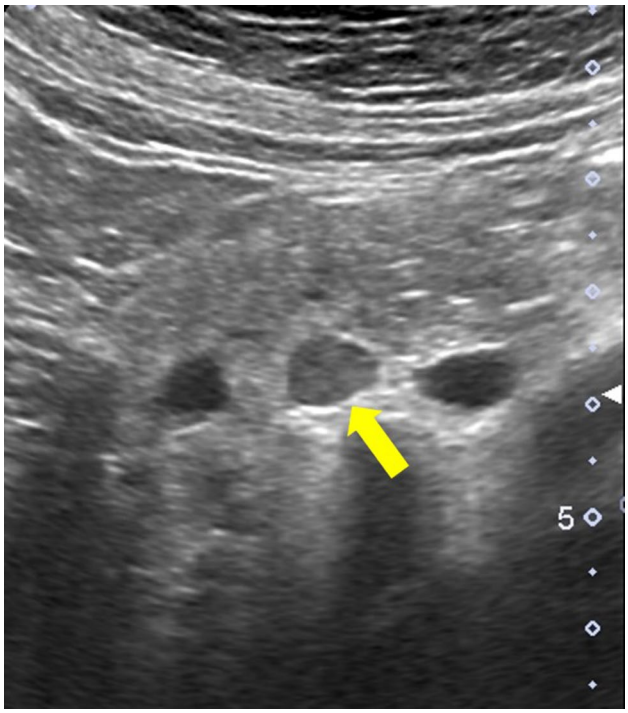

Fig. 1 Abdominal ultrasound revealed a 9-mm, clearly demarcated, hypo-echoic mass within the pancreatic body (arrow)

(Fig. 4). No blood flow within it was observed on Doppler. The mass was uniformly hypo-echoic without any cystic components or calcifications.

The differential diagnosis included a pancreatic neuroendocrine tumor (p-NET), a solid-pseudopapillary neoplasm (SPN), and an atypical pancreatic cancer. Two sessions of EUS-FNA using a 22G needle (Expect™ SlimLine, Boston Scientific Japan, Tokyo, Japan) with 20-mL suction syringe was performed (Fig. 4). On pathology examination, spindle-shaped tumor cells were seen proliferating in bundles. On immunostaining, the lesion was negative for c-kit, CD34, and α-SMA, but positive for S-100 protein. The MIB-1 index was < 2% (Fig. 5).

Based on the above findings, the lesion was diagnosed as a benign pancreatic schwannoma. After discussing options with the patient, he agreed with our recommendation to follow the lesion conservatively. The tumor has not changed significantly after 3 years of follow-up (Fig. 6).

Discussion

Schwannoma is a neurogenic tumor derived from Schwann cells of the peripheral nerves. It was first reported by Verocay et al. [1]. The most common sites are the head and neck, mediastinum, and retroperitoneum. Pancreatic schwannomas are rare [2]. Immunohistochemically, schwannomas stain strongly positive for S100 protein, which is specific to this tumor. In about two out of three cases, degenerative changes such as cystic degeneration, hemorrhage, calcification, and hyalinization are noted, resulting in variable imaging findings [3].

Ma et al. [4] summarized 68 cases of pancreatic schwannoma reported so far in English. The patients' average age was 55.7 years (20–87), with a nearly equal male-to-female ratio (44–56%). About one-third of patients were asymptomatic. For those who had symptoms, abdominal pain was most frequently reported (50%), followed by weight loss (18%), nausea and vomiting (8.8%), indigestion (7.4%), back pain (5.9%), and abdominal mass (4.4%). Anemia, bloody stools, and jaundice were rare, found in only 2.9%. The lesion was in the head of the pancreas in 53% of cases,

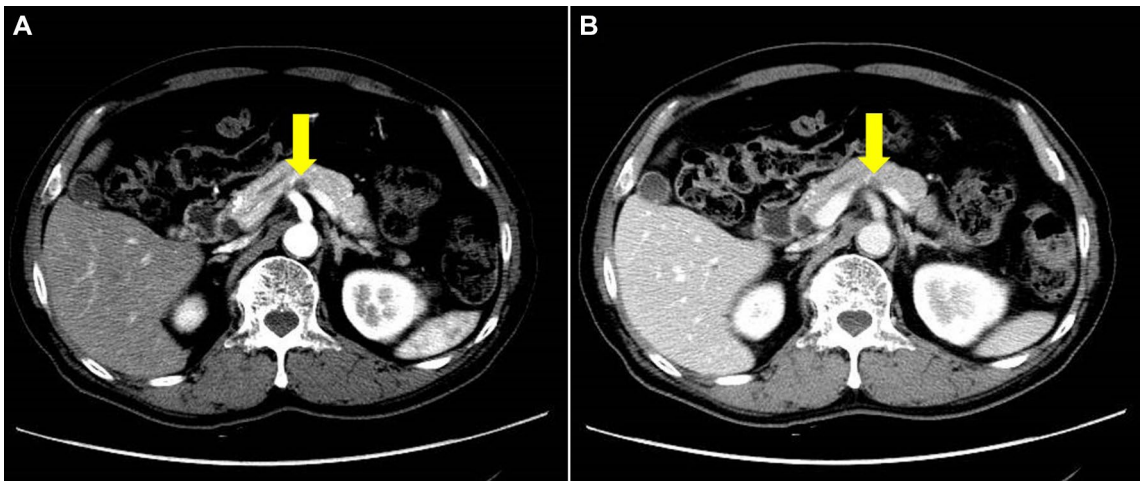

Fig. 2 Abdominal contrast-enhanced computed tomography (CT) showed a 9-mm hypovascular tumor in the arterial phase (arrow, **a**) and the lesion was slightly enhanced in the portal phase (arrow, **b**)

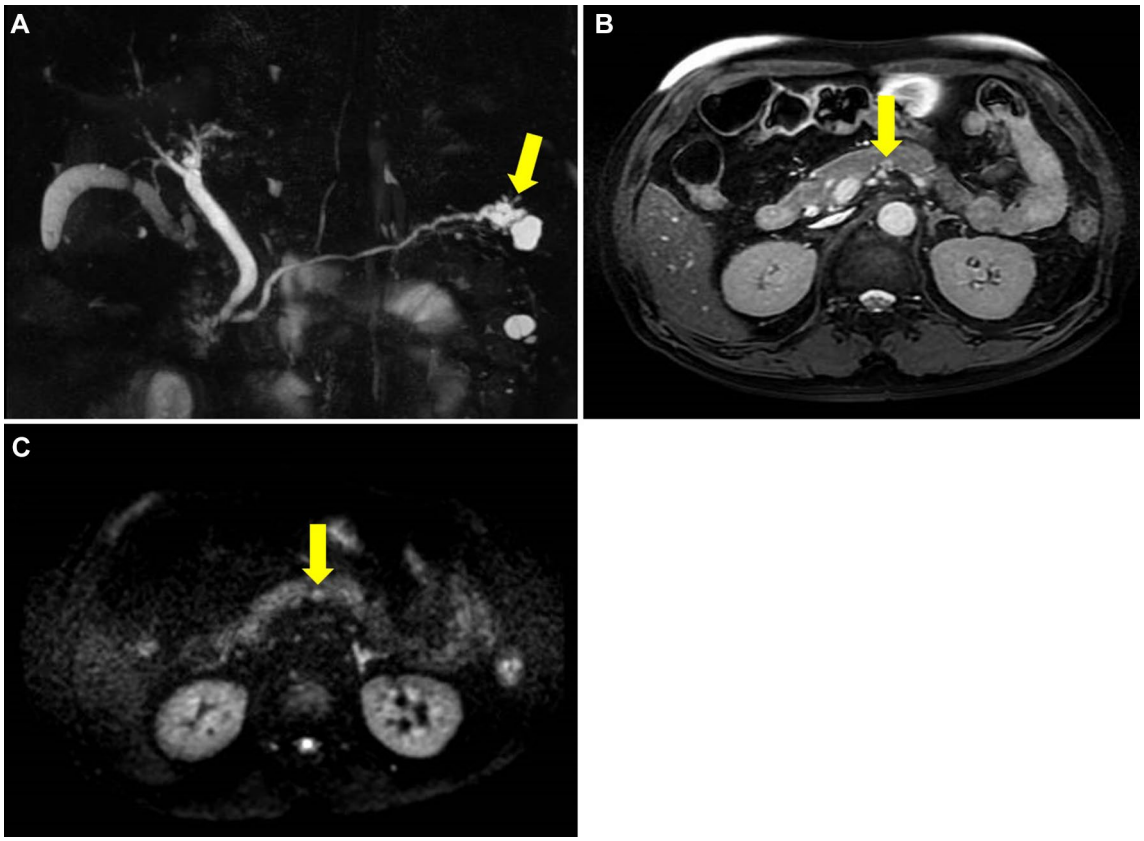

Fig. 3 **a** Magnetic resonance cholangiopancreatography (MRCP) showed a branch duct-type IPMN (17 mm) in the tail of the pancreas (arrow) with no evidence of an increase in size from previous images. **b** Magnetic resonance imaging (MRI) T2-weighted image showed a 9-mm high-intensity mass in the pancreas body (arrow). **c** Diffusion-weighted images (DWI) showed diffusion reduction at the same site (arrow)

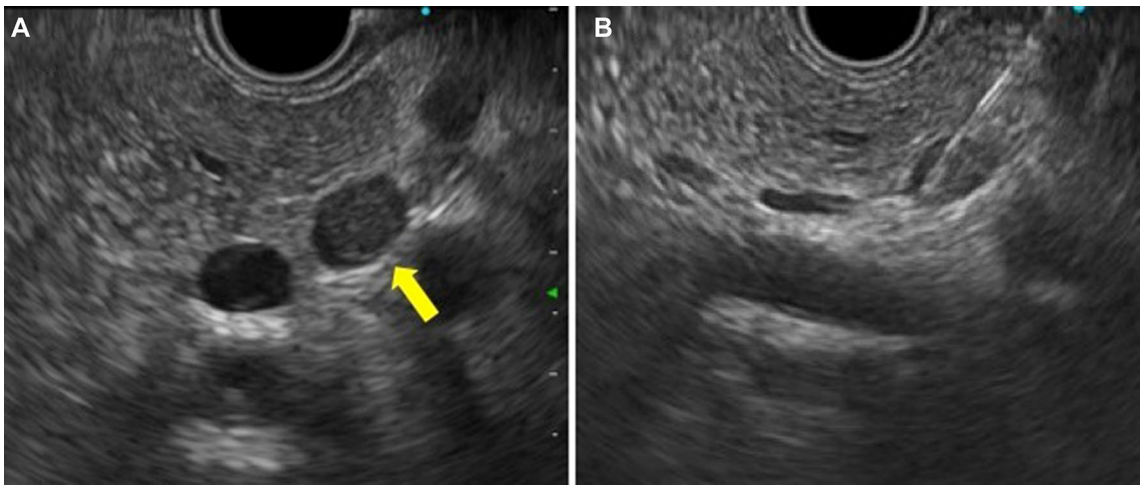

Fig. 4 **a** There was a 9-mm, clearly demarcated, round, solid, hypo-echoic mass in the pancreatic body on EUS (arrow). **b** Two sessions of EUS-FNA using a 22G needle (Expect™ SlimLine, Boston Scientific Japan, Tokyo, Japan) with 20-mL suction syringe was performed

body in 21%, tail in 9%, head and body in 4.5%, and body and tail in 10%.

On imaging, 28% of pancreatic schwannomas were solid, 43% cystic, and 22% solid and cystic mixed. Clear boundaries were present in 80% and calcifications were observed

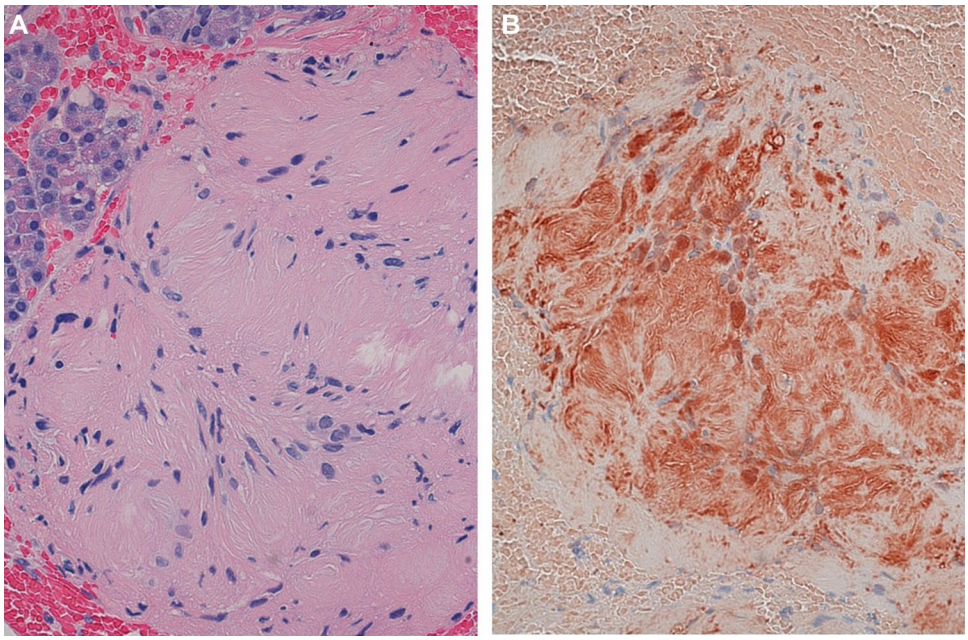

Fig. 5 **a** On pathology examination, spindle-shaped tumor cells were seen proliferating in bundles (H.E., 200×). *H.E.* hematoxylin and eosin. **b** The lesion was positive for S-100 protein (S-100 stain, 200×)

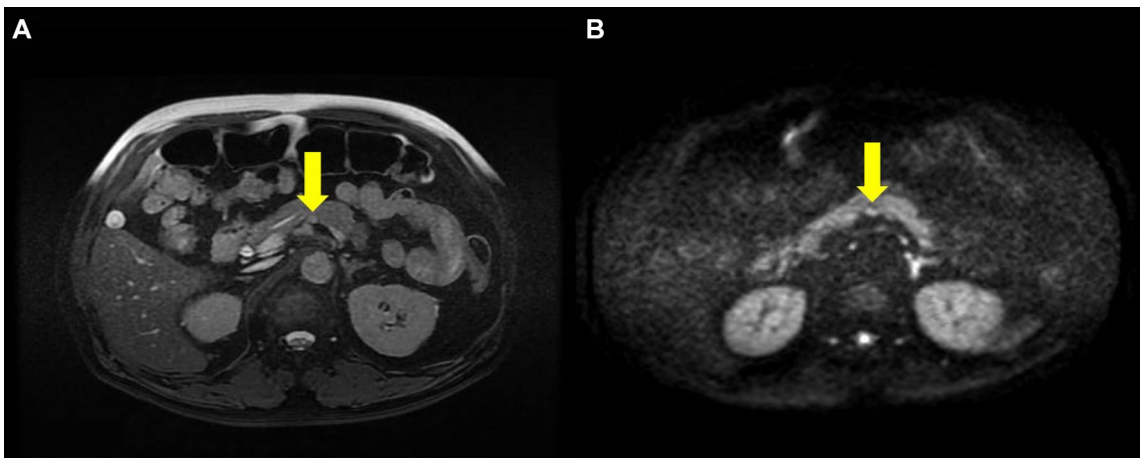

Fig. 6 **a** MRI taken 3 years after follow-up. T2-weighted image showed a 9-mm high-intensity mass in the pancreas body (arrow). **b** Diffusion-weighted images showed slightly high intensity at the same site (arrow). The tumor has not changed significantly after 3 years of follow-up

in only 8.6%. Only 20% of lesions were correctly diagnosed preoperatively, while remaining of them were diagnosed as serous cystic neoplasm (SCN), mucinous cystic neoplasm (MCN), SPN, p-NET, or pseudocyst [4].

Wang et al. summarize the features of imaging study reported so far. According to their reports, in US, 80% hypoechoic, 13% hyperechoic, 7% hyper- and hypo-echoic, in CT, 42% of cystic lesions, 20% of cyst and solid lesion, 25% of solid lesion, septal lesion is 13%, in the MRI (T1 weighted image) 92% low intensity, 8% high intensity, and the MRI (T2-weighted image) 8% low intensity, and 92% high intensity [5].

One of the reasons which preoperative diagnosis is difficult is the lack of characteristic clinical or imaging findings, unlike some other tumors. For example, pancreatic MCN is a cystic tumor that occurs in young to middle-aged women and is usually localized to the body–tail of the pancreas. On imaging, MCN is a cyst that may contain septations (so-called "cyst in cyst" pattern). In the typical example, therefore, diagnosis is not difficult. On the other hand, pancreatic schwannoma has no gender predilection and may occur at any site within the organ. Imaging features include cystic, solid, mixed, and various findings. This lack of specificity makes diagnosis difficult.

Differentiation between pancreatic schwannoma and tumors originating from outside the pancreas (schwannoma, epidermoid cyst, or para-ganglioma) is very difficult. In typical cases, a "beak sign" indicating the origin of the pancreas

Table 1 Literature cases of pancreatic schwannoma diagnosed by EUS-FNA

Case	Age/sex	Location	Tumor size (mm)	Gauge of puncture needle	Treatment	References	Author/publication year
1	44/F	Head	13	25	Follow-up (no change for 5 months)	6	Bruno M/2019
2	35/M	Body	70	N/A	Surgery	7	Crinò SF/2016
3	37/M	Body	16	22	Surgery	7	Crinò SF/2016
4	34/F	Head	70	22	Surgery	7	Crinò SF/2016
5	55/F	Head	10	25	Follow-up	7	Crinò SF/2016
6	83/M	Body	20	25	Follow-up	7	Crinò SF/2016
7	59/F	Head	21	25	Surgery	7	Crinò SF/2016
8	47/F	N/A	15	22	Follow-up (no change for 82 months)	8	Takasumi M/2017
9	72/F	Tail	60	N/A	Surgery	9	Hayashi K/2018
10	37/M	Head	16	22	Surgery	10	Moriya T/2012
11	43/M	Head	N/A	N/A	Surgery	11	Iwano K/2019
12	37/M	Body	13	N/A	Surgery	12	Ammar A/2019
13	79/M	Body	9	22	Follow-up (no change for 36 months)	Our case	

may be seen, but it is not always seen. In this case, the tumor was clearly surrounded by the pancreas by imaging examination (US/CT/MR/EUS) and we diagnosed it arising from pancreas.

Pancreatic schwannoma is usually benign, although of the 68 cases reviewed by Ma et al. [4], 8 (12%) were malignant. They noted that malignant tumors tended to be larger (benign, 5.2 cm vs. malignant, 14 cm) and invaded blood vessels and surrounding organs. The benign and malignant schwannomas did not differ in terms of age, sex, localization, or internal tumor characteristics (solid, cystic, mixed).

Because most pancreatic schwannomas are benign, unnecessary surgery can be avoided if the diagnosis can be made preoperatively.

Similar to this case, a small schwannoma was diagnosed by EUS-FNA, and there were reports of avoiding surgery [6]. If preoperative diagnosis is possible, follow-up is a reasonable option. On the other hand, because preoperative diagnosis is difficult, there is an opinion that actively recommends surgery [5]. However, pancreatectomy has serious complications such as pancreatic fluid leakage, bleeding, and infection, and there are cases in which insulin is required for diabetes after surgery. Unnecessary pancreatectomy should be avoided.

EUS-FNA is a promising powerful diagnostic tool due to the lack of characteristic image findings. Unfortunately, the accuracy rate of EUS-FNA for schwannomas has been reported to be 52.9%, which is not high [6]. If this case cannot be diagnosed by EUS-FNA, it is desirable to conduct a follow-up in a short period of time and to re-consider EUS-FNA if there is tumor growth or morphological change.

We searched PubMed using the terms "Pancreas," "Schwannoma," and "EUS-FNA." We targeted only cases that could be successfully diagnosed by EUS-FNA. The search yielded 12 cases [6–12]. Including ours, the median age of the 13 patients reported was 61.5 years (34–83). There were seven men and six women. The lesion was in the head of the pancreas in six cases, body in five, tail in one, and unknown in one. The puncture needle used was 22G in five cases, 25G in four, and not stated in four. Eight of the patients underwent surgery, while the other five were followed conservatively (Table 1).

Before the advent of EUS-FNA, the possibility of malignancy could not be ruled out without resecting the lesion surgically. In some cases, extensive resection such as pancreatoduodenectomy was done. However, because most schwannomas are benign, even if surgery is performed, enucleation is recommended. On the other hand, if it is possible to make the diagnosis preoperatively, then follow-up without surgery is also a reasonable option.

In our patient, the tumor characteristics suggested a benign lesion, that is, a small tumor diameter ($\leq$ 10 mm), lack of evidence of invasion of other organs, typical pathology findings, and a low MIB-1 index $\leq$ 2%). Therefore, we elected to follow the patient. Three years after diagnosis, no change was seen on imaging, strongly supporting the diagnosis of a benign schwannoma.

EUS-FNA is useful for the diagnosis of pancreatic schwannoma. If a preoperative diagnosis can be made, unnecessary surgery can be avoided. Our case illustrates that EUS-FNA is worth undertaking to evaluate pancreatic tumors that are difficult to diagnose definitively on imaging.

Compliance with ethical standards

Conflict of interest Tetsushi Azami, Yuichi Takano, Fumitaka Niiya, Takahiro Kobayashi, Eiichi Yamamura, Naotaka Maruoka, Tomoko

Norose, Nobuyuki Ohike, and Masatsugu Nagahama declare that they have no conflict of interest.

Human rights All procedures followed have been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

Informed consent Informed consent was obtained from all patients for being included in the study.

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