اپروچ به آقای ۴۶ ساله با یافته پانکراتیت اتوایمیون در سونوگرافی در درمانگاه پزشکی خانواده

ارائه دهنده: فاطمه اصلی، اینترن پزشکی خانواده دانشگاه علوم پزشکی تهران

استاد راهنما: خانم دکتر منصوری، عضو هیئت علمی گروه داخلی دانشگاه علوم پزشکی تهران

Chief complaint:

■ مراجعه با سونوگرافی (TAS) همراه با شواهد مبتنی بر Autoimmune pancreatitis به
 صورت یافته تصادفی به در مانگاه پزشکی خانواده

Code:1291752	Age:
اقائ داؤؤة شادمادنا	Date of type:



مرکز تصویربرداری نصبر نیسوی

Dear Dr: JALALI

On sonography of the liver and biliary tract (transabdominal approach using 3.5 MHz curved probe);

The liver is normal in size and has diffusely increased echogenicity with normal vascular borders suggesting diffuse fatty liver grade I.

No space-occupying lesion is evident in the liver.

The bile ducts and portal vein are of normal diameter

The gallbladder is of normal wall-thickness and is devoid of any stone.

Incidental finding:
Diffuse thickening of the pancreas is noted without obvious
peri pancreatic inflammation and aynamic pancreas MRI for
R/O of autoimmune pancreatitis

Yours sincerely, Dr.Behnejad Radiologist



اسلامشه ب ب ب ب



History of present illness:

- بيمار آقای 46ساله mental retard که با مادر خود زندگی می کند
 - ◄ بیمار شکایت یا علامت خاصی ندارد
 - سابقه بیماری قبلی گزارش نمیشود

Past medical history:

■ سابقه mental retardation از بدو تولد

Drug history:

medication

Sertraline

Chlordiazepoxide

Quetiapine

Social and habitual history:

■ یافته به خصوصی گزارش نشد

Allergy history:

■ حساسیت به مورد خاصی گزارش نشد

Family history:

◄ در سوابق خانوادگی یافته خاصی گزارش نشد

Physical exam:

- آقای میانسال mental retard هوشیار و ارینته
 - در مشاهده بیمار ایکتریک نیست
- ملتحمه بیمار pale نبوده و اسکلرا ایکتریک نیست
 - ح در سمع قلب S1 و S2 بدون سوفل شنیده شد
- ح در سمع ریه ها صدا های نرمال ریوی به صورت قرینه سمع گردید
- در معاینه شکم نرم و بدون دیستنشن، توده قابل لمس، تندرنس، گاردینگ و یافته غیر طبیعی دیگر بود
 - ارگانومگالی مشاهده نشد
 - ◄ معاینه غده تیروئید بیمار نرمال بود و یافته غیر طبیعی نظیر گواتر مشاهده نشد
 - در معاینه غدد بزاقی و اشکی بیمار یافته غیر طبیعی نظیر بزرگی و یا تندرنس مشاهده نشد
 - تندرنس مشاهده نشد CVA -
 - ◄ پوست بیمار در معاینه نرمال بوده و یافته غیرطبیعی مشاهده نگردید
 - لنفادنوپاتی شکمی، سرویکال و یا آگزیلاری در معاینه لمس نشد
 - ◄ نبض های انتهایی اندام ها پر و قرینه لمس شد

Autoimmune pancreatitis (AIP):

- An infrequently recognized disorder of presumed autoimmune etiology
- Characteristic clinical, histologic, and morphologic findings
- Can occur as a primary pancreatic disorder or in association with other disorders of presumed autoimmune etiology, including IgG4 cholangitis, salivary gland disorders, mediastinal fibrosis, retroperitoneal fibrosis, tubulointerstitial disease, and inflammatory bowel disease
- Several publications have indicated that AIP is actually an IgG4 systemic disease

AIP types:

- Autoimmune pancreatitis is now classified into two types. In type 1 AIP, the pancreas is involved as one part of a systemic IgG4-positive disease and meets the HISORt criteria
- Type 2 AIP, or idiopathic duct-centric pancreatitis, is characterized by granulocytic lesions, but without IgG4-positive cells and without systemic involvement.
- In a study of 97 patients with AIP, patients with type 1 AIP were found to have a high relapse rate compared with patients with type 2 AIP who do not experience relapse (49 versus 0 percent)
- In addition, at presentation patients with type 1 AIP were older, had a greater prevalence of IgG4 elevations, and were more likely to have biliary tract disease, retroperitoneal, renal, or salivary gland disease than patients with type 2 AIP. Type 2 AIP is more frequently associated with inflammatory bowel disease than type 1.

What are IgG-4 related diseases?

- The commonly shared features include tumor-like swelling of involved organs, a lymphoplasmacytic infiltrate enriched in IgG4-positive plasma cells, and a variable degree of fibrosis that has a characteristic "storiform" pattern
- In addition, elevated serum concentrations of IgG4 are found in 60 to 70 percent of patients with IgG4-RD
- The majority of patients **respond to glucocorticoids**, particularly in early stages of disease. In some subsets of organ disease (eg, pancreatitis), glucocorticoid responsiveness has been considered one **diagnostic criterion** for the disorder
- Major presentations of this protean condition, which often affects more than one organ, include:
- Type 1 (IgG4-related) autoimmune pancreatitis
- Salivary gland disease
- Orbital disease
- Retroperitoneal fibrosis

Clinical manifestations:

- A number of clinical features have been described that may reflect different stages of autoimmune pancreatitis (AIP)
- The manifestations occur in the pancreas, biliary tract, and other organs

Pancreatic manifestations:

- A pancreatic mass that can be confused with pancreatic carcinoma or lymphoma. A pancreatic mass, enlargement, or prominence on imaging was present in 85 percent of patients in an early report of 26 patients
- Mild abdominal pain with or without attacks of acute pancreatitis and chronic pancreatitis. Recurrent pancreatitis is common (25 percent in a series of 87 patients) and occurs more frequently in patients with focal compared with diffuse pancreatitis (33 versus 13 percent). Although recurrence is common, AIP is not a frequent cause of idiopathic recurrent pancreatitis, at least in Western countries
- Pancreatic duct strictures
- Peripancreatic vascular complications are rare, although in the series cited above they were present in 23 percent of patients with AIP

Biliary tract manifestations:

- The most common presentation of AIP is obstructive jaundice. The term IgG4associated cholangitis (IAC) has been proposed for the biliary tract manifestations of AIP
- An obstructive pattern of liver function tests (ie, a disproportionately elevated serum alkaline phosphatase and minimally elevated serum aminotransferases) is commonly seen in patients with biliary tract involvement. These features can also be seen in patients with pancreatic cancer or cholangiocarcinoma.

Other manifestations:

A number of other organs can be involved in patients with AIP. These include the salivary glands (Sjögren's syndrome), bile duct strictures, lung nodules, autoimmune thyroiditis, and kidney (interstitial nephritis with an IgG4-positive plasma cell infiltrate and IgG4 deposits in the tubular basement membrane)

Diagnosis:

- Correct diagnosis can help avert the consequences of progressive disease and unnecessary surgery
- An important part of the differential diagnosis is distinguishing AIP from pancreatic cancer
- AIP should be considered particularly in those with other autoimmune conditions
- A pancreatic biopsy is usually required to establish the diagnosis
- The diagnostic criteria proposed by the Mayo Clinic (the "HISORt" criteria) are most commonly used in the United States

■ The "HISORt" criteria:

It is defined by the presence of one or more of the following:

- Diagnostic Histology
- Characteristic Imaging on computed tomography
 (CT) and/or pancreatography
- Elevated serum IgG4 levels on Serologic testing
- Other organ involvement
- Response of pancreatic and extrapancreatic manifestations to glucocorticoid therapy

Histology:

- Endoscopic ultrasound (EUS)-guided fine-needle aspiration (FNA) may be an option for obtaining histologic specimens from the pancreas
- The following histologic findings are highly suggestive or diagnostic of AIP:
- A lymphoplasmacytic sclerosing pancreatitis or more than 10 lgG4-positive cells with at least two of the following: periductal lymphoplasmacytic infiltrate, obliterative phlebitis, and acinar fibrosis (type 1)
- Idiopathic duct-centric pancreatitis (IDCP) is characterized by a granulocytic epithelial lesion in the pancreatic duct with minimal IgG4positive cells in the pancreatic parenchyma (type 2)

Imaging:

- AIP is usually first suggested by an imaging test such as contrast-enhanced
 CT or magnetic resonance imaging (MRI)
- The main findings that are diagnostic or highly suggestive of AIP are a diffusely enlarged pancreas with featureless borders and delayed enhancement with or without a capsule-like rim
- By contrast, findings that are highly suggestive or diagnostic of pancreatic cancer include a low density mass, dilatation and/or cutoff of the pancreatic duct, and distal pancreatic atrophy
- A stricture in the common bile duct or the finding of a lesion in the head of the pancreas often prompts consideration of malignancy. Thus, it may not be possible to distinguish AIP from pancreatic cancer based solely upon imaging tests

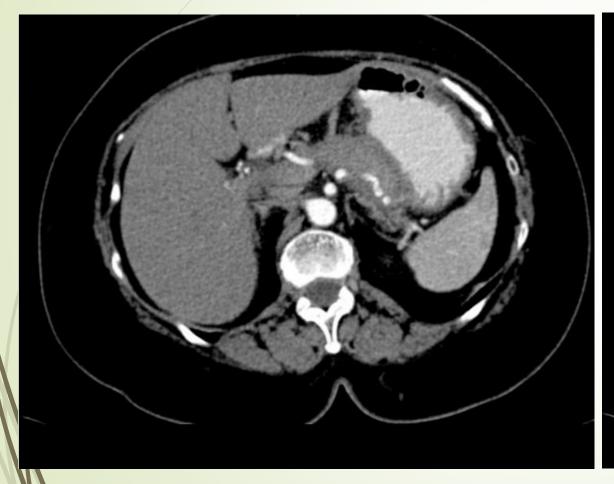
Normal pancrease in CT scan:







AIP in CT scan:

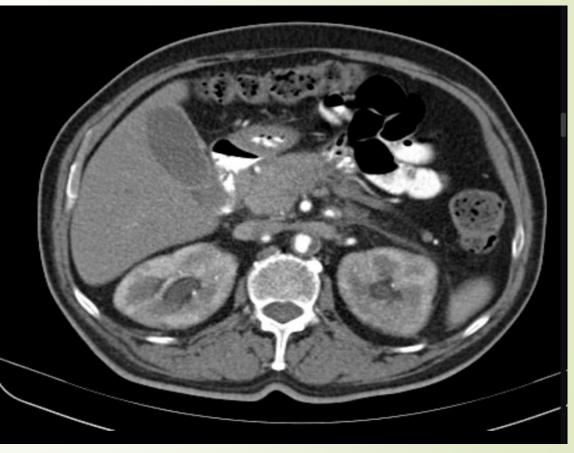




Diffuse enlargement of the pancreas with loss of definition of pancreatic clefts, so-called "sausage-like appearance". Minimal peripancreatic fat stranding, with inflammatory thickening confined to the peripancreatic region. Peripancreatic rim of low attenuation "halo" is well seen in the pancreatic phase. Right hepatic lobe segment VI small enhancing focal

lesion (incidental finding).





The pancreas appears bulky, specifically the pancreatic head and uncinate process, surrounded by minimal peripancreatic fat stranding. Multiple enlarged peripancreatic lymph nodes can be appreciated in the suprapancreatic and pancreaticoduodenal stations.

The common bile duct appears dilated, with evidence of distal common bile duct wall thickening, and associated intrahepatic biliary tree dilatation, more marked in the left lobe of the liver.

The liver, however, is of normal size and contour, with no focal liver lesions.

The abdominal aorta appears diffusely diseased, with multiple atherosclerotic plaques. A low attenuation, circumferential periaortic mass is seen surrounding the abdominal aorta, superior mesenteric artery, right colic artery and inferior mesenteric artery, and extends down to involve both common iliac and internal iliac arteries. This mass appears isodense to the abdominal aorta and IVC in the delayed phase, in keeping with delayed enhancement, likely relating to advanced disease. No significant anterior displacement of the abdominal aorta or the IVC is seen.

Both ureters appear to be embedded within the retroperitoneal fibrotic mass, with medial deviation and abrupt tapering at the level of L4 vertebral body. Mild bilateral hydronephrosis is evident.

No focal lytic or sclerotic lesions are seen in the imaged portions of the spine or the bony pelvis.

Serologic testing for IgG-4:

- Serologic testing for IgG4 is an important component of evaluating a patient suspected of having AIP
- IgG4 accounts for only 5 to 6 percent of the total serum IgG in healthy subjects and is typically less than 140 mg/dL. A serum concentration of IgG4 that is ≥2 times the upper limit of normal is highly suggestive of AIP, but may not rule out pancreatic cancer

- Other serologic factors:
- Anti-PBP peptide antibodies: Anti-plasminogen-binding protein (PBP) peptide antibodies may help diagnose patients with AIP but can not distinguish between AIP and pancreatic cancer
- Other antibodies: antibodies to carbonic anhydrase II antigens (positive in 30 to 59 percent), lactoferrin (positive in 50 to 76 percent), and also rheumatoid factor, perinuclear antineutrophil cytoplasmic antibody, antinuclear antibody, antimitochondrial antibody, anti-smooth muscle antibody, and antithyroglobulin

Treatment:

- the response to glucocorticoid therapy is one of the HISORt criteria for the diagnosis of AIP
- most patients respond to glucocorticoid therapy
- The relapse rate is significant
- Patients are typically followed with serum IgG4 levels, liver biochemical tests, and occasionally by computed tomography (CT) scan while on therapy
- Prednisone at a dosage of 40 mg/day for four to six weeks followed by a taper of 5 mg/week.
- Toward the end of the four- to six-week induction period, we perform repeat imaging tests (typically computed tomography or magnetic resonance imaging, whichever was obtained initially) to evaluate for radiologic improvement
- Most patients destined to respond demonstrate clinical and/or radiologic improvement within the first four to six weeks. Failure to respond in this timeframe should raise the possibility of alternative diagnoses

Our role?



- In history: history of other autoimmune disorders, signs or symptoms suggestive of involvement of other organs
- In physical exam: any palpable mass or swelling, jaundice, etc
- Paraclinical data: abdominal CT scan or MRI, serologic tests like IgG-4 level, liver biochemical tests
- Refer to an internal medicine specialist

سطوح پیشگیری

- ✓ PRIMORDIAL PREVENTION
- ✓PRIMARY PREVENTION
- ✓ SECONDARY PREVENTION
- ✓TERTIARY PREVENTION
- ✓ QUATERNARY PREVENTION

PRIMORDIAL AND PRIMARY PREVENTION

■ با توجه به اینکه این بیماری پروسه اتوایمیون بوده و ریسک فاکتور های شناخته شده ای جز همراهی با سایر بیماری های اتوایمیون برای آن گزارش نشده است در سطوح primordial و primary اقدامات موثری قابل انجام نمی باشد

SECONDARY PREVENTION

- تلاش در جهت تشخیص به موقع و زودرس بیماری جهت جلوگیری از پیشرفت بیماری و نیاز به اقدامات جراحی
 - ◄ بیماران در مراحل اولیه پاسخ بهتری به کورتیکواستروئید ها می دهند

TERTIARY PREVENTION

- → فالوآپ به موقع و مناسب بیماران شناخته شده جهت پیگیری پاسخ به درمان
 - relapse شناخت به موقع بیماران دچار ⊸

QUATERNARY PREVENTION

→ شناخت بیماران دچار عوارض و درمان مناسب جهت بهبود کیفیت زندگی

Lab and paraclinical findings:







(11111) تاریخ پذیرش: ۱۴۰۴/۰۵/۰۹۱۰۱۴ تاریخ اعلام نتیجه: ۱۴۰۴/۰۵/۱۶ بيمه / قرارداد: تأمين اجتماعي

شماره پذیرش: (۵۳۱) ۶۵۷۶۹۶ تاریخ تولد: ۱۳۵۱۰۱۸۰۱ سن: ۶۶۶ نام درخواست کننده: نسیم−عبلاتی نخجیری تفصمی پزشکی فلز



Hematology	-	Units	Reference Rang	ge Differential
Test CBC WBC RBC Hgb HCT MCV MCH MCH RDH RDW-CV	4.95 14.8 43.7 88.3 29.9 33.9	10*3/uL 10*6/uL g/dL % fL pg % 10*3/uL	3.5 - 10.5 4.5-5.6 13.5 - 17.5 41.0 - 52.0 80-96 27-33 33-36 150 - 450	Neutrophils

Riochemistry		1	Jnits	Reference Range
Test FBS	Result 93	-1	mg/dL	IGF(Impaired- Fasting-Glucose) or Diabetic: More than 125 Pre-diabetic: 100-125 Normal: 70-100
			mg/dL	0.7 - 1.4
reatinine	1.1		mg/dL	High: More than 240 Borderline: 200 - 240
holesterol	179		index	Normal: Up to 200 High Risk: More than 6.0 Moderate Risk: 3.1 - 6
Atherogenic index				Low Risk: 0.5 - 3 Normal: Up to 5.0
	A STATE OF THE STA	Н	index	
Risk Factor (For C.A.D)	5.11	The state of the s	mg/dL	Up to 200
NON I POLY	122			20.50

Atherogenic index		H	index	Normal: Up to 5.0
Risk Factor (For C.A.D)	5.11	MR THE STREET	mg/dL	Up to 200
Triglyceride	132		mg/dL	30-50
HDL-Cholesterol	35		mg/dL	Less than 200
LDL-Cholesterol	91		meg/L	135 - 145
Sodium (Na)	144		meq/L	3.5-5.5
Potassium (K)	4.7		mg/dL	0.1-1.2
Total Bilirubin	0.9		mg/dL	0.1 - 0.5
Direct Bilirubin	0.3		U/L	Up to 37
SGOT (AST)	37		U/L	Up to 40
SGPT (ALT)	40		U/L	80 - 306
Alkaline phosphatase (ALP)	305		%	Diabetic: More than 6.4
HbA1C	5.0		index	Pre-Diabetic: 5.7 - 6.4 Normal: Up to 5.7 Diabetic: More than 140 Pre-Diabetic: 115- 140
estimated Average Glucose (eAG)	97			Normal: Up to 115
			g/dL	3.2 - 5.3
Albumin	4.5	The assay are p	erformed by	full automated biochemistry an

تاریخ پذیرش: ۱۴۰۴/۰۵/۱۹۱۱ ۱۴۰۴/۰۵/۱۴۰۱ تاريخ اعلام نتيجه: ٥٥:٥٥ ١٢٥٤/٥٥/١٤ بیمه / قرارداد؛ تأمین اجتماعی تا تا و نام خانوادی: اتای داود سدهای شماره پذیرش: (۳۱) ۶۵۷۶۰۶ تاریخ تولد: ۱۱/۵۱۰/۱۵ سن: ۴۶:

نام درخواست کننده: نسیم-عبادتی نخجیری تخصص پزشکی خانر

4 HBA1C 1403/12/09

Hormon	Anal	ysis
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	Daniela	Units	Reference Range
Test Carcinoembryonic Antigen (CEA)	Result0.2	ng/mL	Non-Smokers: Up to 3.0 Smokers: Up to 5.0
CA 19-9	7.1	IU/lit ng/mL	Up to 37 10 - 220
FerritinVitamin D3		nmol/L	Toxicity: More than 100 Deficient: Less than 20 Sufficient: More than 30 Insufficient: 20 - 30

Miscellaneous		Units	Reference Range
Test	Result	mg/L	24 - 1210
Immunoalobulin G*	1314 *	 mg/L	

Thyroid		Units	Reference Range
Test	Result	u[iU]/mL	0.3 - 5.1
TSH	3.6	d[lO]/IIIL	



BiocemistryII		Units	Reference Range
Test	Result		
Serum amylase	57	U/L	Up to 100
Serum amylase	20	U/L	Up to 60
Serum lipase	28	0,2	



Medical Lab Director Dr. M. Majidi

Medical Lab QC Officer
S. Arjmand



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Date of type:---1404/05/11 15:53



مرکز تصویربرداری نصبر نبسوی (داتر مسابیری)

Dear Dr: Ebadati nakhjiri

On CT-scan of the abdomen and pelvic cavity without contrast:

Evidence of dirty mesenteric sign and LNs with SAD up to 7 mm are seen in mesenteric root suggestive for panniculitis
The liver, spleen and the pancreas have normal size and density.
The gallbladder and biliary tree are within normal limits.
Both kidneys are normal in size and density.

There is not any visible stone in the kidneys and the course of the ureters.

The adrenal glands have normal appearance.

The abdominal aorta is normal in diameter.

Bladder wall thickness is increased.

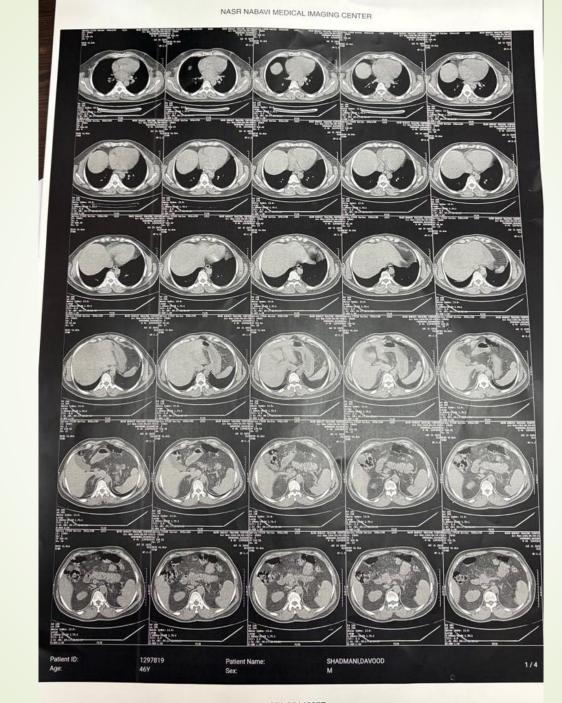
Prostate is enlarged

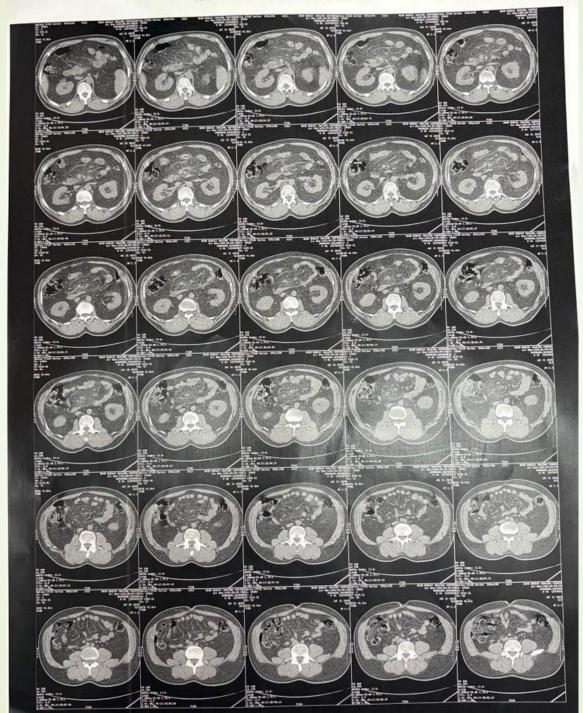
The rectum and other pelvic organs are seemingly normal. There is not any ascites in the peritoneal cavity.

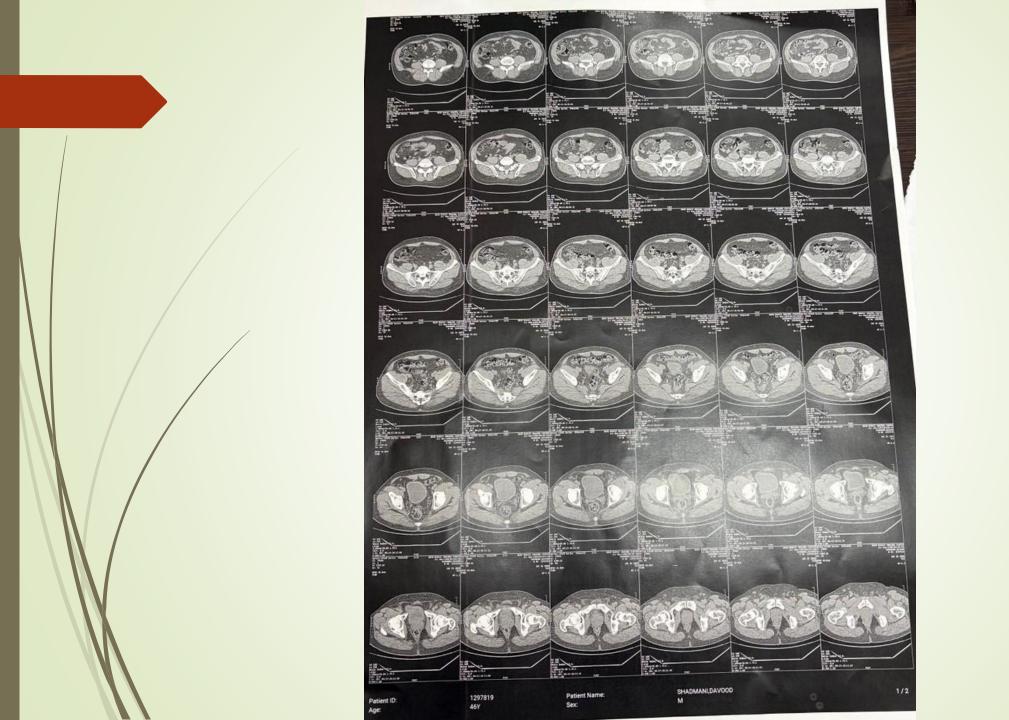
Yours sincerely, Dr.Moradi Radiologist











Jun 45	
1403/12/23	11:04
	1403/12/23



مرکز تصویربرداری نصر نبوی (داتر معد انبری)

Dear Dr: JALALI

On sonography of the abdomen, pelvic cavity and the retroperitoneum (transabdominal approach using 3.5 MHz curved probe);

The liver is normal in size and has diffusely increased echogenicity with normal vascular borders suggesting diffuse fatty liver grade I. No space-occupying lesion is evident in the liver.

The bile ducts and portal vein are of normal diameter.

The gallbladder is of normal wall-thickness and is devoid of any stone.

The spleen is of normal size and shows homogeneous echogenicity.

The pancreas is of normal size and echogenicity.

Neither ascites nor para-aortic adenopathy is visible.

Both kidneys are of normal size and echopattern (RK = 117 mm & LK = 115 mm).

Renal parenchymal thickness is in normal range on both sides (13 mm in right side and 14 mm in left side).

No detectable stone or hydronephrosis is present.

The bladder wall-thickness is normal. No S.O.L is seen within it.

The prostate is normal with 23 cc volume.

Yours sincerely, Dr.Shahrami Radiologist



Thank you for your attention