

Summary of reported cases of pancreatic paraganglioma from 1974 to 2022

No.	Reference	Year of publication	Sex	Age y	Signs/symptoms	Inspection Method	Features	FNA /FS	Location in the pancreas/size, cm	Preoperative diagnosis	Treatment	Functional	Perioperative medication
1	Cope C et al. (10)	1974	F	72	Physical findings	US	Low echo	-	Head; 13*11*7	PCN	TLR	no	no
2	Fujino Y et al. (11)	1998	M	61	Abdominal pain	CT MRI	A solid mass T1 and T2 equal signals	-	Uncinate process; 2.5*4.2*1.8	pNEN	PD	no	no
3	Parithivel VS et al. (12)	2000	M	85	Physical findings	CT	Shadow of a blood-rich, cystic solid mass	FS: NET	Head; 6.0	pNEN	TLR	no	no
4	Ohkawara T et	2005	F	72	Abdominal pain	CT	A solid-cystic	-	Head; 4.0	pNEN	RPH	no	no

al. (13)

tumor
rich in
blood
supply,
no
dilation
of either
biliary
or
pancreat
ic ducts

5	Kim SY et al. (14)	200 8	F	5 7	Lumbar discomfor t	US EUS CT	A hypervascu lar tumor Echogenic mass with several anechoic portions Marked enhanceme nt in the arterial phase, still well- enhancing	-	Head; 6.5*6.6	Panc reatic islet cell tumo r	PPPD	no	no
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							in the portal venous phase, non- enhancing tubular- shaped portions could be seen inside						
6	Tsuka da A et al. (15)	200 8	F	5 7	Physical findings	US CT	Low echo Significantl y stronger clumping shadow	-	Uncinate process; 2.5*2.0	pNE N	TLR	no	no
						MRI	T1 and T2 low signal						
7	Sangs ter G et al. (16)	201 0	M	5 0	Abdomin al pain	CT	Abundant blood supply; Strong uptake;	FNA: poorly differen tiated carcino ma	Uncinate proces	Panc reatic cance r	Surgery; radiother apy	no	no
						PET							
8	He J et al. (17)	201 1	F	4 0	Physical findings	CT	A firm arterioveno us phase	-	Head; 4.5*4.2	-	RPH	no	no

							markedly enhanced and well-defined mass shadow							
9	Lightfoot N et al. (18)	2011	M	66	Abdominal pain	CT PET	Cystic solid Mild metabolic elevation	-	Head and uncinate process; 6.0	-	PD	no	no	
10	Singhi AD et al. (19)	2011	F	61	Abdominal pain	-	-	FNA: pseudo cyst	Tail; 14.0	PCN	DP	no	no	
11	Singhi AD et al. (19)	2011	F	52	Abdominal pain	-	-	FNA: PGL	Body; 14.0	PGL	-	no	no	
12	Singhi AD et al. (19)	2011	F	54	Abdominal pain	-	-	FNA: PGL	Head; 6.5	PGL	DP	no	no	
13	Singhi AD et al. (19)	2011	M	40	Physical findings	-	-	FNA: NET	Body; 5.1	pNE N	TLR	no	no	

14	Singhi AD et al. (19)	2011	F	78	Abdominal pain	-	-	FNA: spindle cell neoplasm	Body; 17.0	-	-	no	no
15	Singhi AD et al. (19)	2011	M	44	Physical findings	-	-	FNA: PGL	Head; 5.5	PGL	-	no	no
16	Singhi AD et al. (19)	2011	M	38	Abdominal pain	-	-	-	Body; 15.0	-	-	no	no
17	Singhi AD et al. (19)	2011	M	47	Abdominal pain	-	-	-	Body; 7.5	pNE N	-	no	no
18	Singhi AD et al. (19)	2011	F	37	Abdominal pain	-	-	-	Tail; 5.7	pNE N	-	no	no
19	Higa B et al. (20)	2012	F	65	Physical findings	CT	A pancreatic mass with enhancement and	-	Uncinate process; 2.0	-	PD	no	no

20	Ganc RL et al. (21)	2012	F	37	Physical findings	EUS	mixed attenuation Low echo	FNA: NET	Head; 4.8*3.2*4.3	pNE N	PD	no	no
21	Al-Jiffry BO et al. (22)	2013	F	19	Abdominal pain	CT	Marked enhancement in the arterial phase and washed out in the venous phase	FNA: NET	Head and neck; 9*5*9.5	pNE N	PD	yes	Intraoperative α and β blockers were injected continuously
22	Borghain Met al. (23)	2013	F	55	Abdominal pain	US CT	Highly vascular A multicystic tumor	-	Tail; 17*19	Pancreatic sarcoma	PD	no	no
23	Straka Met al. (24)	2014	F	53	Abdominal discomfort	US CT	Abundant blood supply An extremely hypervascular	-	Head; 8.1*8.5	-	PPPD	no	no

							lar tumor with abundant collateral vessels from the superior mesenteric artery						
2 4	Zhan g L et al. (25)	201 4	F	5 0	Intermitte nt hypertens ion, headache, sweating, palpitatio ns	CT	A solid tumour with abundant blood vessels and multiple liver metastases	FNA: PGL	Head; 6.0	PGL	-	yes	Postoperat ive 5- fluorouraci l chemother apy, mithramyc in and adriamyci n treatment
2 5	Zhan g L et al. (25)	201 4	M	6 3	Hyperten sion	CT PET	Well- vascularize d tumor Abnormalu ptake	-	Head; 4.0	PGL	surgery	yes	Preoperati ve basal blood pressure lowering
2	Meng	201	F	5	Abdomin	US	An ill-	-	Head; 3*2.5	-	surgery	no	no

							d tumor, strongly enhanced in the arterial phase and still faintly enhanced in the portal vein phase. no dilation of either biliary or pancreatic ducts							
2 9	Ünver M et al. (28)	201 5	F	4 1	Loss of appetite, weight loss, weakness, difficulty breathing during exercise	US MRI	Pancreatic tail and splenic hilum mass Mass invading splenic hilum	-	Tail; 8*7*8	-	TLR	no	no	
3	Gines	201	M	5	Lumbar	CEUS	Arterial	-	Uncinate	pNE	PPPD	no	no	

0	u GC et al. (29)	6		5	pain	and CT	phase hypervascu- larity and slow wash- out		process; 1.3	N				
3 1	Tumu luru S et al. (30)	201 6	F	6 2	Physical findings	CT	Well- defined margin and avid homogene- ous enhanceme- nt. No lymphaden- opathy or metastasis	-	Body; 2.8*2.8*2.7	pNE N	PD	no	no	
3 2	Lin S et al. (31)	201 6	F	4 2	Abdomin- al pain	CT	A solid, low- density, arterial- phase- enhancing mass	-	Body; 5.2*6.3	pNE N	MP	no	no	
3 3	Liang W, Xu S (32)	201 6	M	4 1	Physical findings	CT	Arterioven- ous phase enhanceme	-	Uncinate process;4*6	pNE N	PD	no	no	

							nt (low attenuation)						
						MRI	T1 low signal; T2 slightly high signal; inhomo- geneous enhanceme nt						
34	Furce a L et al. (33)	2017	F	53	Dyspepsi a	US	Inhomogen eous low echo	-	Isthmus; 3.5*2.5*2.5	pNE N	MP	no	no
						CEUS	Intense arterial enhanceme nt, rich arterial vasculariza tion						
35	Nona ka K et al. (34)	2017	F	68	Physical findings	EUS	Low-echoic nodule. No dilation of either biliary or	-	Head; 2.2*2.2*1.7	pNE N	PPPD	no	no

						CT	pancreatic ducts. Enhanced during the arterial phase, weakly enhanced during the portal phase						
						PET	High intake; malignant possibility						
3	Zeng	201	F	5	Abdomin	EUS	Hypoechoic and well-defined mass	FNA: NET	Body; 6.5*6.1*4.4	pNE N	TLR	no	no
6	J et al. (35)	7		8	al pain								
						MRI	Inhomogeneous signal						
3	Zeng	201	F	5	Abdomin	CT	Soft tissue mass	FNA: NET	Head; 2.5*1.7*1.8	pNE N	Surgery	no	no
7	J et al. (35)	7		3	al pain								
3	Nguy	201	F	7	Constipat	EUS	Low echo	FNA:	Tail; 3.6*5*4.5	pNE	TLR	no	no

8	en E et al. (36)	8		0	ion; early satiety	CT	Cystic solid	NET?		N			
3 9	Fite JJ, Malek i Z. (37)	201 8	M	4 0	Lumbar pain; haematur ia	Radiolo gic impress	Abundant blood supply with necrosis	FNA: NET	Peripancreati c; 5.1	pNE N	Surgery	no	no
4 0	Fite JJ, Malek i Z. (37)	201 8	F	2 3	Palpitatio ns	Radiolo gic impress	Heterogene ous mass	FNA: NET	Peripancreati c; 7.0	pNE N	Surgery	no	no
4 1	Chatt oraj AK et al. (38)	201 9	F	3 6	Abdomin al pain	CT	No enhanceme nt	-	Peripancreati c; 7*4	GIST	TLR	no	no
4 2	Zong o N et al. (39)	201 9	M	5 2	Abdomin al pain	CT	Inhomogen eous density	-	Tail; 8.6*8.3	-	DP	yes	Intraoperat ive use of nimodipin e
4 3	Abbas i A et al. (40)	202 0	F	6 1	Physical findings	MRI	T2 hyperinten se; arterial phase enhanceme	FNA: NET	Head and uncinate process; 7.2*6.5	pNE N	PD	yes	no

44	Jiang CN et al. (41)	2021	M	41	Physical findings	EUS CT	nt Low echo Solid; inhomogeneous	FNA: malignant possibility; FS: NET	Body; 4.0*3.9	pNE N	DP	no	no
45	Lanke G et al. (42)	2021	F	73	Physical findings	EUS	Low echo	FNA: PGL	Head; 2.0*1.1	PGL	-	no	no
46	Park, J et al. (43)	2021	M	46	Abdominal discomfort	EUS CT MRI	A well-defined hypoechoic mass An irregular solid mass with marked enhancement in the arterial and portal phases Uneven	FNA: NET or PGL?	Head; 2.8*2.6*3.2	pNE N	PPPD	no	no

							enhancement in the arterial phase, gradual and weak enhancement in the portal and migratory phases, 'pepper appearance' in T1WI and T2WI.							
47	Wang W et al. (44)	2021	F	75	Abdominal discomfort	CT	Abundant blood supply; arterial phase enhancement	-	Neck; 3.1*3.8	PGL; Castleman; pNE N	TLR	no	no	
						MRI	T1WI low or equal signal; T2WI high							

							signal; obvious enhanceme nt						
48	Kim M et al. (45)	2022	M	64	Hip pain	CT	Ovoid, solid, enhanced visualisatio n;	FNA: Atypica l epitheli oid and spindle cell tumour s	Head; 11	Atyp ical epith eloid and spin dle cell tumo urs	TLR	yes	no
						Genetic testing	A RET gene variant of unknown significanc e [c.731C>T (p.T244I)]						
49	Li T et al. (46)	2022	F	26	Abdomin al pain	CT MRI	Mixed density Low signal	-	Body and tail; 8.0*5.0*4.0	pNE N	DP	no	no

							in T1WI, slightly high signal in T2WI						
50	Zhao Z et al. (47)	202 2	F	49	Abdomin al pain	CT MRCP	Low- density mass with marked enhanceme nt in the arterial and portal venous phases; T1 and T2 irregular signal shadows, strong signal on DWI, inhomogen eous enhanceme nt scans	-	Body and neck; 5.0*3.2*4.7	PGL	TLR	no	Preoperati ve use of Phenoxybe nzamine

–: not available or unknown.

Abbreviations: F: female; M: male; CT: computed tomography; CEUS: contrast-enhanced ultrasound; EUS: Endoscopic Ultrasonography; US: ultrasound; CDFI: Color Doppler Flow Imaging; MRCP: Magnetic Resonance Cholangiopancreatography; FNA: fine needle aspiration; FS: frozen section; NET: neuroendocrine tumour; PCN: pancreatic cystadenoma; PNEN: Pancreatic Neuroendocrine Neoplasm; GIST: gastrointestinal stromal tumour;TLR: tumour local resection; PD: pancreaticoduodenectomy; RPH: resection of the pancreas head; PPPD: pylorus preserving pancreaticoduodenectomy; DP: distal pancreatectomy; MP: Middle Pancreatectomy