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Autoimmune Pancreatitis Novelty as Triggered by Sunlight Exposure in a Patient with Verified Causality by Unintentional Re-exposure as Diagnostic Gold Standard: A Case Report

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Abstract

This report describes as novelty the unique case of a patient with painless jaundice due to Autoimmune Pancreatitis (AIP) related to significant sunshine exposure as triggering factor never described before in the literature. The causal association was confirmed by positive unintentional reexpose results viewed as gold standard in clinical diagnostics: (1) male gender aged 76 years and of European ethnicity (Germany); (2) normal serum IgG4 levels excludes attribution of the case to the large group of IgG4 disorders; (3) association with secondary sclerosing cholangitis characterized by prestenotic dilatations above stenoses of the hepatic fork and within intrahepatic bile ducts; (4) treatment with prednisolone and azathioprine was unsuccessful in preventing cellular pancreatic disruption with exocrine pancreatic insufficiency due to organ burnt out and organ atrophy, associated with persisting xerostomia likely due to hyposalivation of exocrine salivary gland insufficiency; and (5) avoiding direct sunlight exposure prevents disease recurrence. In essence, this AIP case with novelty features in form of sunshine exposure as culprit perfectly established regarding causality through a positive unintentional re exposure result differs from current AIP-1 and AIP-2 both lacking an established culprit, and from AIP-3 caused by Immune Checkpoint Inhibitors (ICIs), representing a new AIP type with the option being termed as fourth AIP type observed first in Europe specifically Germany and may call for refining existing AIP guideline recommendations in Japan and Europe based on a mandatory reevaluation of the current AIP classifications but now under inclusion of details provided by the novel AIP case.

Keywords: Sunlight exposure; Autoimmunity; Pancreatic disruption; Autoimmune Pancreatitis (AIP); Secondary Sclerosing Cholangitis (SSC); AIP guidelines

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Introduction

Autoimmune Pancreatitis (AIP) is the result of organ disruption with pancreatic manifestation due to the aberrant autoimmune response against the pancreas, leading to inflammation and fibrosis [1]. Currently, AIP can be classified as three types [1-5]. Accordingly, type 1 AIP (AIP-1) is commonly associated with elevated serum Immunoglobulin G4 (IgG4) levels, is characterized by systemic manifestations [1], and also known as lymphoplasmacytic sclerosing pancreatitis [2]. As opposed, type 2 AIP (AIP-2) typically lacks increased serum IgG4 levels, may coexist with other autoimmune disorders preferentially inflammatory bowel diseases [1,5] and is also termed idiopathic duct-centric pancreatitis [2]. AIP-1 and AIP-2 share the common features of idiopathy, meaning that triggering factors remain undisclosed.

We present as novelty the rare case of a patient diagnosed with AIP who experienced two flares in temporal association with significant sunshine exposure and confirmed causal association based on a positive unintentional preexposure test result commonly viewed as gold standard approach in clinical diagnostics exploring yet unknown causes in diseases. Sunlight exposure as triggering factor in AIP is a novelty.

Case Presentation

Report of a rare novel AIP case triggered by significant sunshine exposure

This report describes the case of a 76-year-old male patient with a high level of expertise in clinical gastroenterology, who was in fairly good health condition until he was diagnosed with AIP. In 2020, he noticed a newly appearing brownish urine, which he attributed to urinary concentration following extensive gardening work under sunshine conditions associated with substantial perspiration due to high temperature and low fluid intake. During the following days, the urine remained largely brownish despite increased fluid intake. Other new symptoms included pruritus, scleral jaundice, general painless jaundice, and voluminous stool in the sense of steatorrhea, which prompted him to seek advice by a gastroenterologist at the local hospital. There was no prior travelling to other countries, no contact with persons suffering from hepatitis, no systemic drug therapy despite of being at a higher age, no exposure to herbal medicines or dietary supplements, and no handling with recreational and occupational hazards including pesticides or herbicides such as glyphosate. There was concern of an obstructive cancer of the pancreas with liver metastases or biliary obstruction. At first presentation in the hospital, laboratory results showed abnormal Liver Test (LTs) of Alanine Aminotransferase (ALT), Aspartate Aminotransferase (AST), Alkaline Phosphatase (ALP), and Gamma-Glutamyl Transferase (GGT), as well as increased Total Bilirubin (TBILI), while serum lipase was normal now and during the following years table 1.

At hospital presentation on 13.02.2020, the abdominal Ultrasound (US) examination showed an enlarged head of the pancreas of unclear dignity associated with a widened common bile duct of 11 mm and signs of intrahepatic cholestasis in an otherwise unremarkable liver without signs of metastases. Spleen and kidneys showed no abnormalities. Small intestine and colon were without abnormalities and showed more specially no wall enlargement in the sense of pathological cockades. The US examination of 15.10.2020 showed a minimal intrahepatic cholestasis of the left liver with normal appearance of the remaining parts of the liver, the common bile duct was unremarkable without dilatation or stenosis, while the head of the pancreas was marginally enlarged with pancreatic atrophy of the body and tail.

Based on a possible cancer of the pancreas head as suspected by US, a contrast-enhanced Computed Tomography (CT) scan with contrast medium was indicated. CT findings included an enlargement of the common bile tract with 12 mm with stop above entrance to the pancreas head. The intrahepatic bile ducts appeared dilatated in an otherwise normal homogenous liver without focal lesions like metastases. Spleen, kidneys, and gastrointestinal tract were without pathological findings. The CT showed an enlarged head of the pancreas and a soft tissue tumescence around the body corpus and tail of the pancreas with a diameter of up to 8 mm in the sense of a sausage-like picture strongly suggestive of AIP (Figure 1).

The CT findings were corroborated by the subsequent Magnetic Resonance Cholangiopancreatography (MRCP), which again showed

Table 1: Case details of the patient with two AIP flares.

Dates	Case details and laboratory data of a new AIP type termed AIP-4
13.02.2020 Prior significant sun exposure	First flare in association with significant sunlight exposure ALT 185 U/L; AST 97 U/L; ALP 302 U/L; GGT 447 U/L; TBILI 12.4 mg/dL; LIP 41 U/L; CA 19-9 149 U/mL; IgG4 0.333 g/L; IgA 161 mg/dL; IgE 45.7 U/mL; IgG 924 mg/dL; IgM 38 mg/dL; IL-6 8.0 pg/mL. Negative titers of serum autoimmune and virus ab. R 0.22,
16.02.2020	Upon treatment with prednisolone (PRED) 40 mg/d, virtually all lab values improved ALT 14 U/L; AST 24 U/L; ALP 66 U/L; GGT 70 U/L; TBILI 0.9 mg/dL.
18.02.2020	ALT 95 U/L; AST 42 U/L; ALP 250 U/L; GGT 320 U/L; TBILI 10.4 mg/dL.
04.03.2020	Fluctuation of laboratory results necessitated switching from PRED to azathioprine (AZA), as shown by lab data ALT 246 U/L; AST 108 U/L; ALP 168 U/L; GGT 142 U/L; TBILI 4.5 mg/dL.
01.04.2020	ALT 39 U/L; AST 27 U/L; ALP 90 U/L; GGT 62 U/L; TBILI 0.9 mg/dL.
06.05.2020	ALT 135 U/L; AST 72 U/L; ALP 257 U/L; GGT 411 U/L; TBILI 9.6 mg/mL.
10.06.2020	ALT 73 U/L; AST 32 U/L; ALP 116 U/L; GGT 256 U/L; TBILI 1.0 mg/dL.
15.07.2020	ALT 19 U/L; AST 24 U/L; ALP 72 U/L; GGT 62 U/L; TBILI 0.7 mg/dL.
23.09.2020	ALT 126 U/L; AST 59 U/L; ALP 176 U/L; GGT 1197 U/L; TBILI 1.4 mg/dL.
30.12.2020	ALT 12 U/L; AST 26 U/L; ALP 72 U/L; GGT 12 U/L; TBILI 0.7 mg/dL.
27.01.2021	ALT 14 U/L; AST 30 U/L; ALP 82 U/L; GGT 121 U/L; TBILI 0.9 mg/dL.
03.03.2021	ALT 16 U/L; AST 26 U/L; ALP 94 U/L; GGT 134 U/L; TBILI 0,7 mg/dL.
28.04.2021	ALT 14 U/L; AST 24 U/L; ALP 93 U/L; GGT 80 U/L; TBILI 1.0 mg/dL.
26.05.2021	ALT 28 U/L; AST 33 U/L; ALP 143 U/L; GGT 298 U/L; TBILI 1.0 mg/dL.
04.08.2021	ALT 15 U/L; AST 22 U/L; ALP 77 U/L; GGT 78 U/L; TBILI 0.9 mg/dL.
08.12.2021	ALT 19 U/L; AST 26 U/L; ALP 56 U/L; GGT 34 U/L; TBILI 0.5 mg/dL.
12.01.2022	ALT 23 U/L; AST 11 U/L; ALP 71 U/L; GGT 22 U/L; TBILI 0.5 mg/dL.
09.02.2022	ALT 11 U/L; AST 24 U/L; ALP 75 U/L: GGT 20 U/L; TBILI 0.6 mg/dL.
O9.03.2022	Treatment stop ALT 12 U/L; AST 23 U/L; ALP 71 U/L; GGT 21 U/L; TBILI 0.5 mg/dL.
11.05.2022	ALT 15 U/L; AST 22 U/L; ALP 78 U/L; GGT 37 U/L; TBILI 0.5 mg/dL.
15.06.2022	ALT 13 U/L; AST 24 U/L; ALP 76 U/L; GGT 28 U/L; TBILI 0.7 mg/dL.
13.07.2022	ALT 8 U/L; AST 21 U/L; ALP 77 U/L; GGT 18 U/L; TBILI 0.7 mg/dL.
08.11.2022	ALT 15 U/L; AST 24 U/L; ALP 86 U/L; GGT 41 U/L; TBILI 0.5 mg/dL.

15.02.2023	ALT 17 U/L; AST 22 U/L; ALP 43 U/L; GGT 17 U/L; TBILI 0.6 mg/dL.
06.09.2023 Prior significant sun exposure +	Second flare due to significant unintentional sunshine re exposure ALT 440 U/L; AST 310 U/L; ALP 470; GGT 77 U/L; TBILI 1.9 mg/dL; R 3.0. Re-introduction of treatment with PRED combined with AZA
11.09.2023 Sun exposure -	ALT 253 U/L; AST 119 U/L; ALP 314 U/L; GGT 777 U/L: TBILI 1.2 mg/dL.
27.09.2023 Sun exposure -	ALT 98 U/L; AST 92 U/L; ALP 275 U/L; GGT 505 U/L; TBILI 0.9 mg/dL.
24.01.2024 Sun exposure -	ALT 354 U/L; AST 217 U/L; ALP 479 U/L; GGT 638 U/L; TBILI 6.8 mg/dL.
21.02.2024 Sun exposure -	ALT 111 U/L; AST 58 U/L; ALP 199 U/L; GGT 319 U/L; TBILI 2.7 mg/dL.
10.04.2024 Sun exposure -	ALT 48 U/L; AST 34 U/L; ALP 95 U/L; GGT 73 U/L; TBILI 0.9 mg/dL.
12.06.2024 Sun exposure -	ALT 16 U/L; AST 24 U/L; ALP 53 U/L; GGT 24 U/L; TBILI 0.7 mg/dL.
21.08.2024 Sun exposure -	Treatment stop ALT 11 U/L; AST 19 U/L; ALP 57 U/L; GGT 17 U/L; TBILI 0.6 mg/dL.
26.3.2025 Sun exposure -	ALT 8 U/L; AST 18 U/L; ALP 74 U/L; GGT 17 U/L; TBILI 0.9 mg/dL.
21.05.2025 Sun exposure -	ALT 11 U/L; AST 20 U/L; ALP 79 U/L; GGT 17 U/L; TBILI 0.6 mg/dL.
15.10.2025 Sun exposure -	ALT 11 U/L; AST 21 U/L; ALP 77 U/L; GGT 16 U/L; TBILI 0.7 mg/dL; HbA1c 5.9%; Total vitamin D 36.6 mg/ml; CA-19-9 8.1 U/L; CEA 2.7 U/L.

The RUCAM-based R (ratio) value mentioned above determines the liver injury pattern with recommendations published earlier in the updated RUCAM report [6]. For the liver injury pattern, the value of the R value was calculated using the multiples of the upper limit of normal (ULN) for ALT: ALP, whereby R ≥5 corresponds to the hepatocellular injury, R ≤2 to the cholestatic liver injury, and 2 < R < 5 to the mixed liver injury pattern. For the current patient, the R value was 0.22 at the first flare and 3.0 at the second flare. Normal ranges: ALT <41 U/L; AST <38 U/L; ALP <129 U/L; HbA1c 4.5-5.7%; TBILI <1.0 mg/dL; LIP <60 U/L; CA 19-9 <39.0, CEA <3.4; IgG4 0.052 - 1.250 g/L; IgA 70 - 400 mg/dL; IgE <140.0 U/mL, IgG 700 -1600 mg/dL; IgM 40 - 239 mg/dL; IL-6 <7.0 pg/mL; total vitamin D level >30.0 mg/ml. The sign "-" means that sunlight exposure was thoroughly avoided. Abbreviations: ALT, Alanine aminotransferase; ALP, Alkaline Phosphatase; ANCA, Antineutrophilic cytoplasmic antibody; AST, Aspartate aminotransferase; AZA, Azathioprine; CA 19-9, Carbohydrate Antigen; CEA, Carcinoembryonic Antigen; GGT, Gamma-Glutamyl Transferase; IgG, Immune Globulin; IL, Interleukin; LIP, Lipase; PRED, Prednisolone; Roussel Uclaf Causality Assessment Method; TBILI, total bilirubin.

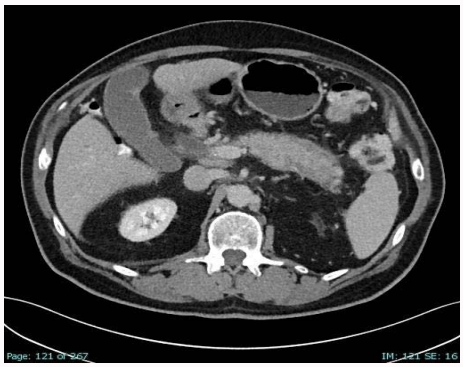

Figure 1: CT portal venous phase of the pancreatic tail with sausage-like appearance and halo sign.

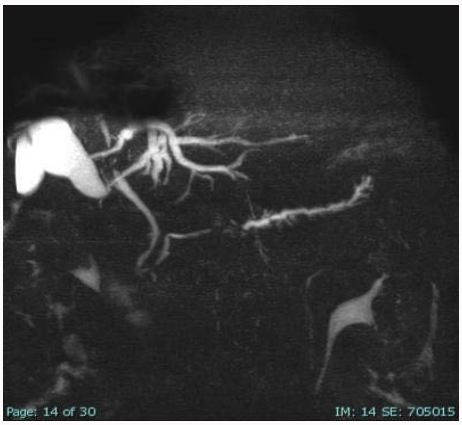

Figure 2: MRCP with focus on intrahepatic secondary sclerosing cholangitis.

an edematous pancreas compatible with AIP [6]. In the further course 15 months after first presentation and still under maintaining therapy with AZA, the MRCP native and with contrast medium revealed apart from a normally appearing large bile duct and lower common bile duct an enlargement of the intrahepatic bile ducts with substantial stenosis of the diameter above the hepatic fork. The upper common bile duct showed a circular, contrast medium uptake by wall thickening causing stenosis of the lumen. Similarly, the right and left intrahepatic bile ducts showed a wall thickening and stenosis. These imaging chances suggested newly developed Secondary Sclerosing Cholangitis (SSC) in the context of the known AIP (Figure 2).

Cytology obtained via Fine Needle Aspiration (FNA) revealed epithelial cells with eosinophilic cytoplasm. The esophago-gastroduodenoscopy showed negative results of helicobacter pylori by histology and the helicobacter urease test. The Endoscopic Ultrasonography (EUS) revealed an enlarged, poorly demarcated pancreas head of 28 × 22 mm appearing as suspected carcinoma.

The treatment of the patient was initiated with Prednisolone (PRED) as tablets of 40 mg daily for 4 weeks with following tapering, which resulted in a prompt improvement of LTs as shown above (Table 1), and was accomplished in line with Japanese guideline recommendations of 2017 that also discussed the use of azathioprine (AZA) for patients with relapse [7]. AZA was given to the current patient at daily doses of 100 mg in an initially overlapping fashion with PRED and later alone for around 3.5 months until recovery of LTs in line with 2017 recommendations from Italy [8]. Regular LT testing was indicated to exclude high values prone to reinstallation of the PRED therapy. The clinical course was characterized by two flares after substantial sun exposure. During the treatment with PRED and AZA, the LTs undulated for a long time despite high AZA doses (Table 1). The LT fluctuation may have had several causes: (1) the current patient had in association with his AIP a severe secondary sclerosing cholangitis of intrahepatic small bile ducts with

stenoses and prestenotic dilatations; (2) the patient might have more benefitted if the treatment with PRED and AZA would have been replaced by immunomodulatory action of Ursodeoxycholic Acid (UDCA) to accelerate bile acid secretion, an option not discussed in earlier consensus recommendations nor in reports published in the years the patient was under medical care; and (3) the patient was under casual low level sunlight exposure because this risk factor was not recognized after the first flare (Table 1).

To overcome the consequences of exocrine pancreatic insufficiency, regular substitution with pancreatic enzymes was accomplished by capsules containing each 25.000 Units lipase activity, 18.750 amylase activity, and 1.125 Units protease activity [9,10]. In addition, Vitamin D capsules 3500 µg each 21 days was taken to restore the lipid soluble vitamin D levels. Prior to the first AIP flare, the body weight was 88 kg, which increased slightly up to 90 kg under PRED treatment and returned down to around 81 kg after treatment with PRED was ceased.

In this special and rare case report, the patient was a retired chief physician of a large Department of Gastroenterology with good expertise allowing for critical evaluation of case data. After the first flare, an internet search was undertaken regarding possible causative agents that may have triggered his initially AIP but in all publications, it was stated that causatives were unknown. As an outdoor fan caring for a large garden, he was often working outside under sunlight exposure, not only prior the first flare but also for the following years. The last AIP flare developed after sitting outside of an Italian restaurant for lunch with his family, exposed to bright sunlight for around two hours. Few days later he observed a dark urine and scleral jaundice. Laboratory data showed recurrence of his AIP that prompted reintroduction of the usual treatment. From now on, however, the patient avoided sunshine exposures after having recognized at least a temporal association of sunlight exposure with the recurrent development of the two AIP flares. This initially observed temporal association was promoted to a causal association because the first flare was also connected with substantial sunshine exposures for several consecutive days. As a result, his gardening was confined since the second flare to non-sunshine conditions using a wide brimmed Panama hat just to be at the safe side. The last treatment was terminated after almost 5 years (Table 1).

For the patient, his novel AIP can clearly be attributed to sunshine exposure classified as a seasonal disease occurring from spring to autumn, the best time for gardening. After years of battling with AIP flares now under conditions of consistent avoidance of sunshine exposure, no flares occurred any more as evidenced by normal LTs. At the occasion of the last ultrasound examination, there were minimal alterations of the intrahepatic bile ducts, but as this was now associated with normal activities of serum GGT, the ultrasound findings were classified as clinically not relevant. However, the pancreas became atrophic as evidenced by ultrasonography and required continued substitution with pancreatic enzymes to replace the exocrine pancreatic function.

The case data of the current patient allows for a critical analysis and helps summarize the 15 key features of this novel AIP: (1) Significant exposure to sunlight as novel and verified cause, not described for any of the three known AIP types; (2) male gender, age 76 years, white ethnicity of European origin, genetic predisposition with respect to immune familiar glaucoma in this special case; (3) not related to IgG4 as based on normal serum IgG4 levels; (4) special immune-based disruption preferentially of the pancreas; (5) clinical signs of painful acute pancreatitis were missing under conditions of normal serum lipase activities; (6) associated with SSC as characterized by prestenotic dilatations above stenoses of the hepatic fork and within intrahepatic bile ducts; (7) association with cholestatic and hepatocellular liver injury as evidenced by increased LTs; (8) related to the R value of the Roussel Uclaf Causality Assessment Method (RUCAM) (Table 1) [6], the liver injury pattern was of the cholestatic type at the first flare but it changed to the mixed hepatocellular-cholestatic type at the second flare; (9) mediators likely ensure cross talking and help function the skin-pancreas axis, pancreas-biliary system axis, pancreas-liver axis, and biliary system-liver axis; (10) due to insufficiently treatable with PRED and/or AZA alone, PRED and Ursodeoxycholic Acid (UDCA) capsules 250 mg bid are likely a better therapeutic option; (11) disruption is responsible for the clinical outcome with exocrine pancreatic insufficiency due to organ burnt out and organ atrophy and remains an issue, in addition to persisting xerostomia likely due to hyposalivation by exocrine salivary gland insufficiency; (12) good news at the end, there was no transition to a vanishing bile duct syndrome; (13) the last HbAc1 value was with 5.9% only minimally above the Upper Limit of Normal (ULN), indicating almost regular insulin secretion by the pancreatic Langerhans islets; (14) with sunshine exposure as established culprit, this novel AIP case cannot be included in cohorts of AIP-1 and AIP-2 defined as diseases without known cause; and finally (15) avoiding direct sunlight exposure prevented disease recurrence.

In a broader sense, AIP cases including AIP-3 caused by Immune Checkpoint Inhibitors (ICIs) [11] belong to the large group of immune and autoimmune disorders comprising liver diseases [12-16] with diagnoses verified by RUCAM [6] in reference to Drug-Induced Autoimmune Hepatitis (DIAIH) [12], idiosyncratic drug-induced anti-CYP autoimmune hepatitis with antibodies against microsomal Cytochrome P450 (CYP) [13,14], HLA-based immune idiosyncratic DILI with reference to Human Leukocyte Antigens (HLA) [14,15], immune idiosyncratic DILI with Stevens-Johnson Syndrome/Toxic Epidermal Necrolysis (SJS/TEN) [16], and immune idiosyncratic DILI by immune checkpoint Inhibitors (ICIs) [11]. On top and interesting is the observation that a subtype of the immune idiosyncratic DILI with SJS/TEN is also triggered by sunlight exposure [16,17] and that ICIs cause not only immune DILI [18] but also AIP-3 associated with SSC [19]. Sunshine exposure is also a causative of the immune Porphyria Cutanea Tarda (PCT) that affects non-skin organs like the liver [20,21]. Under pathogenic and analytical aspects, the current AIP is partially associated other diseases triggered by UV sunshine exposure that leads to internal disorders like the liver but not the pancreas, among these are Porphyria Cutanea Tarda (PCT) that clinically manifests as photosensitive dermal disease and liver involvement. The variability of organ participation is due to a functioning skin-liver axis via cross talking at the molecular immune level involving mediators through a functioning skin-liver axis [21]. A similar pathomechanism may apply for the skin-pancreas axis in the current AIP case.

Discussion

The novelty of the current AIP case report is attributable to the first description that significant sunshine exposure can trigger AIP (Table 1). The temporal association was ascertained by causal association via a positive unintentional exposure test result, a classic approach viewed as gold standard as known from studies in search for drugs as firm causatives such as idiosyncratic drug-induced liver

injury iDILI [6]. The implication of sunlight exposure in AIP was detected by chance in a single patient and is a reminder to carefully search for details in any patient with a disease conflicted by case detail uncertainties. Careful interpretation of the case allowed for the transition of case idiopathy reflecting a disease without ascertained cause to case idiosyncrasy, an unusual individual reaction to an exogen trigger. Despite the actual appreciation based on a single patient, data presented in this case need to be taken with caution and require confirmation by other reports. The current AIP case with established culprit is clearly different form AIP-1 and AIP-2 that are defined by lack of causatives. Previous AIP reports focused on many aspects of AIP, which help provide good medical care for AIP patients [22-30]. However, AIP cohorts are characterized by heterogeneity that impairs uniformity of diagnostic approaches and veritable therapeutic options as standard therapy and for reducing recurrences, requiring yet much clarifying work. Efforts should also consider the use of UDCA to mitigate cholestasis. After all, the AIP community expects updated guidelines in Japan and Europe, globally agreed upon and harmonized between East and West regions.

Conclusion

This report adds to actual knowledge in the AIP field and encourages AIP experts to intensify their focus on possible causes of future AIP cases. Preference should be given to drugs as causatives, an aspect largely neglected so far because medical history in AIP reports often remain vague. However, if patients were elderly with increased comorbidities in need of major comedication, there are good chances to find drugs as culprits, reducing thereby AIP cases conflicted by idiopathy. A comprehensive history includes conventional medications, herbal medicines, and occupational exposure to chemicals. The current new AIP case with its detailed description will open the door for AIP experts to foster updates of existing AIP guidelines via refining.

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