9ο
Περιφερειακό Συνέδριο
Ελληνικής Εταιρείας
Αιμαφαίρεσης

ΠΛΑΣΜΑΦΑΙΡΕΣΗ ΣΕ ΑΣΘΕΝΗ ΜΕ ΘΡΟΜΒΩΤΙΚΗ ΜΙΚΡΟΑΓΓΕΙΟΠΑΘΕΙΑ ΜΕ ΤΕΛΙΚΗ ΔΙΑΓΝΩΣΗ ΚΑΚΟΗΘΕΙΑ. ΠΟΣΟ ΣΥΧΝΑ ΒΡΙΚΟΜΑΣΤΕ ΣΕ ΔΙΛΛΗΜΑ ?

ΠΑΝΑΓΑΚΟΥ ΣΤΕΛΛΑ

ΕΙΔΙΚΕΥΟΜΕΝΗ ΝΕΦΡΟΛΟΓΙΑΣ

ΓΝ ΠΑΠΑΓΕΩΡΓΙΟΥ

ΙΣΤΟΡΙΚΟ

- Θήλυ 68 ετών
- ΤΕΠ 3/2023

<u>Συμπτώματα εισόδου</u>:

✓ Αδυναμία/καταβολή από 10ημέρου
✓ Ήπιο οίδημα κάτω άκρων
✓ Δεκατική πυρετική κίνηση
✓ Θρομβοπενία και αναιμία

<u>Ατομικό αναμνηστικό</u>:

❖ Ρευματοειδής αρθρίτιδα
❖ Αρτηριακή υπέρταση
❖ Λοίμωξη με Covid19 2/2023
❖ Δεν αναφέρονται αλλεργίες

ΕΡΓΑΣΤΗΡΙΑΚΟΣ ΕΛΕΓΧΟΣ

	9/3/2023
WBC	5620
Ουδ/Λεμφ/Μον/Ηω/Βασ	58,9/32,6/6,8/0,8/1 %
PLTs	**21000**
Hb/Hct	**5,5/16,6**
MCV	133
INR/PT/aPTT	1,33/18,4/28,4
LDH	**421**
Cr/UREA	0,96/42
BIL	2,02
TKE	80

PLASMIC Score

Parameter	Result	Score
Platelet count	<30K	1
Creatinine	<2.0	1
INR	<1.5	1
MCV	<90	1
Presence of hemolysis variable	Either: -Retic>2.5% -Undetectable hapto-globin or -iBili>2 mg/dL	1
Abscence of active cancer		1
No prior stem cell or organ transplant		1

Σχιστικύτταρα 7%

- Plasmic score : 6 (high)
- Δείγμα για ενεργότητα ADAMTS 13

Εισαγωγή στην Αιματολογική Κλινική

GUIDELINES ON THE USE OF THERAPEUTIC APHERESIS IN CLINICAL PRACTICE – EVIDENCE-BASED APPROACH FROM THE WRITING COMMITTEE OF THE AMERICAN SOCIETY FOR APHERESIS: THE NINTH SPECIAL ISSUE

TABLE 1 (Continued)

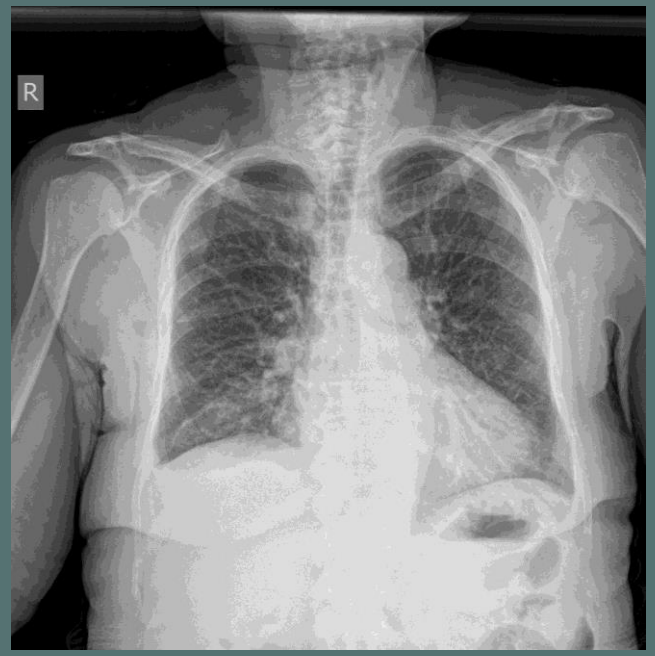
Disease/condition	Indication	Procedure	Category	Grade	Page
		TPE	III	2C	
Thrombocytosis	Symptomatic	Thrombocytapheresis	II	2C	229
	Prophylactic or secondary	Thrombocytapheresis	III	2C	
Thrombotic microangiopathy, coagulation mediated	*THBD*, *DGKE*, and *PLG* mutations	TPE	III	2C	231
Thrombotic microangiopathy, complement mediated	Factor H autoantibody	TPE	I	2C	233
	Complement factor gene mutations	TPE	III	2C	
Thrombotic microangiopathy, drug induced	Ticlopidine	TPE	I	2B	235
	Clopidogrel	TPE	III	2B	
	Gemcitabine	TPE	IV	2C	
	Quinine	TPE	IV	2C	
Thrombotic microangiopathy, infection associated	STEC-HUS, severe	TPE/IA	III	2C	237
	pHUS	TPE	III	2C	
Thrombotic microangiopathy, pregnancy associated	Pregnancy associated, severe	TPE	III	2C	239
	Extremely preterm preeclampsia, severe[a]	TPE/LA	III	2C	
Thrombotic microangiopathy, thrombotic thrombocytopenic purpura		TPE	I	1A	241
Thrombotic microangiopathy, transplantation associated		TPE	III	2C	243
Thyroid storm		TPE	II	2C	245
Toxic epidermal necrolysis	Refractory	TPE	III	2B	247
Transplantation, heart	Cellular rejection	ECP	II	1B	249
	Recurrent rejection	ECP	II	1B	
	Rejection prophylaxis	ECP	II	2A	
	Desensitization	TPE	II	1C	
	Rejection prophylaxis[a]	TPE	II	1C	
	Antibody mediated rejection	TPE	III	2C	

ΘΕΡΑΠΕΙΑ ΠΛΑΣΜΑΦΑΙΡΕΣΗΣ

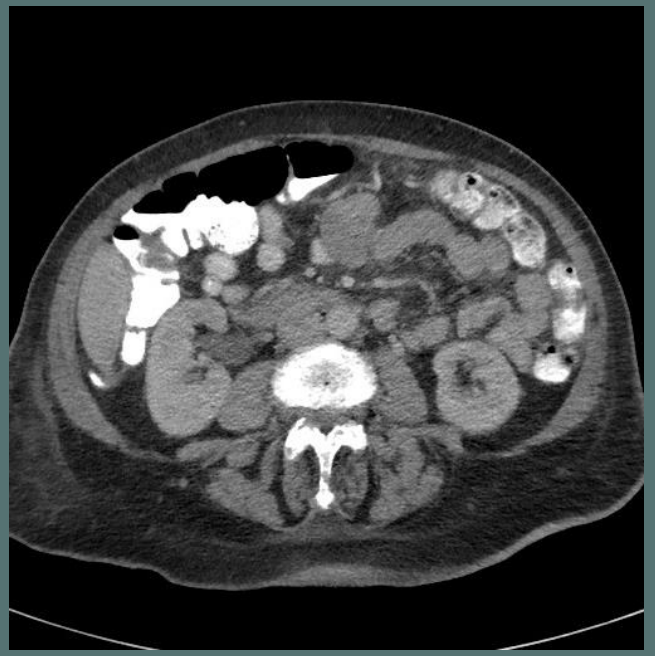
- 8 συνεδρίες PEX με φυγοκέντρηση
- FFPs ως υγρό υποκατάστασης
- Όγκος ανταλλαγής πλάσματος : 4lt
- Χορηγήθηκαν 10mg καπλασιζουμάμπης από αιματολόγους

Επιπλοκές:

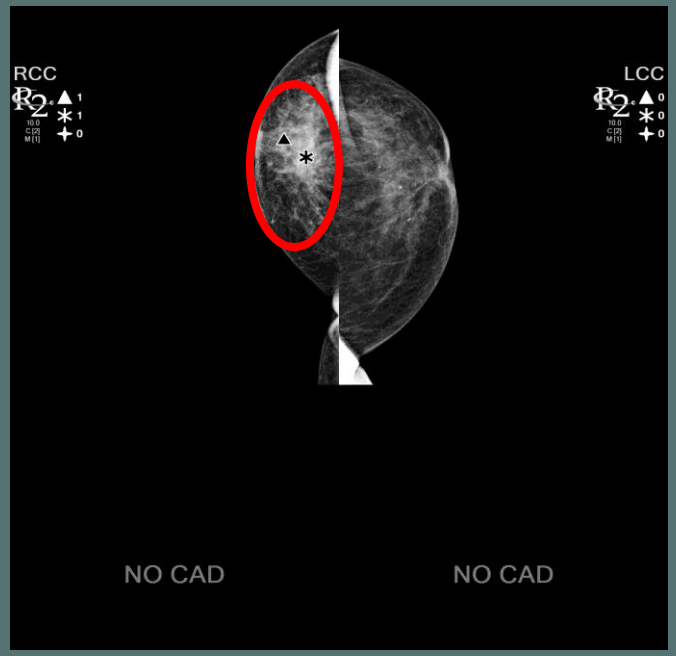
- Αιμορραγία από σημείο εισόδου ΚΦΚ
- Υπασβεστιαιμία



Ro θώρακος: πλευριτική συλλογή ΔΕ



CT ΑΚΚΟ: πάχυνση τοιχώματος θόλου στομάχου



Μαστογραφία: μάζα αυξημένης ακτινοσκιερότητας ΔΕ-Birards 5

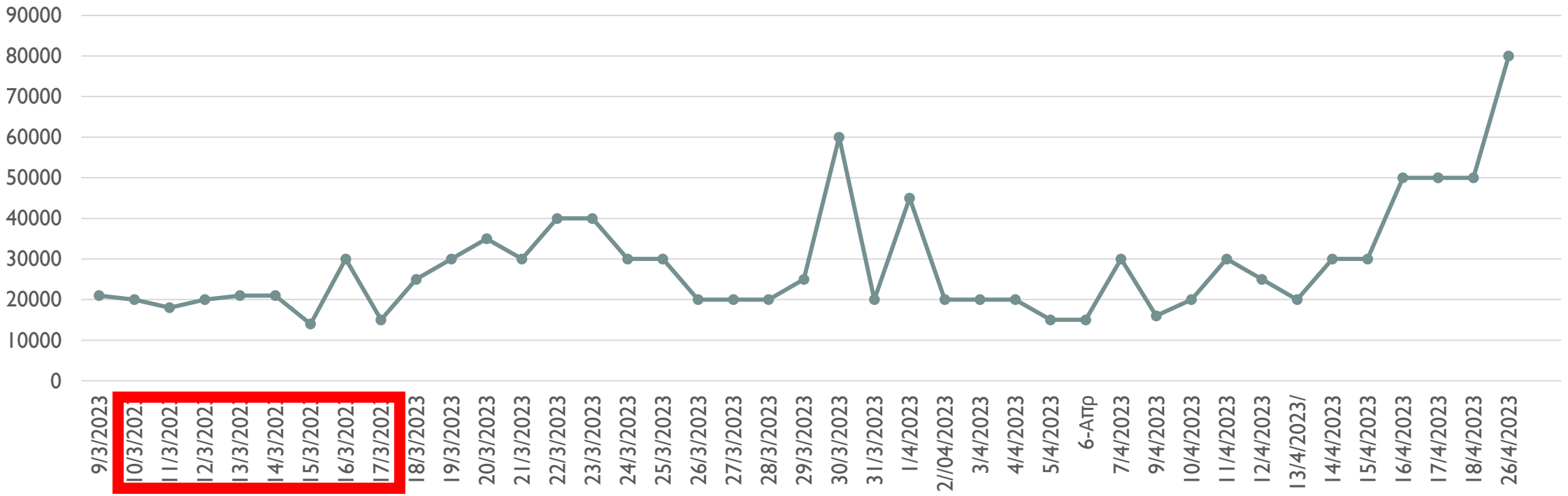
ΑΠΕΙΚΟΝΙΣΤΙΚΟΣ ΕΛΕΓΧΟΣ

ΛΟΙΠΟΣ ΕΛΕΓΧΟΣ

- Triplex **καρδιάς**: Grade II διαστολική δυσλειτουργία-EF>65%
- **Οστεομυελική βιοψία** και **μυελόγραμμα**: Διήθηση μυελου από μεταστατικό καρκίνωμα μέτριας διαφοροποίησης
- Core biopsy **Μαστού:** Μεταστατικό λοβιακό Ca μαστού grade2, ER/PR+, HER2(2+),ki-67 5%

➢ <u>ADAMTS 13: **εφο**</u>

ΑΙΜΟΠΕΤΑΛΙΑ



ΘΕΡΑΠΕΙΑ

Ενδομετακίνηση στην Ογκολογική κλινική

Χορήγηση πακλιταξέλης 140mg εβδομαδιαίως

TMA–malignancy associated

'The development of TMA in patients with underlying malignancies has been widely described.[30,31] This form may represent TMA due to drugs used to treat the underlying malignancy, such as gemcitabine or mitomycin C (as described earlier), or may be a direct consequence of the malignancy. **Criteria for the diagnosis of TMA–malignancy associated include the following: cancer diagnosis, direct antiglobulin test result negative for microangiopathic hemolytic anemia, thrombocytopenia, decreased serum haptoglobin level, and indirect hyperbilirubinemia**.

In a study by Elliot et al,[30] TMA–malignancy associated patients exhibited, in addition to the laboratory parameters provided earlier, elevations in D-dimers in the absence of other markers of disseminated intravascular coagulation, a median serum creatinine level of 1.2 mg/dL, and ADAMTS13 >10%. Patients also exhibited bone pain, respiratory symptoms, anorexia, and weight loss, symptoms not see in TTP or other TMAs. Patients were also older and had a longer history of symptoms than patients with either TTP or other forms of TMA.[32] **The most commonly associated malignancies include those of the stomach, breast, prostate, and lung.** The pathophysiology of TMA–malignancy associated is unclear. As mentioned, it may represent TMA–drug associated or may also be due to expression of tissue factor in widespread tumor.[33]

Evidence supporting efficacy of TPE in this setting is lacking, and the use of TPE may result in a delay in treating the underlying malignancy.[30] ASFA has not categorized the use of TPE in the treatment of this form of TMA.[2]'

THERAPEUTIC APHERESIS AS AN IMMUNOMODULATORY TOOL| DECEMBER 8, 2017

Plasma exchange in thrombotic microangiopathies (TMAs) other than thrombotic thrombocytopenic purpura (TTP)

Jeffrey L. Winters, *Hematology Am Soc Hematol Educ Program* (2017) 2017 (1): 632–638.

Cancer-Related TTP and Considerations of Plasma Exchange

- '…So, without an immediate ADAMTS13 level and concomitant inhibitor assay, in those hours while the diagnosis of TTP is being distinguished from other imitators, including infection and malignancy-associated MA,[7] an immediate initiation of potentially course-altering plasma exchange should be pursued, with the first option of peripheral access for the initial two or three procedures, to gain potentially valuable therapeutic benefits while minimizing the risks of central access…'

Cancer-Related TTP and Considerations of Plasma Exchange

Sep 13, 2011 Dennis A. Gastineau, MD, FACP

Publication Article, **Oncology** ONCOLOGY Vol 25 No 10 Volume **25**, Issue **10**

Cancer-related TTP: Role of plasma exchange

'...TMA associated with disseminated malignancy remains a **challenging and underdiagnosed condition** with very poor prognosis. Plasma exchange has no clinical use is potentially even life-threatening especially when it delays the administration of the appropriate chemotherapy which is the ultimate treatment for the underlying malignancy that resulted in TMA.

We propose that the following **criteria** be used for diagnostic consideration of disseminated malignancy-associated TMA: **evidence of hemolysis; leukoerythroblastic picture on the peripheral blood; coagulopathy consisting of elevated d-dimer, prolonged PT with normal aPTT; extreme elevations in the LDH levels (>1,000 IU/L); and presence of schistocytes in the peripheral blood smear**.

Such patients should be immediately considered for a bone marrow biopsy as chemotherapy should be initiated as soon as possible…'

Cancer-related TTP: Role of plasma exchange

M. H. Farhat , B. de Souza , A. Hanbali

DOI: 10.1200/jco.2009.27.15_suppl.e13527 *Journal of Clinical Oncology* 27, no. 15_suppl


ΕΥΧΑΡΙΣΤΩ ΓΙΑ ΤΗΝ ΠΡΟΣΟΧΗ ΣΑΣ!