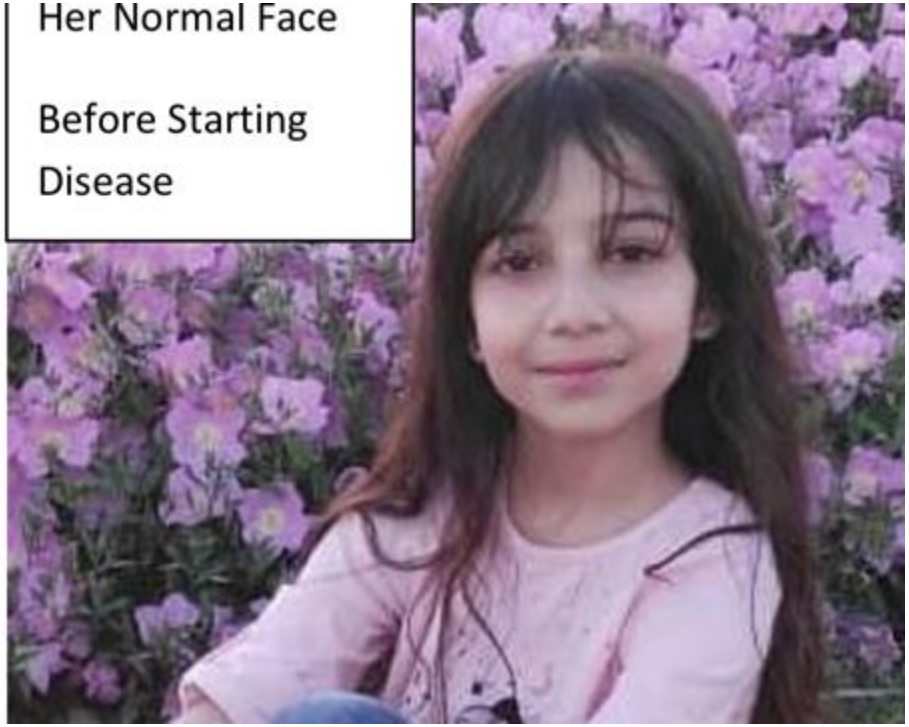

بسم الله الرحمن الرحيم



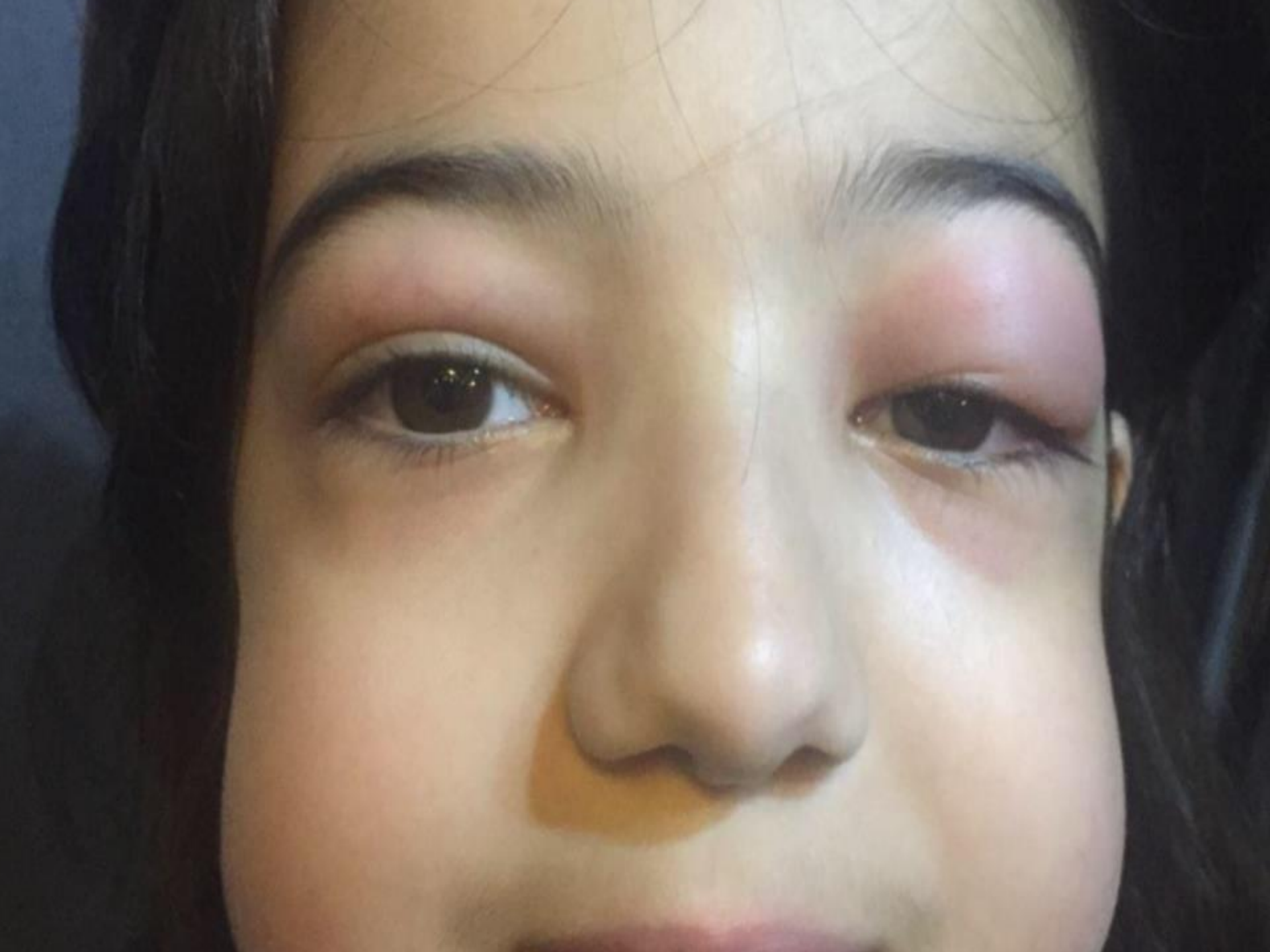
- 2019

CASE PRESENTATION

Our patiant was a ten-year-old girl who had a right periorbital swelling and fever for about one week before referral,

IN INFECTIOUS WARD

- who was initially treated with an antibiotic(*VAN+CEF*) for the diagnosis of pre-septal cellulitis, but gradually fever and left periorbital swelling were added to symptoms.



- *There was a history of systemic lupus erythematous (SLE) in the patient's grandmother and aunt, and grandmother died from a pulmonary complication.*

- *Sonographic evaluation showed extensive soft tissue edema in the face and upper neck particularly in superficial subcutaneous tissue*

- *The study of MIBG scintigraphy was negative.*

نام بیمار : خانم باران قدیری

SONOGRAPHY OF THE NECK , THYROID AND SALIVARY GLANDS:

Extensive soft tissue edema in the face and upper neck particularly in the superficial subcutaneous tissue are seen.

No sign of mass lesion or any obvious pathology is detected in the sites of edema .

No significant adenopathy is seen at anterior and posterior triangles and carotid sheaths and central neck.

Both thyroid lobes have normal size and homogenous echo pattern.

Right lobe measured 29×12×8mm and left lobe 29×12×10mm.

Isthmyc region has normal thickness (1mm) and echo, too.

Salivary glands and particularly parotids and submandibular are symmetric and normal.

No mass or any obvious pathology is defined.

Other cervical structures are symmetric and normal.

Both common carotid arteries, and main branches as well as jugular veins are roughly normal bilaterally.

Concerning the clinical signs and extension of the edema, unusual collusion vascular diseases such as dermatomyositis or polymyositis might be considered.

- ***MRI** images showed inflammatory signal changes in subcutaneous area of bilateral cheek and submandibular region in favor of cellulitis without evidence of collection or lymphadenitis.*

7 May 2019

صویربرداری پزشکی

هار

مرکز تصویربرداری پزشکی هار

تاریخ: ۱۳۹۸/۰۲/۱۷

شماره: ۱۲۴۸۴۱

پیوست:

نام بیمار : باران قدیری (کودک)

سن : ۱۰ سال

با سلام و احترام همکار گرامی جناب آقای دکتر سیامک بسیم فر

Buccal MRI (without contrast)

Dear colleague :

Technique: *Dedicated study is obtained with multiplanar and in different sequences with GE 1.5 tesla machine .*

- ***Inflammatory signal change is seen in subcutaneous areas of bilateral cheek and submandibular region. Cellulitis is first line diagnosis.***
- ***Collection and abscess formation is not presented.***
- ***Maxillary and mandibular bones are normal.***
- ***Submandibular glands and parotid glands are seen as usual.***

- *In laboratory studies only mild Leukopenia (3800) and lymphopenia occurred and other inflammatory markers and markers such as ESR and CRP were mildly elevated.*

- *Bone marrow examinations were also performed for the patient with no evidence of abnormal cell involvement*

BMA AND FLOW

Takht-e Tavous Pathobiology lab
Molecular Diagnostic & flowcytometery Department

آزمایشگاه پاتوبیولوژی تخت طاووس
بخش تشخیص مولکولی و فلوسیتومتری

93-4402	
10y / F	
F-185	

DIAGNOSIS: BM immunophenotyping revealing a lymphoid population about 22.0% of total cells which mostly are mature T-cells with normal CD4/CD8 ratio and remaining are B-cells.

SPECIMEN :	Bone marrow
CLINICAL DATA :	R/O NHL and lymphadenopathy
CLINICAL IMPRESSION :	R/O Malignancy

FLOW CYTOMETRY IMMUNOPHENTYPING ANALYSIS

Gate: 900 (31.5%)

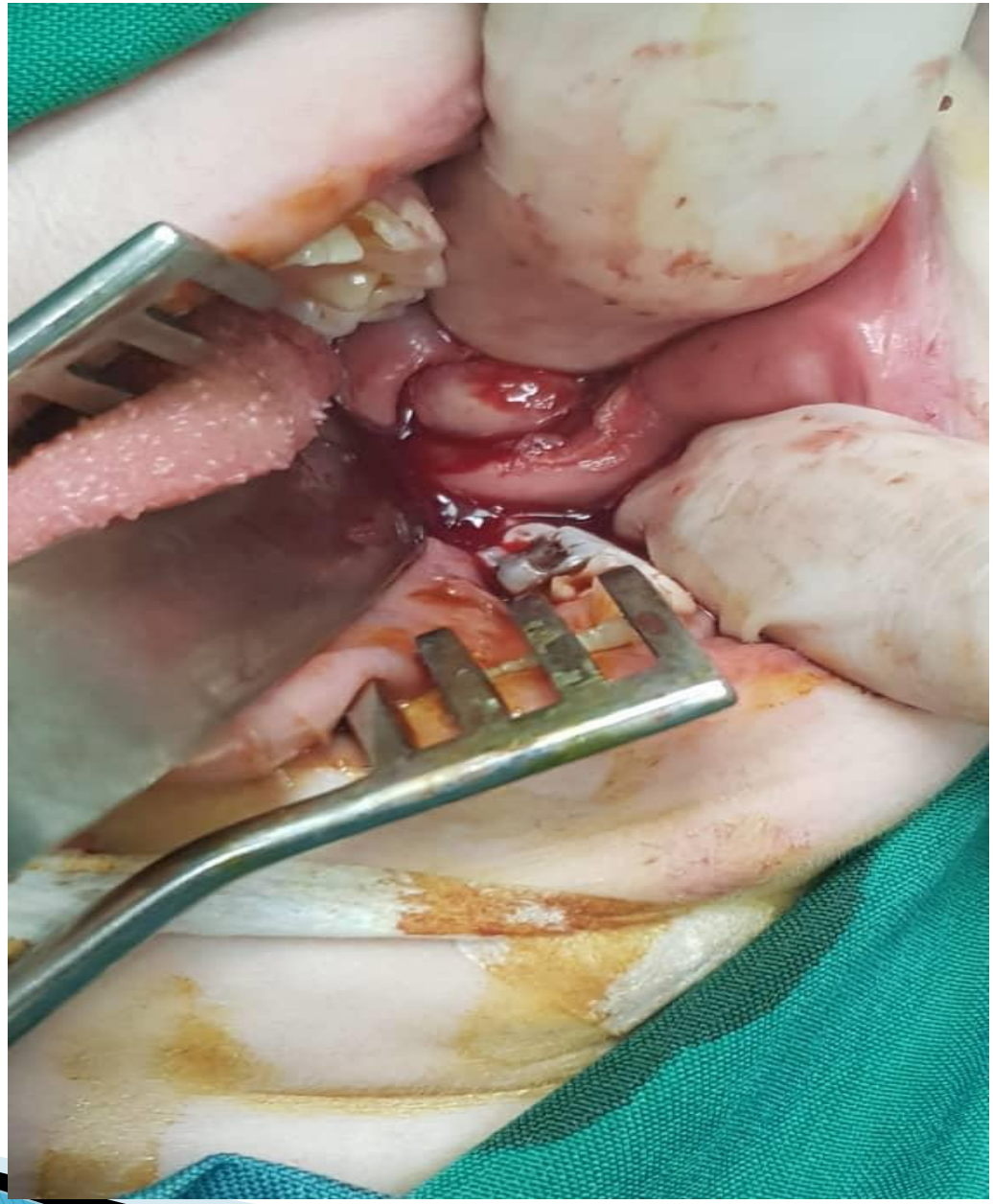
HLA-DR	33.3	CD11	9.1	CD22		CD103	
CD34		CD34		CD45		CD117	
CD2	89.2	CD35		CD56/CD57		CD138	
CD5	81.4	CD36		CD31	13.5	Glycophorin	
CD4	35.0	CD19	17.0	CD34	1.0	CD71	
CD8	56.7	CD20	15.0	CD38		TdT	
CD7	95.4	CD22		CD41		CD1/HLA-DR	10.1
CD4	34.4	FMC7		CD45	95.7	CD15/CD13	
CD8	0.2	cCD3		CD47 dim		CD14/CD64	
CD16		cCD22		CD61		CD65/CD24	
CD1c		cCD79a		CD68			

Myeloperoxidase:	
Viability :	>80.0%
Abnormal cells present by FCM:	-
Percentage of cells with abnormal phenotype:	-
Cell size:	-
PHENOTYPE :	-
MORPHOLOGY :	Diluted BM smear shows myeloid and erythroid series with shift to the left and mild megaloid and dysplastic changes, shift to the left, M/E ratio about 5/1, there are about 22-24% small sized lymphoid cells, presence of 3-4% normal looking plasma cells, few smudge cells, no increase in blasts, megakaryocytes also seen.

M.Aghanipour M.D

F.Jadali M.D
(Clinical & Anatomical Pathologist)
Fellowship in pediatric pathology

- *Finally, the patient underwent a biopsy*



شماره پذیرش : ۰۳-۴۴۷۲ تاریخ پذیرش : ۱۳۹۸/۰۳/۰۲ تاریخ جوابدهی : ۱۳۹۸/۰۳/۲۱ پزشک معالج : جناب آقای دکتر سیدمحمد کاظم

نام مراجعه کننده : **خانم باران قدیری** سن : ۱۰ **سال** شماره پاتولوژی : **S-98-1201**

چ: ۱ ۱

Specimen
Left buccal soft tissue mass

Clinical data
FUO , cervical lymphadenopathy , perioccular swelling , not responsible to antibiotic therapy
R/O lymphoma

Macroscopic
Specimen received in formalin & consists of an ovoid encapsulated elastic mass measuring 18x1.4x1.1cm. The cut surfaces are homogenous tan yellow solid and nodular.Block sum T.S in blocks No:1-3

Microscopic
Serial step sectioning show uniform mature adipocytes with intervening thin fibrous bands resulting nodular pattern infiltrated by polymorphous inflammatory infiltrate including lymphoplasmacells and neutrophils admixed with nuclear dusts. No malignancy.

Diagnosis
Left buccal soft tissue mass , excision:
- Fibrolipoma versus lipomatosis , see the comment please.
- No evidence of malignancy in this specimen.

Comment: According to mixed inflammatory cells infiltrate , infectious diseases should be ruled out.

دکتر مهناز آقائی پور
متخصص آناتومیکوپاتولوژی
عضو هیئت علمی
۲۱۶۵۸

M.Aghaei pour M.D

دکتر روح الله غلامپور شهاب الدینی
متخصص پاتولوژی دهانی و تشریحی
۱۴۲۱۳

R.Gholampour.M.D

Pathologists:

آزمایشگاه پاتوبیولوژی
تخت طاووس

- *The general illness and the persistence of fever and the lack of response to broad-spectrum antibiotics and the negative result of cultures and infectious studies,*

Takht-E-Tavoos pathology laboratory, Tehran
Sections prepared from all the blocks for H-E staining. The blocks were returned to the patient.

MICROSCOPIC DESCRIPTION:

Sections show portions of adipose tissue with patchy areas of necrosis and infiltration by individual and aggreg
of atypical lymphoid cells with small hyperchromatic nuclei showing irregular nuclear contours and scanty
cytoplasm. There is peripheral rimming of adipocytes by some atypical lymphoid cells. There is also marked
infiltration by foamy macrophages.
On IHC staining, there is marked infiltration of adipose tissue by CD3+, CD7+, CD8+, Granzyme B+, lymphoid
with typical peripheral rimming of adipocytes.

SECOND LOOK DIAGNOSIS:

LEFT BUCCAL MASS, SENT FOR SECOND OPINION AND IHC STAINING:
- MORPHOLOGIC STUDY AND IHC STAINING ARE IN KEEPING WITH SUBCUTANEOUS PANNICULITIS LIKE T-CE
LYMPHOMA

ICDO **C76.0**
M-9708/3

NOTE:
Please also see the IHC report AP-98-2332

13 Jun 201

Atieh Hospital
Anatomical & Clinical Pathology
Tehran:Shahrak-E-Gharb
TEL:88086583-84

شماره پرونده: ۷۲-۴۵-۵۰
پزشک معالج: نوربخش
تاریخ مراجعه: ۱۳۹۸/۰۳/۰۷
تاریخ جواب: ۱۳۹۸/۰۳/۲۲

آزاد
سرپایی

"IMMUNOHISTOCHEMICAL STUDIES F

CLINICAL INFORMATION: Not provided
SPECIMEN: Paraffin block number AP-98-2331

IHC MARKERS:
Immunohistochemical staining was done using antibodies against the follo

Markers:	**Description of reaction:**
1 – CD20:	Positive in few scattered small lymphoid cells
2 – CD19:	Positive in few scattered small lymphoid cells
3 – CD3:	Positive in many scattered small lymphoid cells wit
4 – CD7:	Positive in many scattered small lymphoid cells wit
5 – MPO:	Positive in few scattered macrophages
6 – C-kit:	Negative
7 – CD34:	Negative
8 – TdT:	Negative
9 – CD4:	Positive in few scattered lymphoid cells
10 – CD8:	Strongly positive in sheets and individual lymphoid adipocytes
11 – Granzyme B :	Strongly positive in sheets and individual lymphoid adipocytes
12 – CD56:	Negative

INTERPRETATION:
Morphologic study and IHC staining are in keeping with subcutaneous pann

NOTE
Please also see the pathology report AP-98-2331

M.Kadivar MD M.Karimi MD F.Kosari MD
M.Shahidi MD M.Sotoudeh MD M.Tavangar MD

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- *Biopsy showed lobules of fatty tissue with fibrinoid necrosis, small vessels were thrombosed and large vessels were surrounded by a number of lymphocytes, histiocytes and cell debris*

- and in *IHC slides showed the most lymphoid cells were positive for CD3,CD7,CD19,CD20 and strongly positive for CD8 and Granzyme B and negative for CD4,CD34,CD56,c-kit, TdT and MPO , suggesting*
- ***panniculitis and necrotizing leukocytoclastic vasculitis***

آزمایشگاه پاتولوژی بیمارستان توس

Toos Hospital Pathology Lab.

تاریخ گزارش : ۱۳۹۸/۰۴/۰۴

شماره پذیرش : ۹۸۰۴۰۰۰۰۳۵

سن : ۱۱

نام پزشک : دکترباهوش

شماره پاتولوژی :S-98-859

Clinical Data : Acute onset of fever, facial edema and erythema, with epidermal erosion, refractory to AB therapy.

Macroscopic : Received for consultation 3 blocks No 98-1201 with corresponding report from Takht Tavoos lab., dx as "Fibrolipoma vs lipomatosis,…". Moreover, there are 12 IHC slides from Atieh hosp., concluding the case as "..in keeping with subcutaneous panniculitis-like T-cell lymphoma". (No AP-98-2331).

Microscopic : This specimen shows lobules of fatty tissue with diffuse fibrinoid necrosis. The small vessels are thrombosed, the larger vessels are surrounded by a number of lymphocytes, histiocytes and cell debris. The histiocytes show phagocytosis. Necrotic fat cells are surrounded by lymphocytes, a few of them are slightly enlarged.
The IHC slides show that most lymphoid cells are positive for CD3, CD7, CD8 and Granzyme-B; a few CD19 and CD20+ B-cells are also present. CD4, CD34, CD56, c-kit, TdT and MPO are negative.

Diagnosis : Outside blocks No 98-1201 as left buccal mass:

- **NECROTIZING LEUKOCYTOCLASTIC VASCULITIS AND LOBULAR PANNICULITIS**

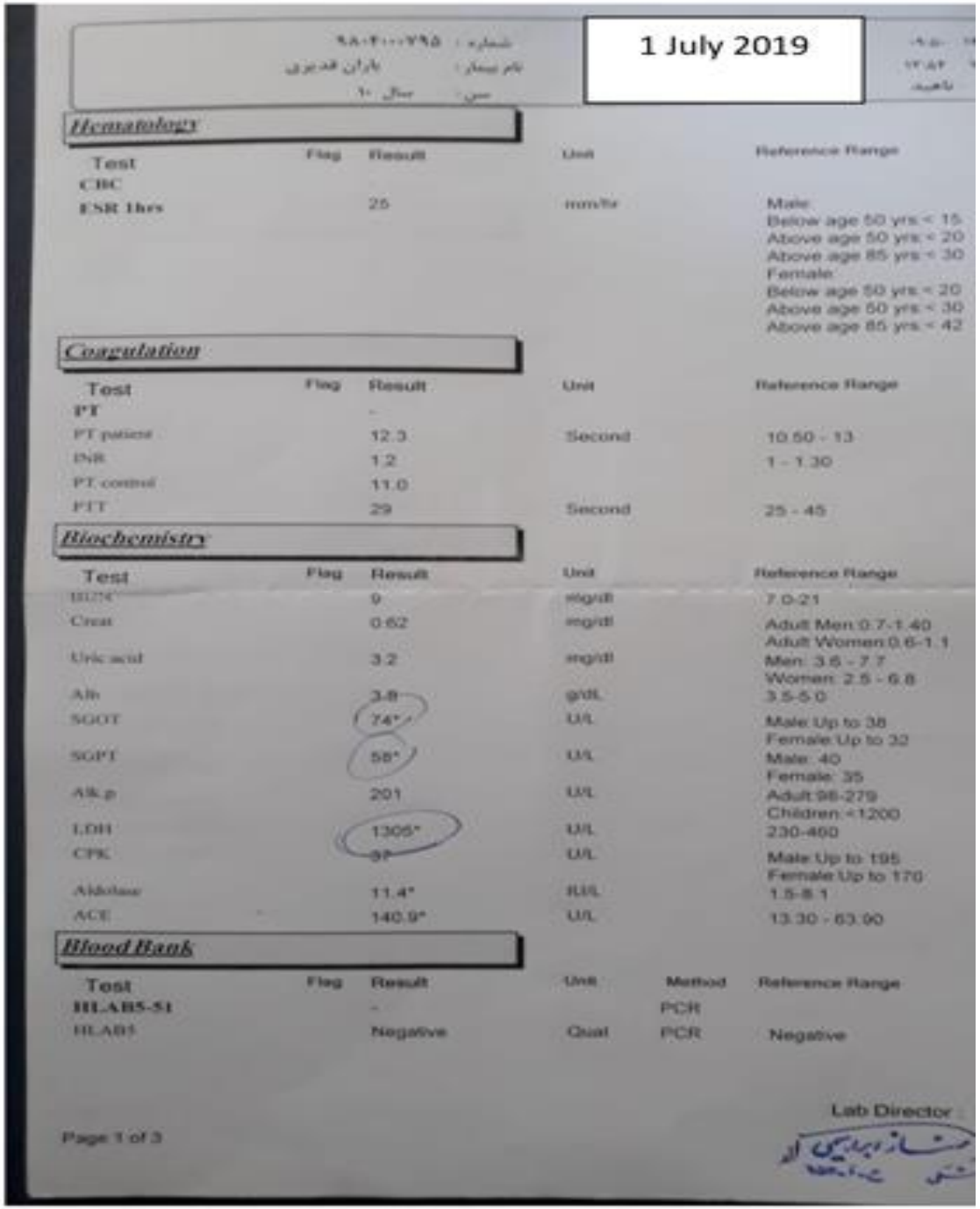
*** Rheumatologist consultation recommended.**

A.Zare MD , AP/CP

Y.Nilipour , Neuromyopathologist

- According to the positive *family history of SLE* in the family, sex of the child and *lymphopenia,* the study was performed for lupus and other rheumatologic diseases

- *In rheumatologic studies, the positive results included FANA with 1/320 titer and ACE 110 , but other autoantibodies and complement were normal.*



شماره : ۹۸۰۴۰۰۰۷۹۵
نام بیمار : باران قدیری
سن : سال ۱۰

1 July 2019

Hematology

Test	Flag	Result	Unit	Reference Range
CBC				
ESR 1hrs		25	mm/hr	Male: Below age 50 yrs < 15 Above age 50 yrs < 20 Above age 85 yrs < 30 Female: Below age 50 yrs < 20 Above age 50 yrs < 30 Above age 85 yrs < 42

Coagulation

Test	Flag	Result	Unit	Reference Range
PT		-		
PT patient		12.3	Second	10.50 - 13
INR		1.2		1 - 1.30
PT control		11.0		
PTT		29	Second	25 - 45

Biochemistry

Test	Flag	Result	Unit	Reference Range
BUN		9	mg/dl	7.0-21
Creat		0.62	mg/dl	Adult Men:0.7-1.40 Adult Women:0.6-1.1
Uric acid		3.2	mg/dl	Men: 3.6 - 7.7 Women: 2.5 - 6.8
Alb		3.8	g/dL	3.5-5.0
SGOT		74*	U/L	Male:Up to 38 Female:Up to 32
SGPT		58*	U/L	Male: 40 Female: 35
Alk.p		201	U/L	Adult:98-279 Children:<1200
LDH		1305*	U/L	230-460
CPK		9*	U/L	Male:Up to 195 Female:Up to 170
Aldolase		11.4*	IU/L	1.5-8.1
ACE		140.9*	U/L	13.30 - 63.90

Blood Bank

Test	Flag	Result	Unit	Method	Reference Range
HLAB5-51		-		PCR	
HLAB51		Negative	Qual	PCR	Negative

Lab Director

شماره : ۹۸۰۴۰۰۰۷۹۵

نام بیمار : باران قدیری

سن : سال ۱۰

1 July 2019

Blood Bank

Test	Flag	Result	Unit	Method	Reference Range
HLAB51		Negative	Qual	PCR	Negative

Serology

Test	Flag	Result	Unit	Reference Range
CRPstar		<2	mg/L	Adult :<10 Newborn Up to 3 weeks:<15
Dir Coombs		Negative	Titer	Nrgative

Immunology

Test	Flag	Result	Unit	Method	Reference Range
Anti ds DNA		7.1	IU/ml	ELISA	Negative <30 Gray Zone 30-35 Positive >35
FANA		-			
Titer		Negative	Titer		Up to 1/40
ANCA		-			
ANCA C		2.5	U/ml	ELISA	Normal Range <12 Equivocal Range 12-18 Positive Range >18
ANCA P		3.1	U/ml	ELISA	Normal Range <12 Equivocal Range 12-18 Positive Range >18
Anti Phospholipid Ab IgG		3.8	U/ml		Negative <10 Positive >=10
Anti Phospholipid Ab IgM		2.4	U/ml		Negative <10 Positive >=10
Anti Cardiolipin IgG		4.0	U/ml	ELISA	Negative <10 Positive >=10
Anti Cardiolipin IgM		4.5	U/ml	ELISA	Negative <10 Positive >=10
Anti coagulant lupus		Negative			
Anti-B2-Glyco(G,M)		-		ELISA	
Anti B2 Glyco(IgG)		<1.0	U/ml		Normal Range <12 Equivocal 12-18 Positive Results >18
Anti B2 Glyco(IgM)		<1.0	IU/ml		Normal Range <12 Equivocal 12-18 Positive Results >18
C3		138	mg/dl		90 - 180
C4		23	mg/dl		10 - 40
CH50		114	U		70-150

Lab Director :


11 July 2019

- *she was treated empirically with intravenous pulse methylprednisolone at a dose of 30mg / kg for three consecutive days, which was temporarily discontinued for several days, but again with recurrent fever and exacerbation of swelling and ulcerative lesions.*


Hospitalization
The Day of Biopsy from Buccal
23 May 2019

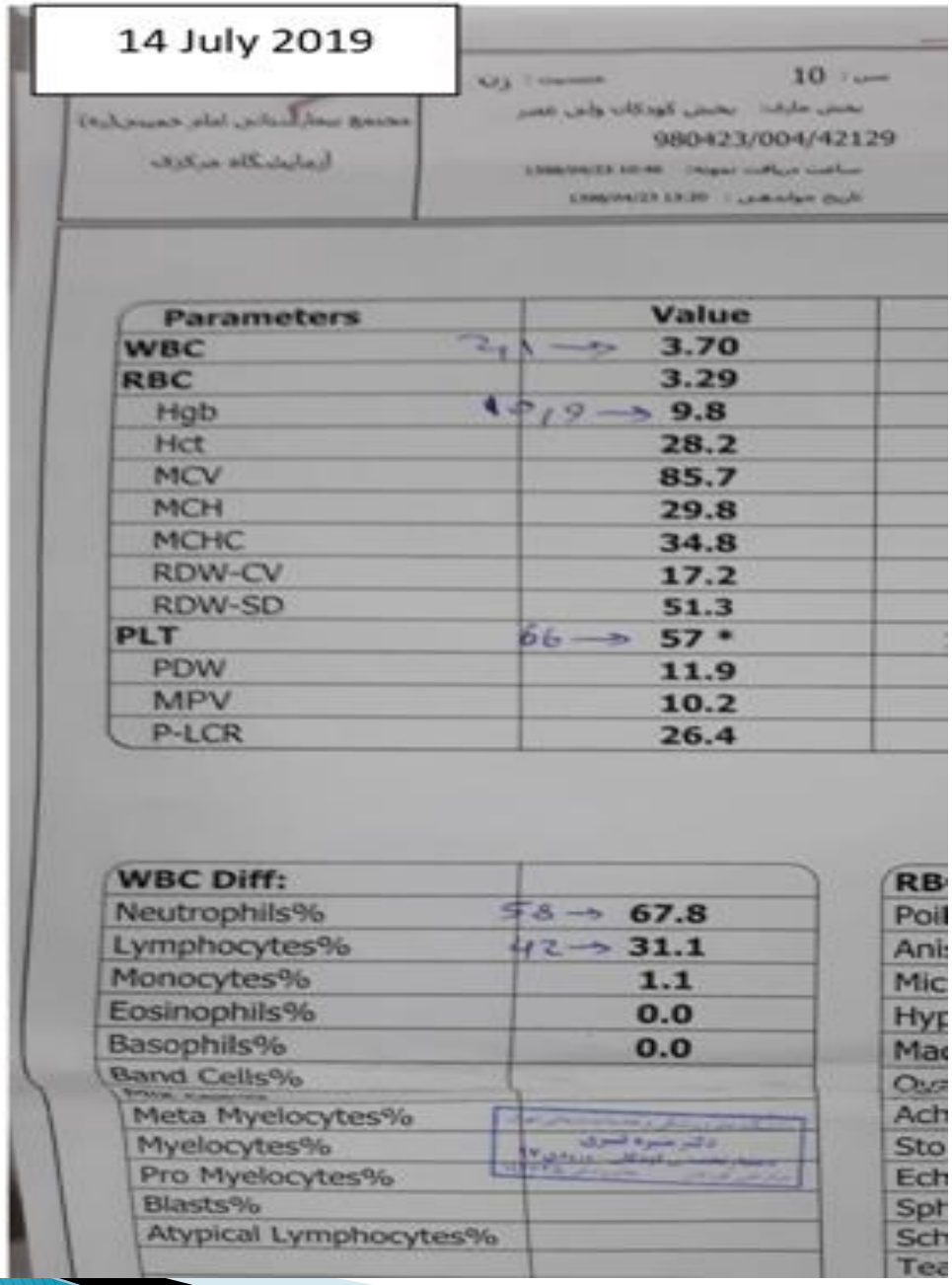
- *according to all the studies and the approximate rejection of possible SLE , with the diagnosis of*

  Subcutanous Like T cell lymphoma,

- *At the start of chemotherapy, the patient developed*
- *High grade fever,*
- *pancytopenia,*
- *high serum ferritin,*
- *High triglycerides level,*
- *Low serum fibrinogen*

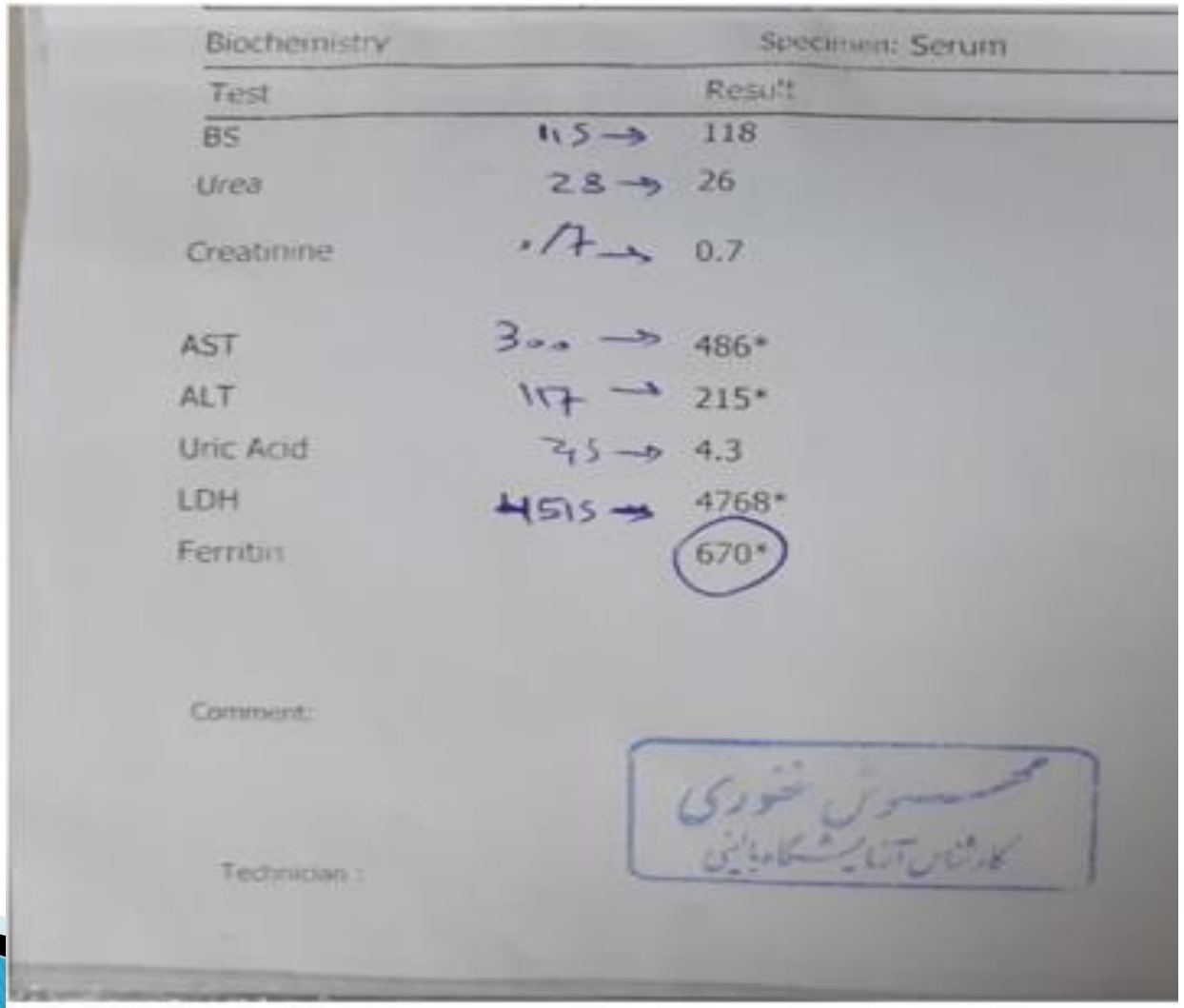

er Reaches
0 everyday

14 July 2019



Parameters		Value
WBC	2.1 →	3.70
RBC		3.29
Hgb	10.9 →	9.8
Hct		28.2
MCV		85.7
MCH		29.8
MCHC		34.8
RDW-CV		17.2
RDW-SD		51.3
PLT	66 →	57 *
PDW		11.9
MPV		10.2
P-LCR		26.4

WBC Diff:		
Neutrophils%	58 →	67.8
Lymphocytes%	42 →	31.1
Monocytes%		1.1
Eosinophils%		0.0
Basophils%		0.0
Band Cells%		
Meta Myelocytes%		
Myelocytes%		
Pro Myelocytes%		
Blasts%		
Atypical Lymphocytes%		



Biochemistry — Specimen: Serum

Test		Result
BS	115 →	118
Urea	28 →	26
Creatinine	.7 →	0.7
AST	300 →	486*
ALT	117 →	215*
Uric Acid	75 →	4.3
LDH	4515 →	4768*
Ferritin		670*

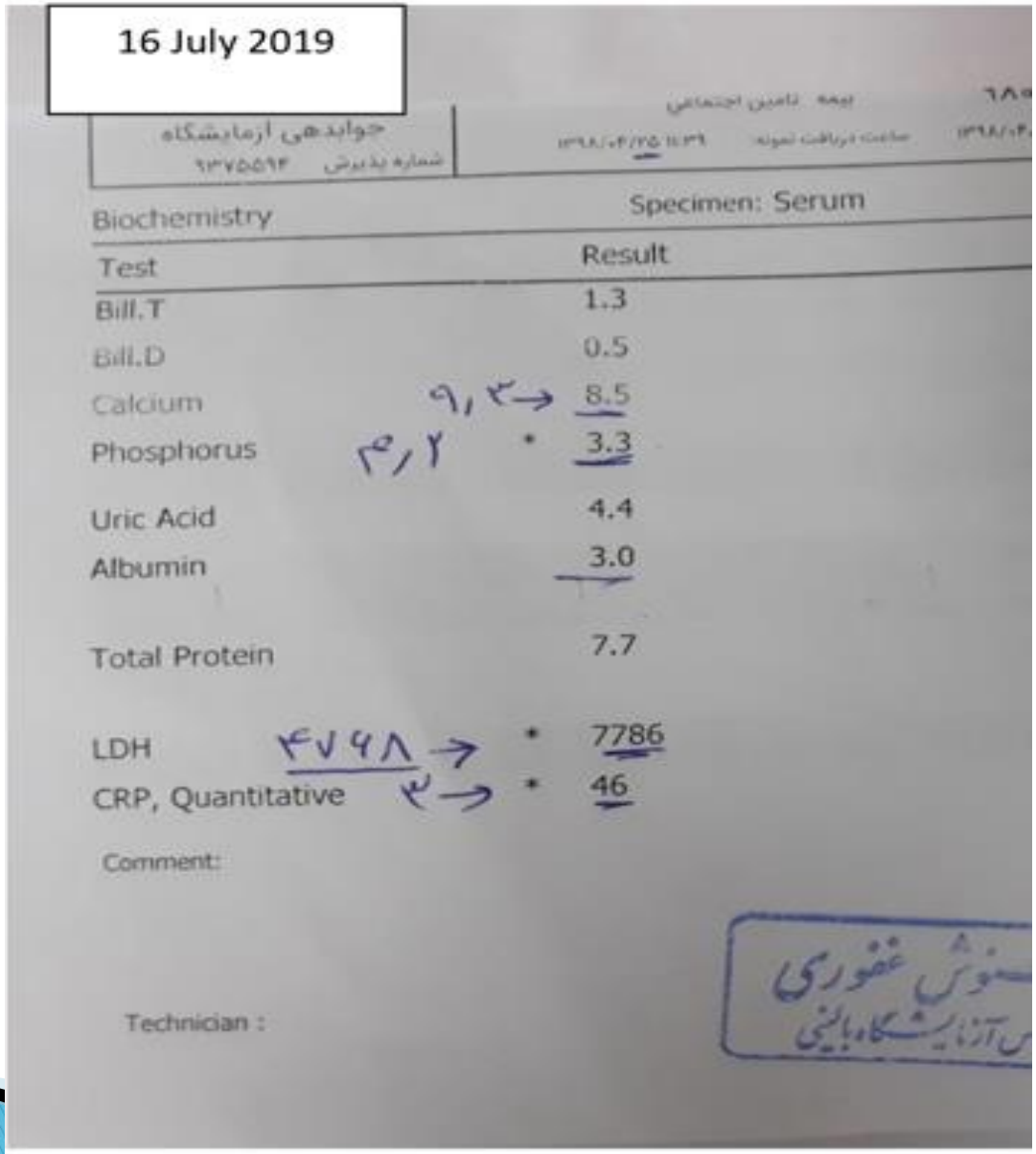
Comment:

Technician :

محسن غفوری
کارشناس آزمایشگاه بالینی

- *and again bone marrow examinations was performed, Hemophagocytosis images were observed, and hemophagocytic syndrome (MAS)*

16 July 2019



جوابدهی آزمایشگاه
شماره پذیرش ۹۳۷۵۶۹۴

بیمه تامین اجتماعی
ساعت دریافت نمونه ۱۳۹۸/۰۴/۲۵ ۱۱:۳۹

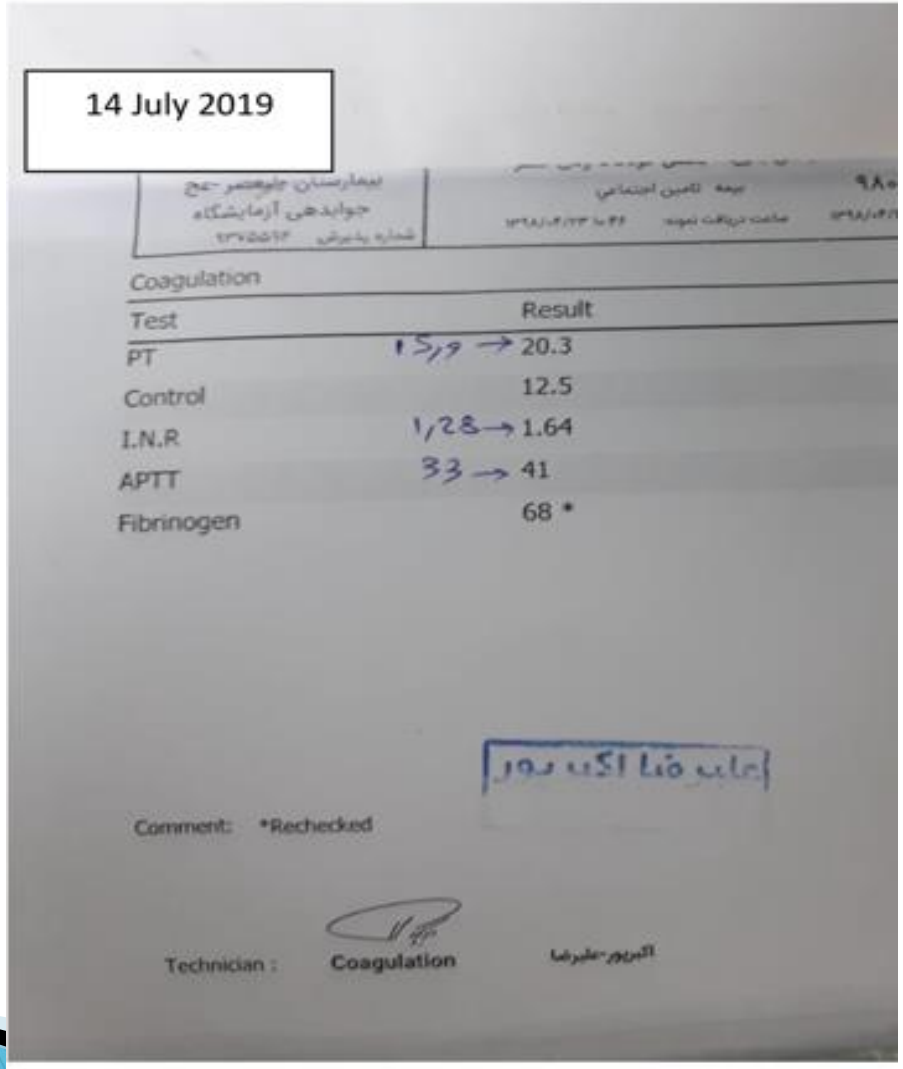
Biochemistry — Specimen: Serum

Test	Result
Bill.T	1.3
Bill.D	0.5
Calcium	8.5
Phosphorus	* 3.3
Uric Acid	4.4
Albumin	3.0
Total Protein	7.7
LDH	* 7786
CRP, Quantitative	* 46

Comment:

Technician :

14 July 2019



بیمارستان جلوهگر -عج
جوابدهی آزمایشگاه
شماره پذیرش ۲۳۰۵۵۶۴

بیمه تامین اجتماعی
مدت دریافت نمونه ۳۹۸/۰۴/۲۳ تا ۴۱

۹۸۰
۳۹۸/۰۴/۲۱

Coagulation

Test	Result
PT	15,9 → 20.3
Control	12.5
I.N.R	1,28 → 1.64
APTT	33 → 41
Fibrinogen	68 *

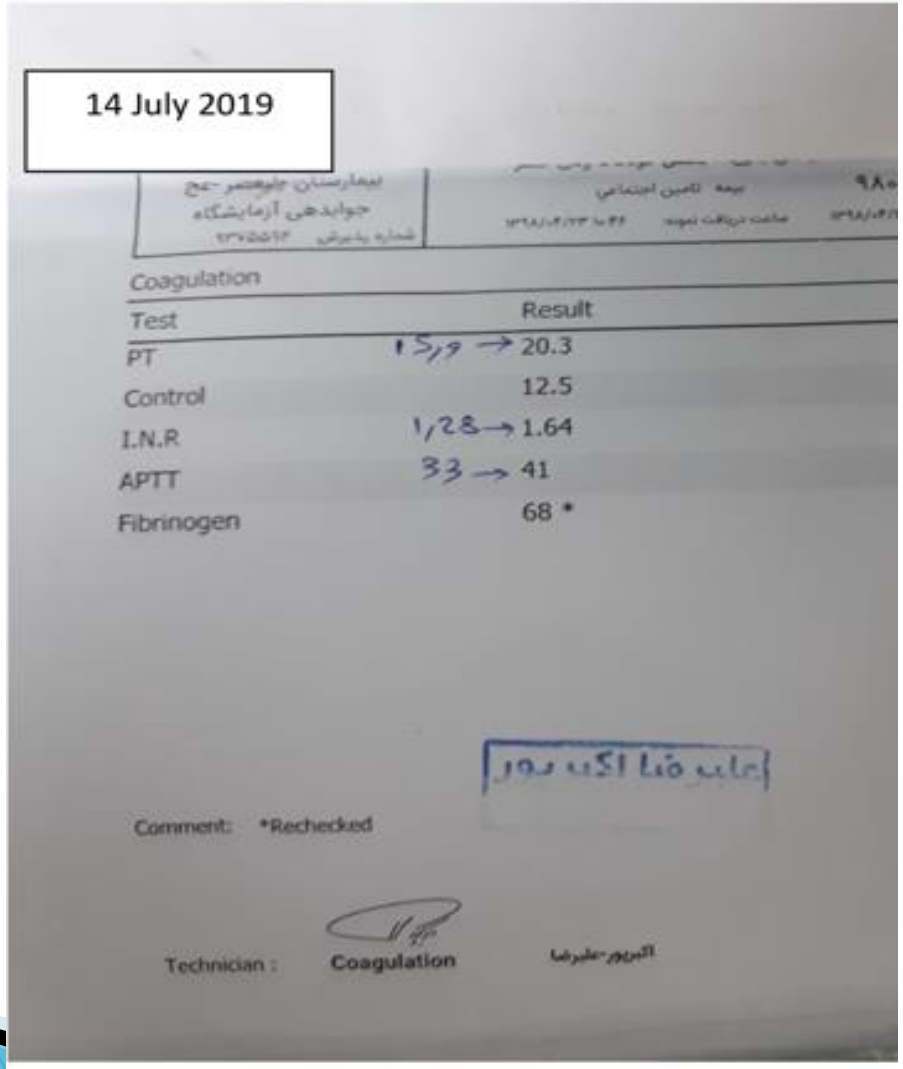
جواب فبا اکتبه بود

Comment: *Rechecked

Technician : Coagulation

اکبرپور-علیرضا

14 July 2019



بیمارستان جلوهگر -عج
جوابدهی آزمایشگاه
شماره پذیرش ۲۳۰۵۵۶۴

بیمه تامین اجتماعی
مدت دریافت نمونه ۳۹۸/۰۴/۲۳ تا ۴۱

۹۸۰
۳۹۸/۰۴/۲۱

Coagulation

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PT	15,9 → 20.3
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APTT	33 → 41
Fibrinogen	68 *

جواب فبا اکتبه بود

Comment: *Rechecked

Technician : Coagulation

اکبرپور-علیرضا

- Treatment with
- pulse-methylprednisolone
- IVIG

- *the chemotherapy according to BFM-NHL protocol for the patient began.*

- *and fortunately improved after two weeks of clinical and laboratory symptoms and continued chemotherapy regimen. Following treatment, fever was discontinued and facial swelling declined markedly and continued with outpatient chemotherapy was discharged.*

- *And in the fallow up, the patient was still afebrile and the swelling and ulcerative facial lesions improved completely.*


op dream

Panniculitis is the inflammation of the subcutaneous fat.

- *In patients with suspected panniculitis, a deep biopsy to include the subcutaneous fat is fundamental.*

Histopathologically, panniculitis can predominantly affect lobules or septa, with/without
vascular involvement.

- *the panniculitis can be classified into following main classes:*
- *A . predominantly septal panniculitis with or without vasculitis,*
- *B . predominantly lobular panniculitis with or without vasculitis and others*

- *. A patchy lymphoplasmacytic infiltrate in the lobules of the subcutaneous fat along with lymphocytic nuclear dust is a clue for the histopathological diagnosis of LE panniculitis (LEP) .*

- *Clinically, LEP and subcutaneous panniculitis-like T-cell lymphoma (SPTCL) are indistinguishable*

- *in SPTCL, the presence of atypical CD8-positive T-lymphocytes and the absence of septal fibrosis, B-cell follicles, and plasma cells clinch the diagnosis.*

SPTCL

- *SPTCL is an uncommon peripheral T-cell lymphoma accounting for less than 1 percent of all non-Hodgkin lymphoma subtypes. The exact incidence and variation in incidence with geography have not been well characterized.*

- *Patients present at a median age of 36 years (range 9 to 79 years) with approximately 20 percent of patients presenting under the age of 20 . There appears to be a female predominance with a male:female ratio of 0.5.*

- *Up to 20 percent of patients have at presentation an associated autoimmune disease such as systemic lupus erythematosus, juvenile idiopathic arthritis, Sjögren's syndrome, or type 1 diabetes mellitus.*

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Universitätsklinikum Schleswig-Holstein
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Kopie

Kiel, den 22.07.2019

Eingangsdatum 16.07.2019
Ausgangsdatum 22.07.2019
wk/grl

Name **Baran**
Vorname **Ghadiri**
Geb.-Datum **24.06.2008**

Journal-Nr. **R/2019/084385**

Farid Kosari MD AP-CP
Associate Professor of Path.
Atie Hospital Laboratory
Shahrake Gharb
Tehran, Iran

Ihre Auftragsnummer 1201
nachrichtlich
Befundauskunft 0431 / 500 15715

Pathologisch-anatomischer Begutachtungsbericht

Dear Farid,

thank you for sending 3 paraffin blocks, macroscopic images and some immunohistochemical stainings as images by E-Mail (your no. 1201/1-3). This young girl suffers from a periorbital edema and skin rash over the face for a couple of months. Despite steroid therapy the findings are getting worse. All serological tests for Lupus have been negative.

The tissue biopsies show abundant necrosis in the fatty tissue. There is a lymphatic infiltration in the fatty tissue of medium sized cells which arrange closely around fatty cells. The cell proliferation is moderate (Ki-67). The cells stain positive for CD3, CD8, Beta-chain of the T-cell receptor (Beta-F1) bunt remain negative for CD30, CD79a and TdT. CD79a is labeling a few plasma cells in this lesion.

This is a difficult case. Generally, I think that a subcutaneous panniculitis-like T-cell lymphoma is the most likeliest diagnosis. The morphology and the proliferation of the T-cells as well as the abundant expression of CD8 argue against a Lupus panniculitis. Furthermore, the B-cell content in this biopsies is relatively low. However, we would expect more B-cells and a more polymorphic infiltrate in Lupus. Nevertheless, we can detect some plasma cells in the CD79a-staining. Therefore, I have tried to confirm by PCR-analysis. We have extracted DNA from Block 3 and performed the BIOMED-2- primers.

T-cell receptor beta chain gene:

A-Multiplex-PCR: Polyklonal
B-Multiplex-PCR: Polyklonal
C-Multiplex-PCR: Polyklonal

T-cell receptor gamma chain gene:

va primer mix: Monoclonal/polyclonal background (164 and 174 bp)
vb primer mix: Irregular polyclonal

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Pathologisch-anatomischer Begutachtungsbericht

I think that all findings are compatible with a subcutaneous panniculitis-like T-cell lymphoma (ICD-O: 9708/3).

With kind regards
Sincerely Yours

Prof. Dr. W. Klapper

Tumorklassifikation
ICD-10: C86.3 unbekannt G
WHO: C44.0 M9708/3 pTX NX