January 2009

Autoimmune Pancreatitis: Disease Evolution, Staging, Response Assessment, and CT Features That Predict Response to Corticosteroid Therapy

Dushyant V. Sahani

Nisha I. Sainani

Vikram Deshpande

Mehrine S. Shaikh

Dmitry L. Frinkelberg

See next page for additional authors

Authors
Dushyant V. Sahani, Nisha I. Sainani, Vikram Deshpande, Mehrine S. Shaikh, Dmitry L. Frinkelberg, and Carlos Fernandez-del Castillo

Autoimmune Pancreatitis: Disease Evolution, Staging, Response Assessment, and CT Features That Predict Response to Corticosteroid Therapy[1]

Dushyant V. Sahani, MD
Nisha I. Sainani, MD
Vikram Deshpande, MD
Mehrine S. Shaikh, MBBS
Dmitry L. Frinkelberg, MD
Carlos Fernandez–del Castillo, MD

Purpose: To evaluate the evolution of morphologic features of autoimmune pancreatitis (AIP) at computed tomography (CT) and to identify imaging features that can predict AIP response to corticosteroid therapy (CST).

Materials and Methods: This HIPAA-compliant retrospective study had institutional review board approval. From among a cohort of 63 patients with AIP, 15 patients (12 men, three women; mean age, 64.7 years; age range, 30–84 years) who underwent sequential CT examinations before treatment were included to assess the evolution of disease by reviewing pancreatic, peripancreatic, and ductal changes. Of these patients, 13 received CST and underwent posttreatment CT; these CT studies were evaluated to determine if there were imaging features that could predict response to CST.

Results: The disease evolved from changes of diffuse (14 of 15 patients) or focal (one of 15 patients) parenchymal swelling, peripancreatic stranding (10 of 15 patients), "halo" (nine of 15 patients), pancreatic duct changes (15 of 15 patients), and distal common bile duct narrowing (12 of 15 patients) to either resolution or development of ductal strictures and/or focal masslike swelling. In 13 patients treated with CST, favorable response to treatment was seen in those with diffuse pancreatic and peripancreatic changes. Suboptimal response was seen in patients with ductal stricture formation (two of 13 patients) and in those in whom focal masslike swellings persisted after resolution of diffuse changes (seven of 13 patients).

Conclusion: CT features like diffuse swelling and halo respond favorably to CST and likely reflect an early inflammatory phase, whereas features like ductal strictures and focal masslike swelling are predictive of a suboptimal response and symbolize a late stage with predominance of fibrosis.

Supplemental material: *http://radiology.rsnajnls.org/cgi/content/full/2493080279/DC1*

[1] From the Departments of Radiology (D.V.S., N.I.S.), Pathology (V.D.), Gastroenterology (D.L.F.), and Surgery (C.F.), Massachusetts General Hospital–Harvard Medical School, 55 Fruit St, White 270, Boston, MA 02114; and Aga Khan University, Karachi, Pakistan (M.S.S.). Received February 11, 2008; revision requested April 22; revision received June 10; accepted June 17; final version accepted July 11. **Address correspondence to** D.V.S. (e-mail: *dsahani@partners.org*).

Autoimmune pancreatitis (AIP) is being increasingly recognized because of growing awareness of the disease and improved understanding of its imaging features. Surgical resection, which was once frequently performed for management of AIP, has provided an opportunity to study the histopathologic features of a number of specimens (1–4). This, in turn, has led to an improved understanding of the disease's biology and to increased use of corticosteroid therapy (CST) (5). The varied clinical manifestations of AIP range from mild, nonspecific complaints to the more ominous symptom of obstructive jaundice mimicking pancreatic malignancy (6). Although clinically the disease might resemble malignancy of the pancreas or, occasionally, other forms of pancreatitis, the characteristic imaging, clinical, and laboratory features that have been described, as well as its response to CST, have allowed classification of this pathologic process as a separate disease entity (6). A recent study (7) investigated the benefits of CST over mere observation in terms of complete remission rate, recurrence rate, and period of time needed to achieve complete remission, and clear advantages with the use of CST were found; CST has therefore been recommended as a standard initial therapy for AIP.

Advances in Knowledge

- Features of autoimmune pancreatitis (AIP) at CT reflect various phases of disease evolution; the features associated with inflammatory changes of the disease can resolve completely with timely institution of corticosteroid therapy (CST).
- Response to CST can be monitored with CT and improvement in disease status can be measured within a few weeks of the institution of CST.
- Improvements in imaging features at CT are also supported by the resolution of clinical symptoms and laboratory findings.

The features of AIP at computed tomography (CT) have also been reported to be quite varied. The presence of diffuse or focal enlargement of the pancreas, with or without "halo," and the presence of featureless homogeneous morphology are considered characteristic of AIP (8–11). In few cases, a focal masslike swelling, often in the pancreatic head, can be present (8–12). Other features, such as retraction of the pancreatic tail, segmental or diffuse narrowing of the pancreatic duct (PD), involvement of the distal common bile duct (CBD), and multiple cholangitis-like bile duct strictures, have been described (8,9,11,13). This spectrum of imaging findings raises the question of a sequential progression of the disease over time. Moreover, the response of AIP to CST has also been variable, from complete resolution of symptoms and imaging findings to variable degrees of partial response and recurrent disease (14–19). We hypothesize that these differences in imaging features and responses to CST may be related to different phases of AIP. Understanding the sequence of events associated with AIP can enable early diagnosis of the disease and identify morphologic features that can predict response to medical treatment.

We undertook this retrospective analysis to evaluate the evolution of morphologic features of AIP at CT and to identify imaging features that can predict AIP response to CST.

Implications for Patient Care

- Recognition of certain imaging features such as diffuse pancreatic swelling and peripancreatic halo and/or stranding at CT can predict a favorable outcome after CST and therefore can be crucial in deciding appropriate care and avoiding surgery for these patients.
- AIP is prone to chronicity and recurrence, which can be suggested by imaging features such as either partial resolution of swelling, loss of lobularity, or halo or reappearance of these findings after an initial response; this appearance of chronicity or recurrence can determine the need for a longer or repeat dosage of corticosteroids at appropriate times to achieve complete resolution.

Materials and Methods

Patient Selection

The institutional review board of our hospital approved this Health Insurance Portability and Accountability Act–compliant retrospective study and waived the need for informed consent. An electronic database of 63 patients with a diagnosis of AIP on the basis of the combination of clinical findings and imaging features (CT or magnetic resonance imaging) that was supported by laboratory parameters, results of endoscopic retrograde cholangiopancreatography (ERCP), and/or results of endoscopic ultrasonography (US) with fine-needle aspiration cytology (14) in our hospital from July 1995 to July 2007 was retrospectively reviewed (N.I.S., with 6 years of experience). The diagnosis in these patients was confirmed with percutaneous biopsy (n = 18), analysis of surgical specimens (n = 25), or re-

Published online before print
10.1148/radiol.2493080279

Radiology 2009; 250:118–129

Abbreviations:
AIP = autoimmune pancreatitis
CBD = common bile duct
CST = corticosteroid therapy
ERCP = endoscopic retrograde cholangiopancreatography
IgG = immunoglobulin G
IgG4 = immunoglobulin G4
PD = pancreatic duct

Author contributions:
Guarantors of integrity of entire study, D.V.S., C.F.; study concepts/study design or data acquisition or data analysis/interpretation, all authors; manuscript drafting or manuscript revision for important intellectual content, all authors; manuscript final version approval, all authors; literature research, N.I.S., M.S.S., D.L.F.; clinical studies, D.V.S., N.I.S., V.D., M.S.S., D.L.F., C.F.; statistical analysis, N.I.S., M.S.S., D.L.F.; and manuscript editing, D.V.S., V.D., C.F.

Authors stated no financial relationship to disclose.

Table 1

Findings at Pre-CST CT Examinations Showing Evolution of AIP in 15 Patients

Patient No./ Age (y)/Sex	Time Frame*	Clinical Features†	Serologic Results		Parenchymal Changes					Peripancreatic Changes		Ductal Changes	
			IgG Level‡	IgG4 Level§	Swelling	Enhancement	Lobularity	Focal Masslike Swelling	Tail Cutoff	Stranding	Halo	PD	CBD‖
1/71/M	Baseline	1, 2, 3, 4	++	++++	Diffuse	Heterogeneous	Featureless	Absent	Present	Absent	Absent	Diffuse attenuation	DNPD
	4 Mo	1, 2, 3, 4	++	++++	Increase	Increase	Increase	Absent	No change	Absent	4 mm	No change	Stent
2/57/F	Baseline	1, 2, 3	N	NE	Diffuse	Homogeneous	Featureless	Head	Present	Absent	Absent	Diffuse attenuation	DNPD
	1 Mo	1, 2, 3	NE	NE	Increase	No change	Increase	No change	Increase	Absent	Absent	No change	Stent
3/58/M#	Baseline	1, 2, 3, 4	N	+	Diffuse	Heterogeneous	Featureless	Absent	Absent	Absent	6 mm	Diffuse attenuation	DNPD
	2 Mo	1, 2, 3, 4	N	+	Increase	No change	Increase	Absent	Present	Absent	6 mm	Increase	No change
	6 Mo	1, 2, 3, 4	N	++	Increase	Increase	Increase	Head	Increase	Absent	8 mm	No change	Increase
	6 Mo	1, 2	N	++	No change	No change	No change	No change	Increase	Absent	10 mm	No change	No change
4/77/M**	Baseline	1, 3, 4	N	+	Diffuse	Heterogeneous	Featureless	Head	Present	Present	4 mm	Prominent	DNPD
	1 Mo	1, 3, 4	N	+	Increase	Increase	Increase	No change	No change	Increase	6 mm	Diffuse attenuation	No change
5/30/M	Baseline	1, 3, 4, 5	NE	NE	Diffuse	Homogeneous	Featureless	Absent	Absent	Absent	Absent	Diffuse attenuation	Absent
	2 Mo	1, 3, 4, 5	NE	NE	Diffuse	No change	No change	Absent	Present	Absent	Absent	No change	Absent
	2 Mo	1, 3, 4, 5	NE	NE	Increase	No change	Increase	Absent	Increase	Absent	Absent	Increase	Absent
	3 Mo	1, 3, 4, 5	NE	NE	No change	No change	No change	Absent	No change	Absent	Absent	No change	Absent
6/65/M	Baseline	2	N	++	Diffuse	Heterogeneous	Featureless	Head	Present	Present	4 mm	Irregular attenuation	DNPD
	2 Mo	2	N	+++	No change	No change	No change	No change	No change	No change	4 mm	No change	No change
	2 Mo	2	N	+++	Increase	No change	Increase	No change	Increase	Increase	6 mm	Increase	Increase
	6 Mo	2	N	+++	Increase	Increase	Increase	No change	No change	No change	6 mm	No change	No change
7/84/F#	Baseline	1, 2, 4, 5	N	++	Diffuse	Homogeneous	Normal	Absent	Present	Present	1 mm	Diffuse attenuation	DNPD
	2 Mo	1, 2, 4, 5	N	++	No change	No change	No change	Absent	No change	No change	3 mm	No change	No change
	2 Mo	1, 2, 4, 5	NE	NE	Increase	No change	Featureless	Absent	No change	Increase	5 mm	No change	Stent
8/72/M	Baseline	2, 3, 4, 5, 6	NE	NE	Normal	Normal	Normal	Tail	Absent	Absent	Absent	Diffuse attenuation	DNPD
	9 Mo	2, 3, 4, 5, 6	++	+++++	Diffuse	Heterogeneous	Partial	No change	Present	Present	1 mm	No change	No change
	10 Mo	2, 3, 4, 5, 6	NE	NE	Increase	Increase	Featureless	Absent	No change	Increase	3 mm	Increase	Stent
	12 Mo	2, 3, 4, 5, 6	NE	NE	Increase	Increase	Increase	Absent	No change	Increase	5 mm	Increase	Stent
9/83/M	Baseline	1, 3, 5	N	+	Diffuse	Heterogeneous	Partial	Absent	Absent	Present	Absent	Diffuse attenuation	DNPD
	2 Mo	1, 3, 5	N	+	Increase	Increase	Featureless	Absent	Present	Increase	Absent	No change	Stent
10/61/M	Baseline	1, 2, 5	N	N	Diffuse	Homogeneous	Partial	Absent	Present	Present	Absent	Diffuse attenuation	DNPD
	1 Mo	1, 2, 5	NE	NE	Increase	No change	Featureless	Absent	No change	No change	Absent	No change	No change
11/41/M	Baseline	3, 6, 7	N	+	Diffuse	Heterogeneous	Featureless	Head	Present	Present	3 mm	Diffuse attenuation	DNPD
	1 Mo	3, 6, 7	N	+	Increase	Increase	Increase	No change	Increase	Increase	5 mm	No change	No change
12/64/F	Baseline	1	+	NE	Diffuse	Heterogeneous	Partial	Head	Absent	Present	Absent	Prominent	Absent
	6 Mo	1	+	NE	No change	No change	No change	No change	Absent	No change	Absent	No change	Absent
	7 Mo	1	NE	NE	Increase	Increase	Featureless	No change	Present	No change	Absent	Diffuse attenuation	Absent
13/61/M**	Baseline	1, 2, 3	N	NE	Fullness	Heterogeneous	Normal	Head	Present	Absent	Absent	Prominent	DNPD
	2 Mo	1, 2, 3	NE	NE	No change	No change	Normal	No change	No change	Absent	Absent	Diffuse attenuation	Stent
14/78/M††	Baseline	1, 2	N	NE	Diffuse	Homogeneous	Partial	Head	Absent	Present	Absent	Diffuse attenuation	DNPD
	1 Mo	1, 2	N	NE	No change	No change	No change	No change	Present	No change	3 mm	No change	Stent
	1 Mo	1, 2	NE	NE	No change	No change	Featureless	No change	No change	No change	3 mm	No change	Stent

(Table 1 continues)

Table 1 (continued)

Findings at Pre-CST CT Examinations Showing Evolution of AIP in 15 Patients

Patient No./ Age (y)/Sex	Time Frame*	Clinical Features†	Serologic Results		Parenchymal Changes					Peripancreatic Changes		Ductal Changes	
			IgG Level‡	IgG4 Level§	Swelling	Enhancement	Lobularity	Focal Masslike Swelling	Tail Cutoff	Stranding	Halo	PD	CBD‖
15/69/M‡‡	Baseline	8	NE	NE	Diffuse	Heterogeneous	Partial	Tail	Absent	Present	2 mm	Prominent	Absent
	1 Mo		NE	NE	No change	No change	No change	No change	Absent	No change	2 mm	Diffuse attenuation	Absent
	1 Mo		NE	NE	No change	No change	No change	No change	Absent	No change	2 mm	No change	Absent

Note.—"Increase," "Decrease," and "No change" are relative to the findings at the previous examination. NE = not examined.

* Time interval in months for each follow-up CT examination is in relation to the preceding examination and not the baseline examination.

† 1 = Abdominal pain, 2 = jaundice, 3 = fatigue, 4 = weight loss, 5 = nausea and/or vomiting, 6 = pruritus, 7 = back pain, 8 = AIP incidentally detected at CT.

‡ N = normal (600–1500 mg/dL [6.0–15.0 g/L]), + = 1501–2000 mg/dL (15.1–20.0 g/L), ++ = >2000 mg/dL (>20.0 g/L).

§ N = normal (8–140 mg/dL [0.08–1.40 g/L]), + = 141–340 mg/dL (1.41–3.40 g/L), ++ = 341–540 mg/dL (3.41–5.40 g/L), +++ = 541–740 mg/dL (5.41–7.40 g/L), ++++ = 741–940 mg/dL (7.41–9.40 g/L), +++++ = >940 mg/dL (>9.40 g/L).

‖ DNPD = distal narrowing with proximal dilatation.

History of known non–insulin-dependent diabetes mellitus.

** New onset of non–insulin-dependent diabetes mellitus.

†† This patient underwent choledochojejunostomy with biopsy 2 months after baseline.

‡‡ This patient underwent distal pancreatectomy 2 months after baseline.

sponse to CST at follow-up (n = 20). Patients either were medically treated with CST (n = 25), underwent surgery (n = 25), or did not receive any treatment (n = 13). The radiology department, in collaboration with the surgical, endoscopy, and pathology departments, had been maintaining a database since 2000, and cases had been added to the database as and when a diagnosis was made. Fifteen patients who underwent pretreatment serial contrast material–enhanced CT examinations were identified and were included in this study for evaluating the evolution of CT features before CST. Of these patients, 13 were treated with CST; data in these patients were used to assess response to CST.

Patients and Treatment Strategies

The 15 patients with serial CT studies included in this investigation ranged in age from 30 to 84 years (mean age, 64.7 years). Twelve patients were men (mean age, 63.8 years; age range, 30–83 years), and three were women (mean age, 68.3 years; age range, 57–84 years). Thirteen of these patients were treated with CST, and two underwent surgery. The protocol for CST therapy included 40 mg of prednisone per day for 4 weeks, followed by gradual tapering by 5 mg every 2 weeks until the dose reached 15 mg per day and then gradual tapering again by 2.5 mg every 4–8 weeks. Three of the 13 patients who received CST later underwent surgery because of persisting clinical symptoms. Two patients were treated solely with surgery after follow-up examinations because of a clinical suspicion of a mass in the pancreatic head (choledochojejunostomy with biopsy of head lesion) or tail (distal pancreatectomy).

Clinical Features and Laboratory Data

Clinical symptoms at presentation such as abdominal pain, jaundice, weight loss, and their improvement or exacerbation after CST were recorded in a standardized database (N.I.S.). Any history of previous pancreatitis, alcohol abuse, and autoimmune disease was also recorded. Serum immunoglobulin G (IgG) and immunoglobulin G4 (IgG4) levels were

Table 2

Findings at Post-CST CT Examinations for Response Assessment in 13 Patients

Patient No. and Time Frame (mo)*	Clinical Features†	Serologic Results		Parenchymal Changes					Peripancreatic Changes		Ductal Changes	
		IgG Level‡	IgG4 Level§	Swelling	Enhancement	Lobularity	Focal Masslike Swelling	Tail Cutoff	Stranding	Halo	PD	CBD
1												
2	1, 2	N	+	Decrease	Decrease	Decrease	Absent	Normal	Absent	Normal	Decrease	Stent
3	N	N	+	Normal	Normal	Normal	Absent	Normal	Absent	Normal	Normal	Stent
3	N	N	N	Normal	Normal	Normal	Absent	Normal	Absent	Normal	Normal	Normal
2												
2	N	NE	NE	Decrease	No change	Decrease	No change	No change	Absent	Absent	Decrease	Decrease, stent
3‖												
3	1, 2	N	+	Decrease	No change	No change	No change	No change	Absent	10 mm	Decrease	Decrease
6	1	N	+	Decrease	Decrease	Decrease	No change	No change	Absent	4 mm	Decrease	Decrease
4#												
2	N	N	NE	Decrease	Decrease	Decrease	Decrease	No change	Normal	Normal	Normal	Normal
2	N	N	NE	Normal	Normal	Normal	No change	No change	Normal	Normal	Normal	Normal
5												
4	1, 5	NE	NE	Decrease	No change	Decrease	Absent	No change	Absent	Absent	Decrease	Absent
3	N	NE	NE	No change	No change	No change	Absent	No change	Absent	Absent	No change	Absent
6												
1.5	2 (Decrease)	N	+	Decrease	No change	Decrease	Decrease	Decrease	Decrease	4 mm	Decrease	Decrease
4**	2	N	N	No change	No change	No change	Tail	Normal	Normal	Normal	No change	No change
2	2 (Decrease)	NE	NE	Decrease	Decrease	Decrease	Decrease	Normal	Normal	Normal	Decrease	Decrease
7‖												
1	1, 2, 5	N	N	Decrease	No change	No change	Absent	No change	Decrease	2 mm	Decrease	Decrease
3	N	NE	NE	Normal	Normal	Normal	Absent	Normal	No change	2 mm	Normal	No change
8												
4	2, 5	NE	NE	Decrease, atrophy	No change	Decrease	Normal	Normal	Normal	1 mm	Decrease	Decrease
8	N	NE	NE	Atrophy	Normal	Normal	Normal	Normal	Normal	Normal	Normal	Normal
9												
5	N	N	N	Normal	Normal	Normal	Absent	Normal	Normal	Absent	Normal	Normal
10												
3	N	N	N	Normal	No change	No change	Absent	Normal	Normal	Absent	Normal	Stent
6	N	NE	NE	Atrophy	Normal	Normal	Absent	Normal	Normal	Absent	Normal	Normal
11												
4††	1, 6	N	+ (Decrease)	Increase	Normal	Increase	No change	No change	Normal	Normal	Stricture	Normal
12												
6	1 (Decrease)	NE	NE	Decrease	No change	Decrease	No change	No change	Decrease	Absent	No change	Absent
5††	1 (Decrease)	N	NE	Atrophy	Normal	Normal	No change	No change	Normal	Absent	No change	Absent
12	1 (Decrease)	NE	NE	Atrophy	Normal	Normal	Absent	No change	Normal	Absent	No change	Absent

(Table 2 continues)

Table 2 (continued)

Findings at Post-CST CT Examinations for Response Assessment in 13 Patients

Patient No. and Time Frame (mo)*	Clinical Features†	Serologic Results		Parenchymal Changes					Peripancreatic Changes		Ductal Changes	
		IgG Level‡	IgG4 Level§	Swelling	Enhancement	Lobularity	Focal Masslike Swelling	Tail Cutoff	Stranding	Halo	PD	CBD
13#												
5‡‡	1 (Decrease)	N	N	Decrease	Decrease	Normal	No change	No change	Absent	Absent	Stricture	Decrease, stent
8	1 (Decrease)	NE	NE	Atrophy	Normal	Normal	Absent	No change	Absent	Absent	Stricture	Absent

Note.—"Increase," "Decrease," and "No change" are relative to the findings at the previous examination. NE = not examined.

* Time interval in months for each follow-up CT examination is in relation to the preceding examination and not the baseline examination.

† 1 = Abdominal pain, 2 = jaundice, 3 = fatigue, 4 = weight loss, 5 = nausea and/or vomiting, 6 = pruritus, 7 = back pain, 8 = AIP incidentally detected at CT.

‡ N = normal (600–1500 mg/dL [6.0–15.0 g/L]), + = 1501–2000 mg/dL (15.1–20.0 g/L), ++ = >2000 mg/dL (>20.0 g/L).

§ N = normal (8–140 mg/dL [0.08–1.40 g/L]), + = 141–340 mg/dL (1.41–3.40 g/L), ++ = 341–540 mg/dL (3.41–5.40 g/L), +++ = 541–740 mg/dL (5.41–7.40 g/L), ++++ = 741–940 mg/dL (7.41–9.40 g/L), +++++ = >940 mg/dL (>9.40 g/L).

‖ History of known non–insulin-dependent diabetes mellitus.

New onset of non–insulin-dependent diabetes mellitus.

** CST was reinstituted in this patient at this point.

†† This patient underwent surgery with the Whipple procedure at this point.

‡‡ This patient underwent choledochojejunostomy at this point.

noted before and after treatment. Changes in the values at follow-up were assessed and correlated with imaging features (N.I.S.).

Multidetector CT and Image Analysis

All examinations were performed with a multidetector CT scanner with 4–16 detectors (LightSpeed; GE Medical Systems, Milwaukee, Wis) before and after intravenous administration of nonionic iodinated contrast material (iopamidol, Isovue 300; Bracco Diagnostics, Princeton, NJ). The scanning was performed with a maximum section thickness of 5 mm and a minimum section thickness of 1.25 mm. Postprocessed reconstructed images of the pancreas in coronal and oblique planes were available for 10 of 15 patients.

A single reader (D.V.S., with 13 years of radiology experience with special expertise in pancreatic imaging) who was aware of the purpose of this study performed the image analysis. The CT studies of each patient were interpreted sequentially, and the reader was kept blinded to the radiology reports, clinical and laboratory details, and treatment information. Image analysis was performed on a picture archiving and communication system (Agfa, Richmond, Va), and the readings were recorded in a systematic fashion on a predesigned template, which was later transferred to a standardized database (N.I.S.).

Pre-CST CT studies.—Pretreatment sequential multidetector CT studies were evaluated for all 15 patients to study the disease evolution (D.V.S.). Presence of pancreatic swelling, whether focal (when only a part of the pancreas was involved) or diffuse (involving the entire pancreas), was recorded. The pancreas was considered to be swollen subjectively when there was partial or complete obscuration of lobular architecture. A complete loss of pancreatic lobulation and absence of normal pancreatic clefts was described as a "featureless" pancreas. Focal masslike swelling was defined as focal enlargement without pancreatic or peripancreatic inflammatory changes. Enhancement of the pancreatic parenchyma was

subjectively described as homogeneous or heterogeneous. Involution of the pancreatic tail, or "tail cutoff," was considered to be present when there was abrupt blunting and retraction of the tail. The presence of parenchymal atrophy, calcification, or stones was assessed. The peripancreatic region was evaluated for stranding (represented by streaking of peripancreatic fat), a hypoattenuating rim surrounding the pancreas (referred to as a "halo"), and any fluid collections. The PD and CBD were assessed for focal or diffuse changes such as narrowing or stricture, dilatation, and abnormal enhancement. In addition, the presence of peripancreatic lymphadenopathy and the status of peripancreatic vessels were evaluated.

Quantitative analysis was performed on the CT study available just before the start of CST and included anteroposterior measurements of the pancreatic head (at the level of the portosplenic confluence), neck (anterior to the portal vein), body (to the left of the superior mesenteric artery at the level of the left renal vein), and tail (about 1 cm proximal to the distal end of the pancreas). The widest thickness of any halo and the short-axis diameter of the lymph nodes were also recorded. The widest diameter of the dilated CBD was measured on the baseline study (N.I.S.). These measurements were then transferred to the standardized database (N.I.S.).

Post-CST CT studies.—To evaluate the response to CST, CT findings before and after CST were compared in 13 patients (D.V.S.). Favorable response was suggested by partial or complete resolution of focal or diffuse swelling, recovery of pancreatic lobularity and tail, resolution of peripancreatic stranding and halo, and recovery of PD and CBD morphology. Partial response also included resolution of some features, with persistence or stability of others. Increase in the above-mentioned findings, development of focal PD or CBD stricture with upstream dilatation, or evidence of any new findings that were not present on previous studies indicated disease progression. Quantitative analysis of pancreatic size was performed on the last available CT study for each patient after they had received CST (N.I.S.). The change in pancreatic size before and after CST was analyzed for a significant difference by using the Wilcoxon signed rank test. $P < .05$ was considered to indicate a statistically significant difference.

Figure 1

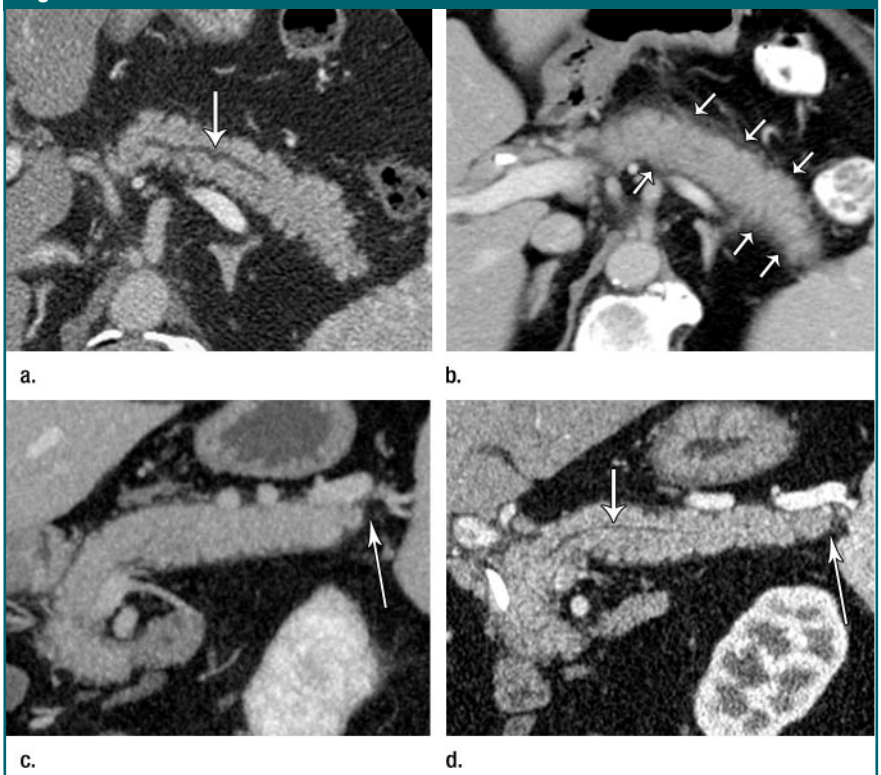

Figure 1: Serial contrast-enhanced CT images through pancreas in 84-year-old woman who presented with abdominal pain, jaundice, and weight loss and had complete response to CST. **(a)** Axial image from initial examination reveals normal pancreas morphology but minimal prominence of PD (arrow). **(b)** Axial and **(c)** coronal images from subsequent study performed at 2-month interval shows diffuse pancreatic swelling with loss of lobularity and a subtle halo around the pancreas (arrows in **b**). The PD is now attenuated (not visualized), and the pancreatic tail appears foreshortened (arrow in **c**). **(d)** Coronal image obtained after 2 months of CST reveals near complete resolution of pancreatic swelling and peripancreatic changes. Note that parenchymal lobularity has reappeared and that the PD (short arrow) is now visualized. Additionally, recovery of the pancreatic tail (long arrow) is observed.

Results

Clinical Features and Laboratory Data

Pretreatment.—The symptoms at presentation included abdominal pain ($n = 11$), jaundice ($n = 9$), fatigability ($n = 9$), weight loss ($n = 6$), nausea and/or vomiting ($n = 5$), pruritus ($n = 2$), back pain ($n = 1$), new onset of diabetes ($n = 2$), and no symptoms ($n = 1$). None of the patients had a history of pancreatitis or alcohol abuse. Four patients had associated autoimmune diseases (primary sclerosing cholangitis, Sjögren syndrome, Behçet disease, and rheumatoid arthritis). Serum IgG levels (available in 13 patients) were elevated in three patients, while IgG4 levels (available in nine patients) were elevated in eight patients (Tables 1, E1 [*http://radiology.rsnajnls.org/cgi/content/full/2493080279/DC1*].

Posttreatment.—After CST, complete resolution of symptoms was seen in eight of 13 patients, while in five patients, partial resolution was recorded

(Tables 2, E1 [*http://radiology.rsnajnls.org/cgi/content/full/2493080279/DC1*]. The latter five patients had persistent abdominal pain or varying degrees of other reported symptoms (jaundice in one patient, pruritus in another). Complete resolution of symptoms occurred within a 2–12-month interval (mean, 5.4 months). Gradual improvement in IgG and IgG4 levels was also noted in the patients when these levels were available, except in two patients with partial resolution of clinical symptoms in whom mild IgG4 elevation persisted (Tables 2, E1 [*http://radiology.rsnajnls.org/cgi/content/full/2493080279/DC1*].

Multidetector CT Image Analysis

Pre-CST CT studies.—The changes observed before institution of CST on CT studies in 15 patients in the evaluation of disease evolution are listed in Tables 1 and E1 (*http://radiology.rsnajnls.org/cgi/content/full/2493080279/DC1*). The number of pretreatment CT examinations and the time interval at which these examinations were performed varied for each patient. All patients underwent a minimum of two CT examinations, whereas four patients underwent four pretreatment examinations; the imaging interval ranged from 1 to 31 months (mean, 6.5 months).

At the initial CT examination, varying degrees of diffuse pancreatic swelling with loss of lobularity were observed in 14 of 15 patients (Figs 1, 2); one patient had fullness of the pancreatic parenchyma with preserved lobularity. These features gradually evolved to increase in swelling and/or loss of lobularity manifesting as a featureless "sausage-shaped" pancreas. In one patient (patient 8), the disease started as a focal swelling in the tail and then, over a period of 19 months, progressed to diffuse swelling and a featureless pancreas (Fig 3). In 10 patients, gradually increasing heterogeneous enhancement was observed in the swollen pancreas. Pancreatic tail retraction was noted at the baseline CT examination in eight patients and was observed to develop on follow-up CT studies in the other six patients. Retraction increased at follow-up examinations in five patients and remained stable in seven. Progression of disease thus incited or augmented retraction of the tail (Figs 1, 2). Peripancreatic findings (stranding [$n = 10$] and/or halo [$n = 9$]) noted at the baseline examination in nine patients paralleled the pancreatic changes, evolving and/or becoming more severe at follow-up examinations (Figs 1, 2).

Quantitatively, the maximum anteroposterior dimensions of the pancreas at the CT examination performed prior to the initiation of CST were as follows: head, 24–48 mm (mean, 32.2 mm ± 6.9 [standard deviation]); neck, 6–24 mm (14.3 mm ± 5.6); body, 15–31 mm (22 mm ± 5.1); and tail, 11–30 mm (21.8 mm ± 6.8). The thickness of the halo ranged from 1 to 10 mm (4.7 mm ± 2.8). The dimensions of the pancreas and the thickness of halo increased as the disease evolved. There was associated attenuation of the PD in 10 of 15 patients that was caused by swelling in the surrounding pancreatic parenchyma (Figs 1, 2). In four patients, the PD initially appeared prominent, and, as the parenchymal swelling progressed, diffuse attenuation of the PD occurred (Fig 1), whereas in one

Figure 2

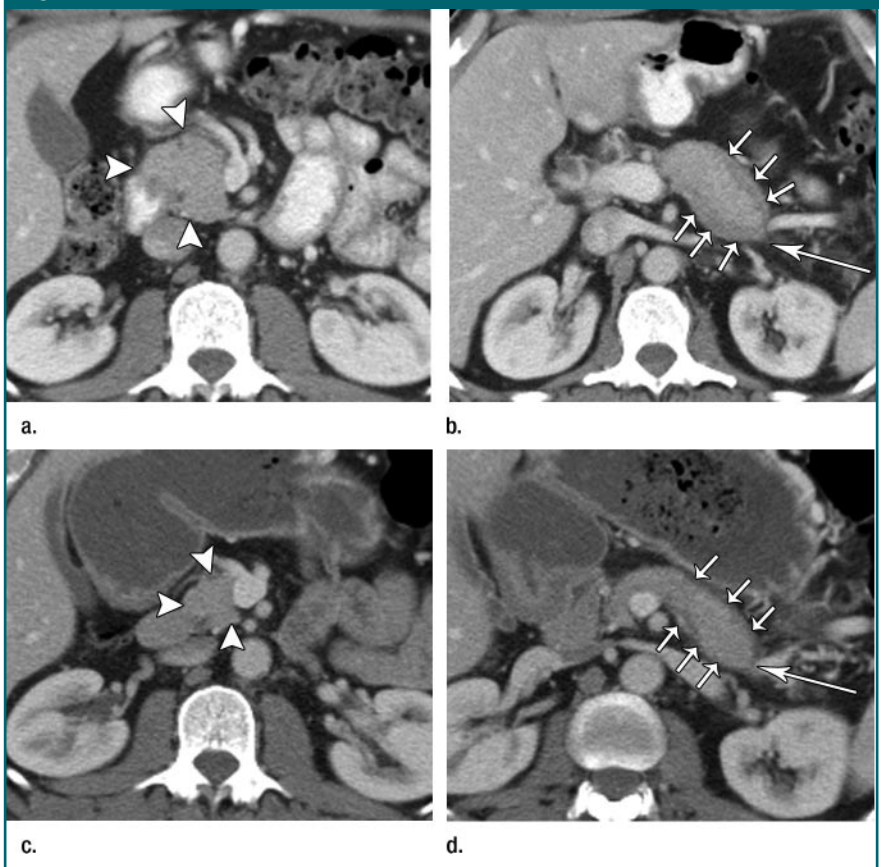

Figure 2: **(a–d)** Axial contrast-enhanced CT images through pancreas in 58-year-old man who presented with jaundice and abdominal pain and who demonstrated a partial response to CST. Images **a** and **b** were obtained before and **c** and **d** were obtained 8 months after CST. **(a, b)** Focal prominent swelling is seen in the head of the pancreas (arrowheads); diffuse swelling, loss of lobularity, halo (short arrows) in the region of the body and tail, and retraction of the tail (long arrow) are also seen. **(c, d)** Follow-up images reveal reduction in the size of the focal swelling in the pancreatic head (arrowheads) and partial resolution of pancreatic swelling and halo (short arrows). The retraction of the tail (long arrow) persisted.

patient, the PD was irregularly narrowed. The distal CBD showed smooth tapering due to swelling in the pancreatic head in 12 patients, in three of whom thick enhancing walls of the CBD were noted. There were associated variable degrees of dilatation of the proximal CBD (widest diameter range, 8–16 mm; mean, 11.4 mm [as measured at the baseline examination]) and intrahepatic biliary radicals in all of these patients. Eight of the 12 patients underwent stent placement for palliative relief of biliary obstruction, and in the remaining patients, these obstructive changes either gradually progressed ($n = 2$) or remained stable ($n = 2$). None of the patients had parenchymal calcification, stones, or atrophy at the pretreatment CT examinations. One patient (patient 8) developed a cystic lesion with attenuation of fluid in the body of the pancreas during the course of the disease; this lesion, however, resolved at follow-up imaging before CST. Peripancreatic lymph node enlargement was observed in eight of 15 patients during the course of the disease. The size of the lymph nodes ranged from 6 to 13 mm.

Post-CST CT studies.—The changes observed at CT examinations in 13 patients treated with CST are listed in Tables 2 and E1 (*http://radiology.rsnajnls.org/cgi/content/full/2493080279/DC1*). The number of posttreatment CT examinations and the time interval at which these examinations were performed for each patient varied. All patients underwent a minimum of one posttreatment CT examination, while in two patients, three posttreatment CT studies were

Figure 3

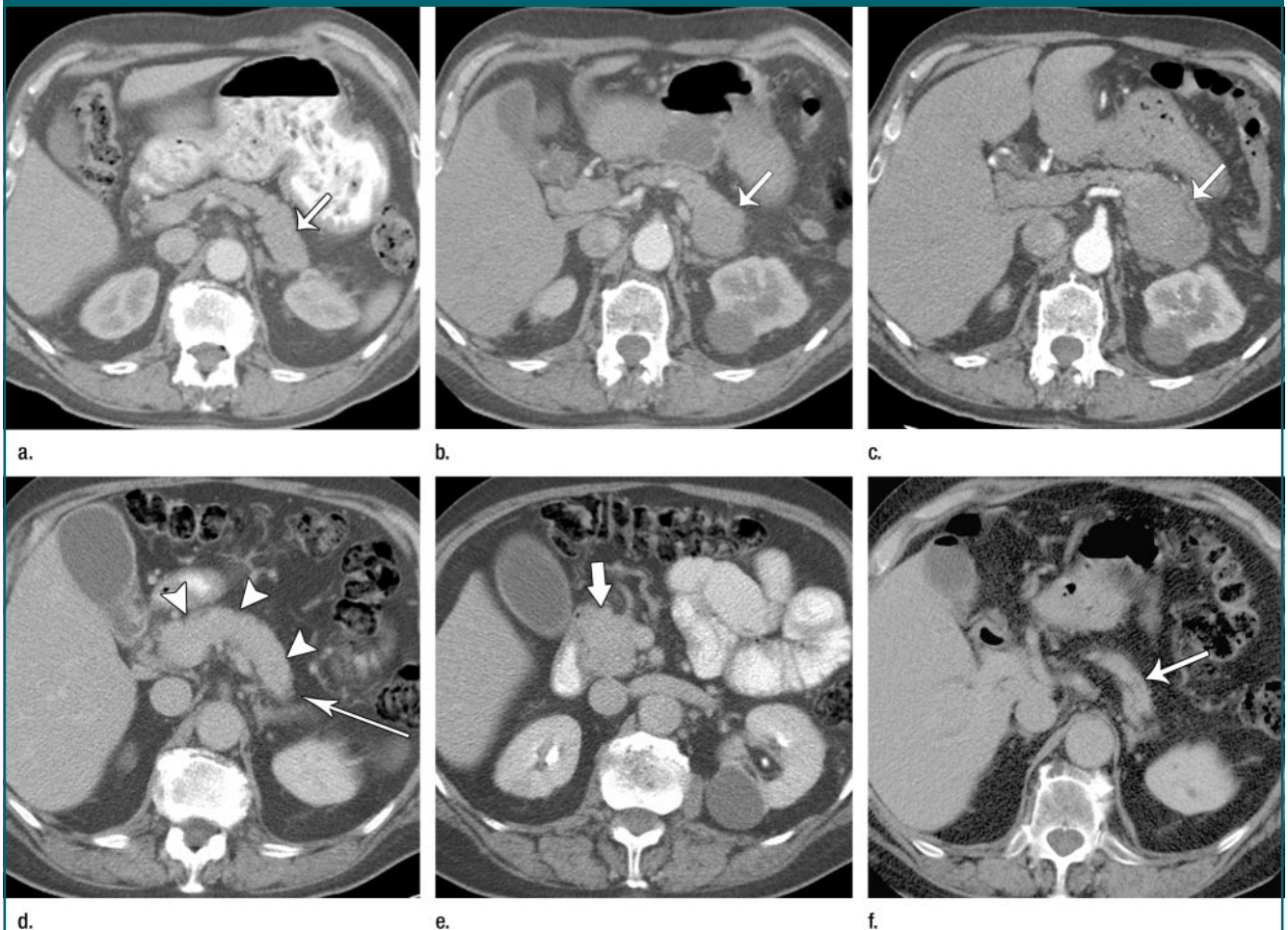

Figure 3: Sequential axial CT images through pancreas in 72-year-old man who eventually developed atrophy of the pancreas after CST. **(a)** Image from initial examination shows mild fullness in the tail (arrow) that worsened gradually over an 8-month period on **(b, c)** images from two subsequent CT examinations (6 and 8 months from baseline, respectively). **(d)** The disease in the pancreas then evolved over next 18 months into diffuse swelling (arrowheads), retraction of the tail (arrow), and loss of lobularity. The patient had jaundice and pruritus at this stage. **(e)** The head of the pancreas at this stage (18 months from baseline) also appears enlarged (arrow), but the focal changes in the tail noted on prior studies have resolved. **(f)** A course of CST was given that resulted in complete resolution of pancreatic swelling 22 months from baseline, with changes of atrophy in the pancreas (arrow).

available. The total posttreatment time duration ranged from 2 to 23 months (mean, 8.3 months).

Complete resolution of pancreatic changes (swelling, loss of lobularity, heterogeneity, tail retraction), peripancreatic changes, and ductal changes was recorded in four (patients 1, 8, 9, and 10) of 13 patients (Fig 1) over a period of 5 months to 1 year (mean, 7.7 months). Among these patients, improvement in parenchymal swelling and peripancreatic stranding and/or halo was the most notable finding. These changes gradually improved to complete resolution, and, finally, the pancreas appeared slightly atrophied than usually expected in two patients (patients 8 and 10) (Fig 3). The atrophy developing in some patients at follow-up evaluation has been attributed to acinar loss (20,21). The ductal changes also gradually resolved in parallel with the parenchymal swelling (Fig 1). Regional lymphadenopathy resolved in six of eight patients (3,22,23). These described imaging manifestations that favorably responded to CST have been attributed to inflammatory changes (13,20,24). Complete resolution of clinical and serologic parameters (when available) was noted in these patients (Tables 2, E1 [*http://radiology.rsnajnls.org/cgi/content/full/2493080279/DC1*].

Partial response was observed in nine of 13 patients at follow-up examinations over a period of 2–23 months (mean, 8.2 months). The parenchymal changes (swelling, loss of lobularity, heterogeneity) partially improved in five of the nine patients, and peripancreatic changes improved in two of the nine patients (Fig 2). In seven of nine patients with partial response, retraction of the tail persisted (Fig 2). In another seven of nine patients, diffuse swelling in the body and tail showed obvious improvements; however, a more focal masslike swelling in the head either remained unchanged (in five of seven patients) or showed minimal improvement (in two of seven patients) (Figs 2, 3). Three of these patients with persistent swelling in the head (patients 11, 12, and 13) later underwent surgery to relieve symptoms. The manifestations in the CBD and PD also resolved partially in five of nine and seven of nine patients, respectively (Fig 3). In two of the patients with PD changes (patients 11 and 13), the PD changes progressed to a focal stricture in the head (disease progression); in these patients, there were other associated chronic changes (persisting focal masslike swelling and/or atrophy) (3,13). Focal masslike swelling, persistent retraction of the tail, and ductal strictures were the features observed late in the evolution of disease and have been attributed to chronicity and the inception of fibrotic changes (3,13). In one patient with diffuse changes in the pancreas and an associated focal masslike lesion in the head (patient 6), substantial improvement was noted at imaging and in clinical symptoms 6 weeks after CST; therefore, CST was terminated. However, at follow-up CT examination 6 months after the institution of CST, focal swelling developed in the tail, and CST was therefore reinstituted. Subsequent CT studies over 2 months demonstrated marked improvement in the pancreatic changes. Eight patients with CBD dilatation underwent stent placement; the stents were removed later as the patients' conditions improved clinically and radiologically (at multidetector CT and/or ERCP). Among nine patients with partial response at imaging, partial resolution of symptoms was noted in five patients (two patients with focal swelling in the pancreatic head and PD stricture, one patient with focal swelling in the pancreatic head, and two other patients), while in the remaining patients, the symptoms resolved completely. Results of serologic examination, when available, revealed improvement after CST (Tables 2, E1 [*http://radiology.rsnajnls.org/cgi/content/full/2493080279/DC1*].

The quantitative dimensions of the pancreas on the last available CT studies for the patients treated with CST were as follows: pancreatic head, 17–28 mm (23.1 mm ± 4.2); neck, 6–16 mm (10.4 mm ± 3.2); body, 10–28 mm (17.5 mm ± 4.6); and tail, 11–25 mm (17.1 mm ± 3.9). Differences in the size of the pancreas measured before and after CST were statistically significant (head, $P = .003$; neck, $P = .007$; body, $P = .0002$; and tail, $P = .003$).

Discussion

AIP, first described by Sarles et al in 1961 (25) as "primary inflammatory sclerosis" of the pancreas, was later understood to be a variant of chronic pancreatitis invoked by an autoimmune process; hence, the currently accepted terminology of "autoimmune pancreatitis" has come into existence (16,22). The widely accepted criteria for the diagnosis of AIP are those developed by the Japan Pancreas Society in 2002 and revised in 2006, which include imaging, serologic, and pathologic criteria (25, 26). The HISORt criteria of the Mayo Clinic in 2006 (13) include extrapancreatic manifestations and response to CST in addition to the above-mentioned criteria. Although the diagnostic criteria for AIP have been well described, the evolution of the disease is not well understood, despite its potential relevance in selecting an appropriate treatment strategy. In our study, CST was initiated in the patients because of strong suspicion of AIP on the basis of clinical manifestation, laboratory values (elevated IgG4 levels), and imaging features such as sequential multidetector CT findings over time and findings at ERCP and/or endoscopic US with fine-needle aspiration cytology that were consistent with diagnosis. Invasive procedures such as biopsy were not routinely performed for histopathologic confirmation in the cohort of patients included in our study (5,13,27–29). The favorable response to CST objectively monitored with multidetector CT further supports the fact that imaging features, in conjunction with clinical and relevant laboratory data (when reviewed cautiously), in patients with a high level of suspicion of AIP can be regarded as sufficient evidence to initiate a trial of CST.

The spectrum of imaging features of AIP at CT have been described in the literature (8–10). Moreover, differences in the degree of response to CST, from normalization of pancreatic and ductal changes (7,9,13,16,30) to persistence of focal masslike lesions and develop-

ment of ductal strictures, have been reported (8,30). These observations suggest that this array of imaging manifestations could represent different phases in disease evolution, analogous to the staging proposed by Zamboni et al (3) on the basis of differences in the extent of inflammatory changes and fibrosis. Analyzing the course of events at CT, we observed that the disease manifested as diffuse or focal swelling in the pancreas. Without treatment, these changes progressed into worsening pancreatic swelling and complete loss of parenchymal lobularity, resulting in a featureless, sausage-shaped pancreas (8,11). Concurrent progression of pancreatic tail retraction, peripancreatic findings, and ductal changes was also noted. The disease started as a focal swelling in one patient that later advanced to involve the gland diffusely. This finding suggests that AIP can start as a focal, low-grade inflammation in the pancreas that results in nonspecific symptoms for which the patients may not seek medical attention. As the disease progresses into a diffuse form, patients can experience symptoms that require a visit to the physician. Similar observation of the evolution of focal into diffuse changes was also made by Hirano et al (30). As in earlier studies, in our study, diffuse pancreatic swelling, peripancreatic changes, and ductal obliteration or thickening (when present) responded favorably to CST treatment in all patients in a short time frame (1 month in a few patients), thus supporting the hypothesis of the presence of an underlying active inflammatory process (13,16,20,24). Incomplete resolution of the discussed changes observed in a few patients in our study could be attributed to a slower responder or to inadequate availability of follow-up studies. Focal masslike swelling (when present without peripancreatic inflammatory changes), persistent focal ductal narrowing (stricture formation), and tail retraction in some cases reflected suboptimal response to CST and exemplify late events in the disease evolution that are likely due to the inception of fibrosis or diffuse sclerosis (3,13,31). Persisting focal masslike swelling can masquerade as pancreatic carcinoma clinically and at imaging and often requires surgical resection to alleviate the patient's symptoms (3,17,20,24,29). Also, to avoid any delay in treating a veiled malignancy, surgical exploration has been recommended for corticosteroid-treated focal masslike swellings that do not show improvement at follow-up imaging, as observed in some of our patients (13). The observation of resolution of ductal narrowing in some cases and lack of resolution with development of stricture despite treatment in others suggests that CST, if instituted early in the course of the disease, results in a favorable outcome as it can avert evolution to the late stage of disease. Hirano et al (30) also found CST to be particularly useful in preventing ductal lesions. Development of a pancreatic cyst in association with AIP that disappears naturally, which was seen in one of our patients, has also been reported by Nakazawa et al (10) and has been attributed to ductal stenosis with formation of a retention cyst.

The duration of CST is currently not standardized, as in a few patients, more than one course of therapy may be required because of recurrence or development of new imaging findings at cessation of CST, as observed in our study. Other investigators (7,14,30) have also made similar observations, and patients with partial response to CST may require reinstitution of CST at similar or higher doses. CST may also be needed after surgery because of the development of new imaging findings; a new intrahepatic biliary stenosis was reported in one patient after pancreatoduodenectomy by Nakazawa et al (10). For the same reason, regular clinical, serologic, and imaging follow-up is prudent for monitoring disease status in patients with incomplete response (14,30).

Our study had a number of limitations. We did not have control over several factors in this retrospective study. The scanning parameters and protocol, the type and volume of intravenous contrast material, and the types of prednisone used were not consistent. Laboratory parameters, especially serologic results, were not available in all patients or at all time points when CT examinations were performed to establish correlation. Specific inclusion criteria resulted in a small cohort, which limited correlating demographic (age and sex) patterns with CT results. Detection of disease at a different phase at the baseline CT examination in each patient and the variable time interval at which the pretreatment and posttreatment examinations were performed for all patients prevented relating development or evolution of a specific imaging feature with a specific time interval. However, this situation is very commonly encountered in this disease, as many patients present in the postacute phase, a variable time interval after the onset of the disease process (10,13). Moreover, the partial response observed in some patients could be partly related to the unavailability of follow-up studies until complete resolution. Finally, a single reader aware of the purpose of the study evaluated all the imaging studies.

In conclusion, knowledge of the morphologic evolution of AIP and recognition of the imaging features at CT that can predict outcome after CST might be crucial in deciding the appropriate treatment for patients with this disease. In our experience, pancreatic swelling with peripancreatic changes, including the halo, represents an early or inflammatory dominant phase of the disease that responds favorably to CST. Focal masslike swelling, especially in the pancreatic head, without diffuse pancreatic and peripancreatic changes and development of focal PD strictures are manifestations of a late disease stage with predominance of fibrosis and less degree of inflammation and are predictive of a suboptimal outcome after CST, frequently requiring surgical intervention.

References

1. Notohara K, Burgart LJ, Yadav D, Chari S, Smyrk TC. Idiopathic chronic pancreatitis with periductal lymphoplasmacytic infiltration: clinicopathologic features of 35 cases. Am J Surg Pathol 2003;27:1119–1127.

2. Weber SM, Cubukcu-Dimopulo O, Palesty JA, et al. Lymphoplasmacytic sclerosing pancreatitis: inflammatory mimic of pancre-

atic carcinoma. J Gastrointest Surg 2003;7:129–137.

3. Zamboni G, Luttges J, Capelli P, et al. Histopathological features of diagnostic and clinical relevance in autoimmune pancreatitis: a study on 53 resection specimens and 9 biopsy specimens. Virchows Arch 2004;445:552–563.
4. Yadav D, Notahara K, Smyrk TC, et al. Idiopathic tumefactive chronic pancreatitis: clinical profile, histology, and natural history after resection. Clin Gastroenterol Hepatol 2003;1:129–135.
5. Kim KP, Kim MH, Kim JC, Lee SS, Seo DW, Lee SK. Diagnostic criteria for autoimmune chronic pancreatitis revisited. World J Gastroenterol 2006;12:2487–2496.
6. Wakabayashi T, Kawaura Y, Satomura Y, et al. Duct-narrowing chronic pancreatitis without immunoserologic abnormality: comparison with duct-narrowing chronic pancreatitis with positive serological evidence and its clinical management. Dig Dis Sci 2005;50:1414–1421.
7. Ito T, Nishimori I, Inoue N, et al. Treatment for autoimmune pancreatitis: consensus on the treatment for patients with autoimmune pancreatitis in Japan. J Gastroenterol 2007;42(suppl 18):50–58.
8. Sahani DV, Kalva SP, Farrell J, et al. Autoimmune pancreatitis: imaging features. Radiology 2004;233:345–352.
9. Yang DH, Kim KW, Kim TK, et al. Autoimmune pancreatitis: radiologic findings in 20 patients. Abdom Imaging 2006;31:94–102.
10. Nakazawa T, Ohara H, Sano H, et al. Difficulty in diagnosing autoimmune pancreatitis by imaging findings. Gastrointest Endosc 2007;65:99–108.
11. Finkelberg DL, Sahani D, Deshpande V, Brugge WR. Autoimmune pancreatitis. N Engl J Med 2006;355:2670–2676.
12. Koga Y, Yamaguchi K, Sugitani A, Chijiiwa K, Tanaka M. Autoimmune pancreatitis starting as a localized form. J Gastroenterol 2002;37:133–137.
13. Chari ST, Smyrk TC, Levy MJ, et al. Diagnosis of autoimmune pancreatitis: the Mayo Clinic experience. Clin Gastroenterol Hepatol 2006;4:1010–1016.
14. Kamisawa T, Yoshiike M, Egawa N, Nakajima H, Tsuruta K, Okamoto A. Treating patients with autoimmune pancreatitis: results from a long-term follow-up study. Pancreatology 2005;5:234–238.
15. Hamano H, Kawa S, Horiuchi A, et al. High serum IgG4 concentrations in patients with sclerosing pancreatitis. N Engl J Med 2001;344:732–738.
16. Yoshida K, Toki F, Takeuchi T, Watanabe S, Shiratori K, Hayashi N. Chronic pancreatitis caused by an autoimmune abnormality: proposal of the concept of autoimmune pancreatitis. Dig Dis Sci 1995;40:1561–1568.
17. Kamisawa T, Egawa N, Nakajima H, Tsuruta K, Okamoto A, Kamata N. Clinical difficulties in the differentiation of autoimmune pancreatitis and pancreatic carcinoma. Am J Gastroenterol 2003;98:2694–2699.
18. Nishino T, Toki F, Oyama H, Shimizu K, Shiratori K. Long-term outcome of autoimmune pancreatitis after oral prednisolone therapy. Intern Med 2006;45:497–501.
19. Czako L, Hegykozi E, Palinkas A, Lonovics J. Autoimmune pancreatitis: functional and morphological recovery after steroid therapy. World J Gastroenterol 2006;12:1810–1812.
20. Deshpande V, Mino-Kenudson M, Brugge W, Lauwers GY. Autoimmune pancreatitis: more than just a pancreatic disease? a contemporary review of its pathology. Arch Pathol Lab Med 2005;129:1148–1154.
21. Kamisawa T, Okamoto A. Prognosis of autoimmune pancreatitis. J Gastroenterol 2007;42(suppl 18):59–62.
22. Okazaki K, Chiba T. Autoimmune related pancreatitis. Gut 2002;51:1–4.
23. Kloppel G, Luttges J, Sipos B, Capelli P, Zamboni G. Autoimmune pancreatitis: pathological findings. JOP 2005;6:97–101.
24. Irie H, Honda H, Baba S, et al. Autoimmune pancreatitis: CT and MR characteristics. AJR Am J Roentgenol 1998;170:1323–1327.
25. Sarles H, Sarles JC, Muratore R, Guien C. Chronic inflammatory sclerosis of the pancreas: an autonomous pancreatic disease? Am J Dig Dis 1961;6:688–698.
26. Pearson RK, Longnecker DS, Chari ST, et al. Controversies in clinical pancreatology: autoimmune pancreatitis—does it exist? Pancreas 2003;27:1–13.
27. Okazaki K, Kawa S, Kamisawa T, et al. Clinical diagnostic criteria of autoimmune pancreatitis: revised proposal. J Gastroenterol 2006;41:626–631.
28. Ghazale A, Chari ST, Smyrk TC, et al. Value of serum IgG4 in the diagnosis of autoimmune pancreatitis and in distinguishing it from pancreatic cancer. Am J Gastroenterol 2007;102:1646–1653.
29. Kwon S, Kim MH, Choi EK. The diagnostic criteria for autoimmune chronic pancreatitis: it is time to make a consensus. Pancreas 2007;34:279–286.
30. Hirano K, Tada M, Isayama H, et al. Long-term prognosis of autoimmune pancreatitis with and without corticosteroid treatment. Gut 2007;56:1719–1724.
31. Suda K, Takase M, Fukumura Y, Kashiwagi S. Pathology of autoimmune pancreatitis and tumor-forming pancreatitis. J Gastroenterol 2007;42(suppl 18):22–27.