ΔΩΜΑ

Διαφοροδιάγνωση περιστατικού

Νεφρολογική κλινική

Ε΄παθολογική κλινική

ΓΝΑ « Ο Ευαγγελισμός»

Ασθενής 67 ετών με παρατεινόμενο εμπύρετο απο 4 μήνου χωρίς άλλα συμπτώματα και :

- Λιγοστά παθολογικά ευρήματα απο την κλινική εξέταση (συστολικό φύσημα κορυφής,μη μουσικοί ρόγχοι άμφω και μείωση αψ σύστοιχα)

- Αρνητικό εξωτερικό απεικονιστικό έλεγχο (CT θώρακος με εμφυσηματικές αλλοιώσεις , U/S κοιλίας με αύξηση της ηχογένειας του φλοιού των νεφρών)

- Σε νέο απεικονιστικό έλεγχο με U/S κοιλίας παρουσία ασκιτικής συλλογής και πλευτικών συλλογών

- Εργαστηριακός έλεγχος

- Αναιμία ορθόχρωμη (Hb 7.7 g/dl), θρομβοπενία (20.000/μL), σχιστοκύτταρα (4 ΚΟΠ) απουσία σφαιροκυττάρων, αυξημένα ΔΕΚ και LDH, ευρήματα συμβατά με MAHA

- ΟΝΑ (Ur 171 mg/dl, Cr 3.9 mg/dl) με άφθονα ερυθρά (χωρίς παρουσία κυλίνδρων ή δύσμορφων RBCs) και παρουσία λευκώματος και αιμοσφαιρίνης στην γενική ούρων.

- Χαμηλή CRP-ΤΚΕ, ήπια παράταση aPTT και χαμηλό ινωδογόνο και χαμηλό συμπλήρωμα, RF(+) και ANA 1:640 με ANCA(-)

Συνοψίζοντας

- Παρατεινόμενο εμπύρετο
- ΜΑΗΑ
- ΟΝΑ

Αίτια παρατεινόμενου εμπυρέτου

TABLE 288-7 FREQUENCY OF SELECTED CHRONIC FEBRILE ILLNESSES

INFECTION, 25-50%	MALIGNANCY, 20-30%	CONNECTIVE TISSUE DISEASE, 15-30%	MISCELLANEOUS, 10-20%	UNDIAGNOSED, 10-30%
Cytomegalovirus	Carcinomatosis	Polyarteritis nodosa	Drug-induced fever	
Endocarditis	Leukemia	Rheumatoid arthritis	Granulomatous hepatitis	
Intra-abdominal	Local tumor	Still's disease	Inflammatory bowel disease	
Mycoses	Lymphoma	Systemic lupus erythematosus	Pancreatitis	
Occult abscess		Temporal arteritis	Pulmonary embolism	
Tuberculosis				

Αίτια ΟΝΑ

Prerenal ARF

I. Hypovolemia
- A. Increased extracellular fluid losses: hemorrhage
- B. Gastrointestinal fluid loss: vomiting, diarrhea, enterocutaneous fistula
- C. Renal fluid loss: diuretics, osmotic diuresis, hypoadrenalism, nephrogenic diabetes insipidus
- D. Extravascular sequestration: burns, pancreatitis, severe hypoalbuminemia (hypoproteinemia)
- E. Decreased intake: dehydration, altered mental status

II. Altered renal hemodynamics resulting in hypoperfusion
- A. Low cardiac output state: diseases of the myocardium, valves, and pericardium (including tamponade); pulmonary hypertension or massive pulmonary embolism leading to right and left heart failure; impaired venous return (e.g., abdominal compartment syndrome or positive pressure ventilation)
- B. Systemic vasodilation: sepsis, antihypertensives, afterload reducers, anaphylaxis
- C. Renal vasoconstriction: hypercalcemia, catecholamines, calcineurin inhibitors, amphotericin B
- D. Impairment of renal autoregulatory responses: cyclooxygenase inhibitors (e.g., nonsteroidal anti-inflammatory drugs), angiotensin-converting enzyme inhibitors, or angiotensin II receptor blockers
- E. Hepatorenal syndrome

Intrinsic ARF

I. Renovascular obstruction (bilateral, or unilateral in the setting of one kidney)
- A. Renal artery obstruction: atherosclerotic plaque, thrombosis, embolism, dissection aneurysm, large vessel vasculitis
- B. Renal vein obstruction: thrombosis or compression

II. Diseases of the glomeruli or vasculature
- A. Glomerulonephritis or vasculitis
- B. Other: thrombotic microangiopathy, malignant hypertension, collagen vascular diseases (systemic lupus erythematosus, scleroderma), disseminated intravascular coagulation, preeclampsia

III. Acute tubular necrosis
- A. Ischemia: causes are the same as for prerenal ARF, but generally the insult is more severe and/or more prolonged
- B. Infection, with or without sepsis syndrome
- C. Toxins:
  1. Exogenous: radiocontrast, calcineurin inhibitors, antibiotics (e.g., aminoglycosides), chemotherapy (e.g., cisplatin), antifungals (e.g., amphotericin B), ethylene glycol
  2. Endogenous: rhabdomyolysis, hemolysis

IV. Interstitial nephritis
- A. Allergic: antibiotics (β-lactams, sulfonamides, quinolones, rifampin), nonsteroidal anti-inflammatory drugs, diuretics, other drugs
- B. Infection: pyelonephritis (if bilateral)
- C. Infiltration: lymphoma, leukemia, sarcoidosis
- D. Inflammatory, nonvascular: Sjögren's syndrome, tubulointerstitial nephritis with uveitis

V. Intratubular obstruction
- A. Endogenous: myeloma proteins, uric acid (tumor lysis syndrome), systemic oxalalosis
- B. Exogenous: acyclovir, gancyclovir, methotrexate, indinavir

Postrenal ARF (Obstruction)

I. Ureteric (bilateral, or unilateral in the case of one kidney): calculi, blood clots, sloughed papillae, cancer, external compression (e.g., retroperitoneal fibrosis)

II. Bladder neck: neurogenic bladder, prostatic hypertrophy, calculi, blood clots, cancer

III. Urethra: stricture or congenital valves

Αίτια ΜΑΗΑ

- TTP/HUS
- Φάρμακα
- Κακοήθειες
- ΔΕΠ
- Αγγειίτιδες
- Κακοήθης υπέρταση
- Σκληρόδερμα
- ΣΕΛ
- APS/cAPS

Φαρμακα

Φάρμακα συσχετιζόμενα με ΜΑΗΑ

1) Quinine

2) Χημειοθεραπευτικά (Cisplatin,Bevacizumab)

3) ανοσοκατασταλτικοί παράγοντες (cyclosporine,tacrolimus)

- Ο ασθενής ωστόσο δεν ελάμβανε καμιά φαρμακευτική αγωγή

ΔΕΠ

- Παράταση aPTT χαμηλό ινωδογόνο και πυρετό.

- Το καλό επίπεδο επικοινωνίας, το γεγονός ότι ήταν αιμοδυναμικά σταθερός με καλό γαλακτικο και η απουσία πορφυρικού εξανθήματος, αιμορραγικων εκδηλώσεων και πολυοργανικής συμμετοχής μαλλον μας απομακρύνουν απο acute DIC.

➢ Χρόνια ΔΕΠ ??

TABLE 178-1 MAJOR CAUSES OF DISSEMINATED INTRAVASCULAR COAGULATION

INFECTIONS
Gram-negative bacterial sepsis
Other bacteria, fungi, viruses, Rocky Mountain spotted fever, malaria
IMMUNOLOGIC REACTIONS
Transfusion reactions (ABO incompatibility)
Transplant rejection
OBSTETRIC COMPLICATIONS
Amniotic fluid embolism
Retained dead fetus
Abruptio placentae
Toxemia, preeclampsia
Septic abortion
MALIGNANCIES
Pancreatic carcinoma
Adenocarcinomas
Acute promyelocytic leukemia
Other neoplasms
LIVER FAILURE
ACUTE PANCREATITIS
ENVENOMATION
RESPIRATORY DISTRESS SYNDROME
TRAUMA, SHOCK
Brain injury
Crush injury
Burns
Hypothermia/hyperthermia
Fat embolism
Hypoxia, ischemia
Surgery
VASCULAR DISORDERS
Giant hemangioma (Kasabach-Merritt syndrome)
Aortic aneurysm
Vascular tumors

Κακοήθειες

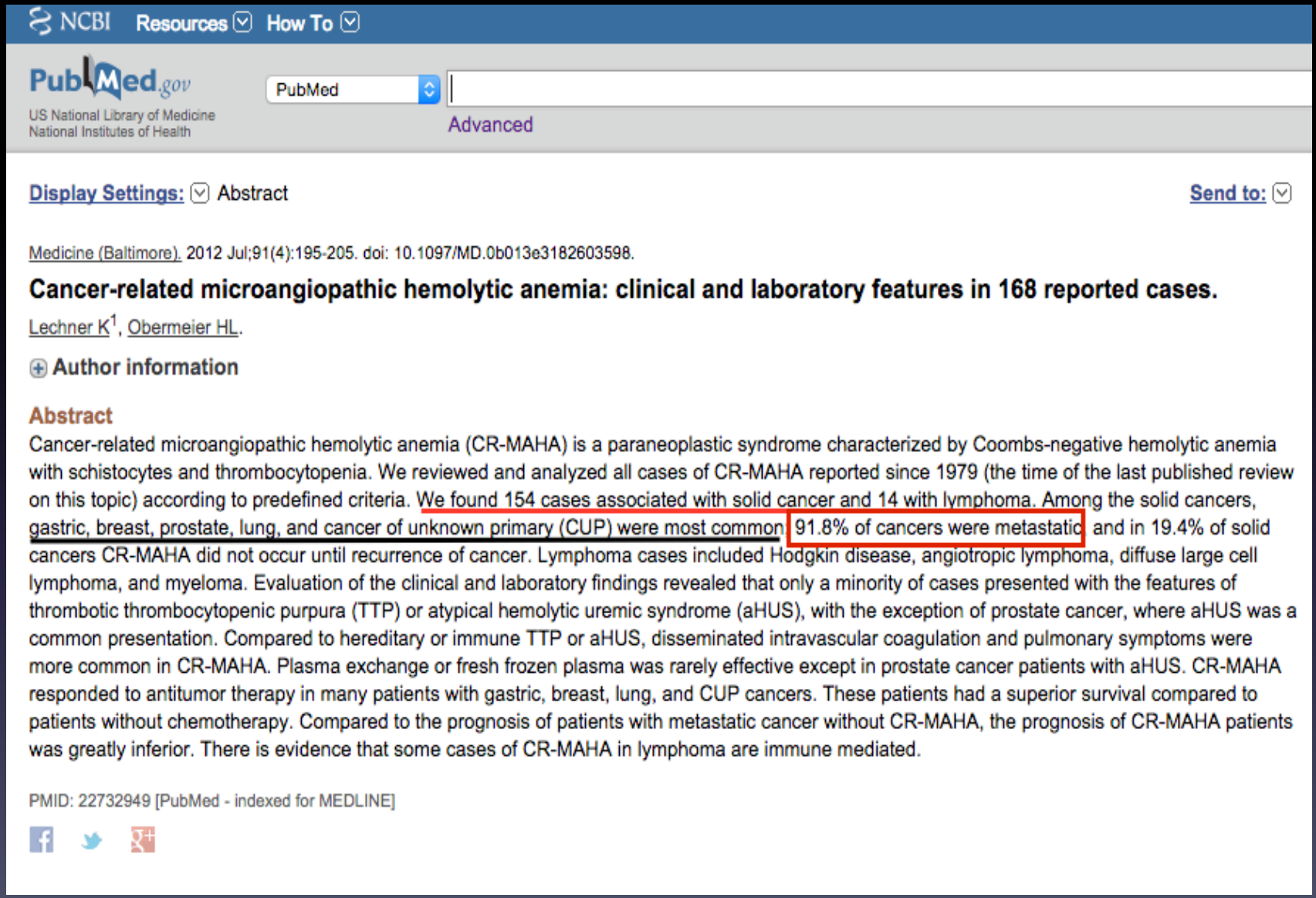
Table 1 Systemic Malignancies Associated With Microangiopathic Hemolytic Anemia and Thrombocytopenia

	Data Sources						
Malignancy	Brain 1962[2] (N = 5)	Antman 1979[5] (N = 55)	Oklahoma 2007[6] (N = 10)	Multiple Reports From the Literature[6] (N = 19)	Oberic 2009[9] (N = 20)	Elliott 2010[10] (N = 7)	Total (N = 65)
Gastric carcinoma	3	30		5	4	1	43
Breast carcinoma		7	2	1	7	4	21
Lung carcinoma	1	4	2	1	3	1	12
Prostate carcinoma	1	2		4	2		9
Carcinoma of unknown origin		5		3	1		9
Colon carcinoma		1		1	1		3
Hepatoma		1			2		3
Pancreatic carcinoma		2	1				3
Anal squamous cell carcinoma				2			2
Non-Hodgkin lymphoma			1	1			2
Acute lymphocytic leukemia			1				1
Cholangiocarcinoma		1					1
Kaposi sarcoma			1				1
Multiple endocrine neoplasia type 1				1			1
Myelodysplastic syndrome			1				1
Neuroendocrine tumor						1	1
Ovarian carcinoma		1					1
Renal carcinoma			1				1
Seminal vesicle tumor		1					1

In the patients reported from the Oklahoma TTP-HUS Registry[6] and the Mayo Clinic,[10] the systemic malignancies were not suspected when plasma exchange treatment for the diagnosis of TTP (thrombotic thrombocytopenic purpura) was begun. Many patients in the other reports had clinically overt and recognized malignancies when the microangiopathic hemolytic anemia and thrombocytopenia occurred. The data from Antman et al[5] include 4 patients whom they reported on and 51 previously reported cases. Patients from the Oklahoma literature review[6] are from a systematic review through January 2006 to identify all previously reported patients[1] in whom TTP or HUS (hemolytic-uremic syndrome) was initially suspected[2] and malignancy was not considered as an initial diagnosis.[3] The patients had not received chemotherapy, nor was there evidence of malignancy within the previous year,[4] and there was no evidence of disseminated intravascular coagulation. None of the patients identified in the Oklahoma[6] review were previously identified in the review by Antman et al.[5]

Oncology (Williston Park). 2011 sep;25(10):908-14

Κακοήθειες



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Medicine (Baltimore). 2012 Jul;91(4):195-205. doi: 10.1097/MD.0b013e3182603598.

Cancer-related microangiopathic hemolytic anemia: clinical and laboratory features in 168 reported cases.

Lechner K[1], Obermeier HL.

Author information

Abstract

Cancer-related microangiopathic hemolytic anemia (CR-MAHA) is a paraneoplastic syndrome characterized by Coombs-negative hemolytic anemia with schistocytes and thrombocytopenia. We reviewed and analyzed all cases of CR-MAHA reported since 1979 (the time of the last published review on this topic) according to predefined criteria. We found 154 cases associated with solid cancer and 14 with lymphoma. Among the solid cancers, gastric, breast, prostate, lung, and cancer of unknown primary (CUP) were most common. 91.8% of cancers were metastatic, and in 19.4% of solid cancers CR-MAHA did not occur until recurrence of cancer. Lymphoma cases included Hodgkin disease, angiotropic lymphoma, diffuse large cell lymphoma, and myeloma. Evaluation of the clinical and laboratory findings revealed that only a minority of cases presented with the features of thrombotic thrombocytopenic purpura (TTP) or atypical hemolytic uremic syndrome (aHUS), with the exception of prostate cancer, where aHUS was a common presentation. Compared to hereditary or immune TTP or aHUS, disseminated intravascular coagulation and pulmonary symptoms were more common in CR-MAHA. Plasma exchange or fresh frozen plasma was rarely effective except in prostate cancer patients with aHUS. CR-MAHA responded to antitumor therapy in many patients with gastric, breast, lung, and CUP cancers. These patients had a superior survival compared to patients without chemotherapy. Compared to the prognosis of patients with metastatic cancer without CR-MAHA, the prognosis of CR-MAHA patients was greatly inferior. There is evidence that some cases of CR-MAHA in lymphoma are immune mediated.

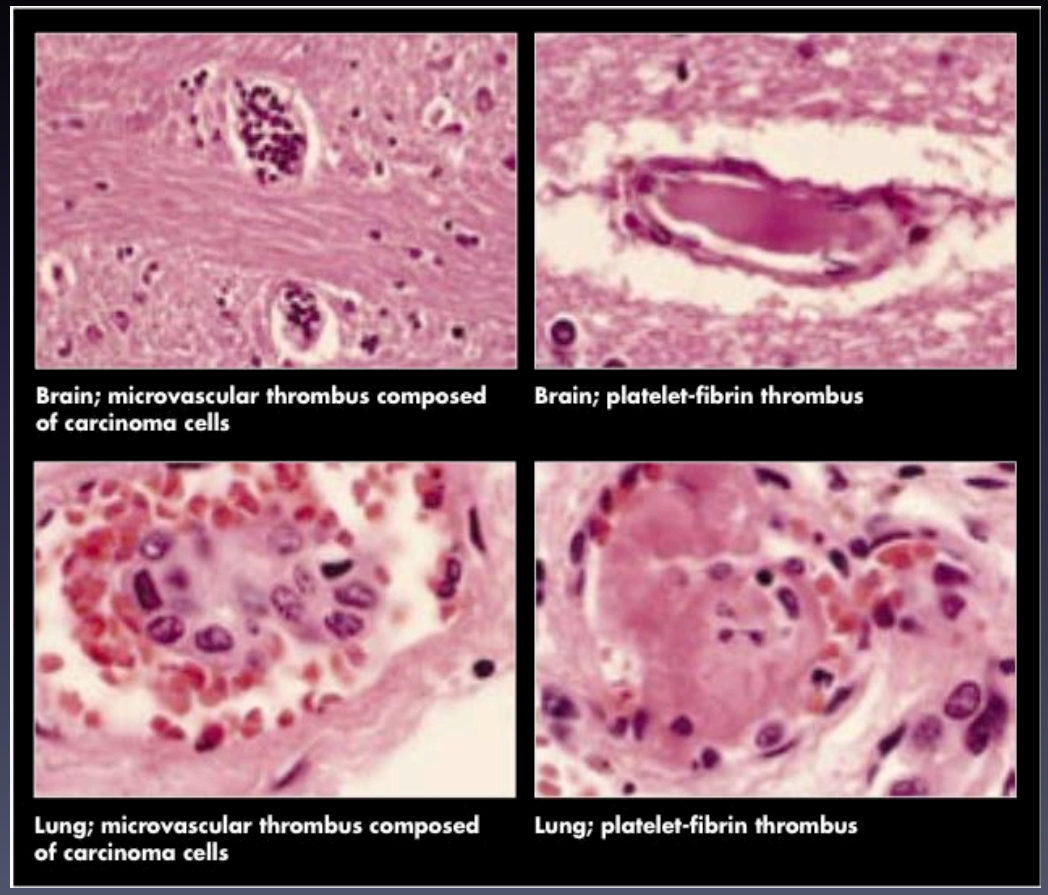
PMID: 22732949 [PubMed - indexed for MEDLINE]

Κακοήθειες

Προκαλούν ΜΑΗΑ με δυό πιθανούς μηχανισμούς

1) Chronic DIC

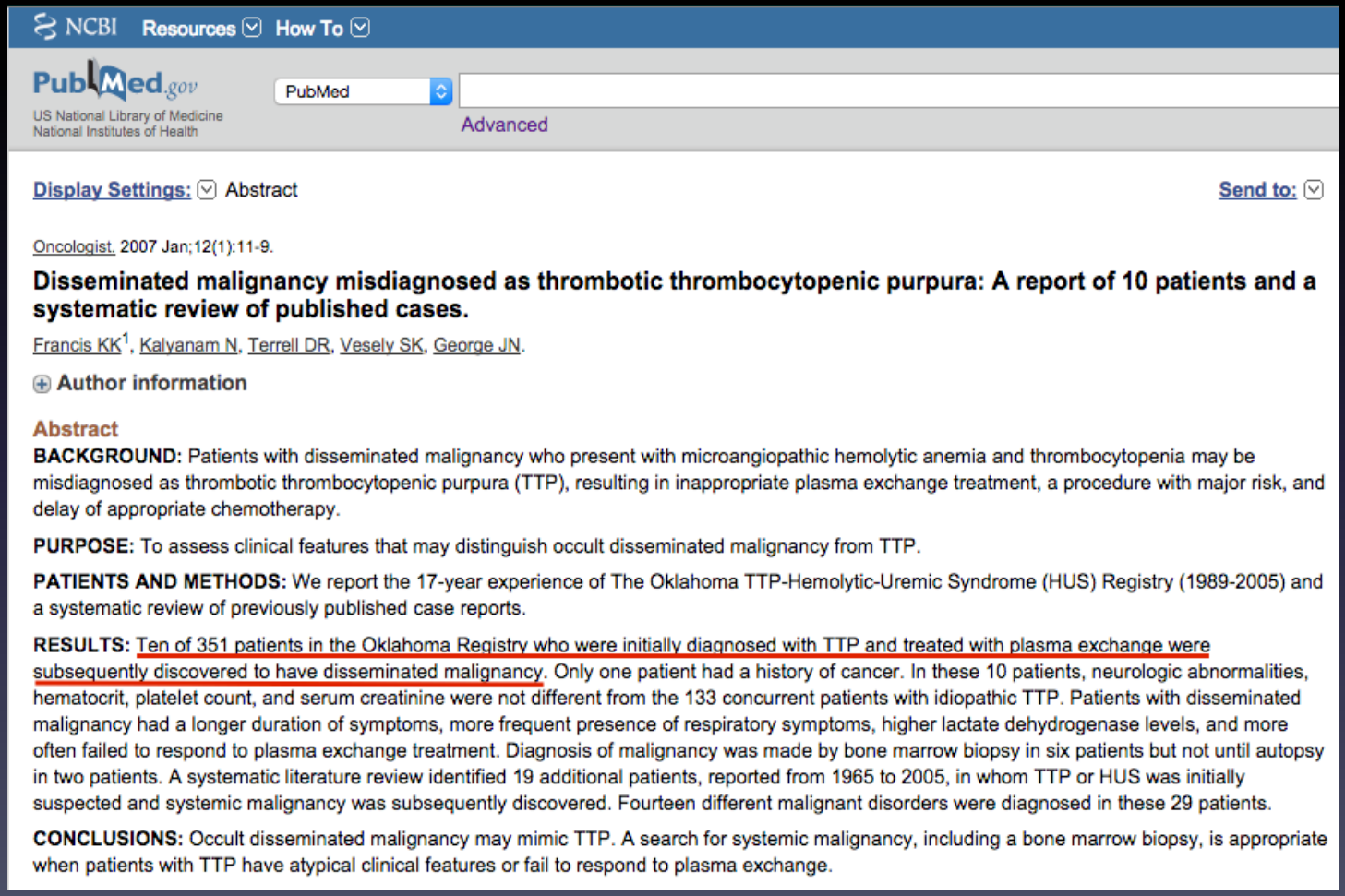
2) Μερική απόφραξη των αρτηριολίων απο tumor cell-emboli



Κακοήθειες

- Στην πλειονότητα των περιπτώσεων το νεόπλασμα είναι ήδη γνωστό
- Συνήθως είναι μεταστατικό
- Σπανιότερα η πρώτη εκδήλωση του νεοπλάσματος μπορεί να είναι αυτή της θρομβωτικής μικροαγγειοπάθειας

Κακοήθειες



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Oncologist. 2007 Jan;12(1):11-9.

Disseminated malignancy misdiagnosed as thrombotic thrombocytopenic purpura: A report of 10 patients and a systematic review of published cases.

Francis KK[1], Kalyanam N, Terrell DR, Vesely SK, George JN.

Author information

Abstract

BACKGROUND: Patients with disseminated malignancy who present with microangiopathic hemolytic anemia and thrombocytopenia may be misdiagnosed as thrombotic thrombocytopenic purpura (TTP), resulting in inappropriate plasma exchange treatment, a procedure with major risk, and delay of appropriate chemotherapy.

PURPOSE: To assess clinical features that may distinguish occult disseminated malignancy from TTP.

PATIENTS AND METHODS: We report the 17-year experience of The Oklahoma TTP-Hemolytic-Uremic Syndrome (HUS) Registry (1989-2005) and a systematic review of previously published case reports.

RESULTS: Ten of 351 patients in the Oklahoma Registry who were initially diagnosed with TTP and treated with plasma exchange were subsequently discovered to have disseminated malignancy. Only one patient had a history of cancer. In these 10 patients, neurologic abnormalities, hematocrit, platelet count, and serum creatinine were not different from the 133 concurrent patients with idiopathic TTP. Patients with disseminated malignancy had a longer duration of symptoms, more frequent presence of respiratory symptoms, higher lactate dehydrogenase levels, and more often failed to respond to plasma exchange treatment. Diagnosis of malignancy was made by bone marrow biopsy in six patients but not until autopsy in two patients. A systematic literature review identified 19 additional patients, reported from 1965 to 2005, in whom TTP or HUS was initially suspected and systemic malignancy was subsequently discovered. Fourteen different malignant disorders were diagnosed in these 29 patients.

CONCLUSIONS: Occult disseminated malignancy may mimic TTP. A search for systemic malignancy, including a bone marrow biopsy, is appropriate when patients with TTP have atypical clinical features or fail to respond to plasma exchange.

Κακοήθειες

Χαρακτηριστικά των ασθενών αυτών αποτελούν :

- Μεγάλη ηλικία
- Σταδιακή έναρξη των συμπτωμάτων
- Συμπτωματολογία απο αναπνευστικο
- Λευκοερυθροβλαστική αντίδραση
- Ιδιαίτερα αυξημένη LDH
- Διαταραχές πήξης
- Μη καλή ανταπόκριση στην πλασμαφαίρεση

Oncology (Williston Park). 2011:25(10):908

Κακοήθειες

Περαιτέρω έλεγχος με :

- Απεικόνιση κοιλίας-πυέλου με CT
- Ενδοσκόπηση πεπτικού
- Νεοπλασματικοί δείκτες
- Οστεομυελική βιοψία

Κακοήθης υπέρταση

Τιμές αρτηριακής πίεσης (BP >210/130mmHg) με συνοδές βλάβες σε όργανα στόχους (εγκέφαλος, νεφροί, καρδιά) και ευρήματα ΜΑΗΑ

- Ο ασθενής είχε φυσιολογικές τιμές αρτηριακής πίεσης και πυρετό

Σκληρόδερμα

Νεφρική κρίση σκληροδέρματος

- Οξεία ολιγουρική νεφρική ανεπάρκεια
- Παθολογικό ίζημα ούρων
- Αυξημένη τιμή αρτηριακής πίεσης (μετρια-επίπεδα κακοήθους υπέρτασης)
- ΜΑΗΑ
- Συχνότερη στο dcSSc

Σκληρόδερμα

- Αν και σπάνια, η πρώτη εκδήλωση του σκληροδέρματος μπορεί να είναι η νεφρική κρίση

- Ορισμένοι επίσης ασθενείς μπορει να παραμείνουν νορμοτασικοί η απλά να εμφανίζουν τιμές BP μεγαλύτερες του baseline.

- Το φύλο του ασθενή σε συνδυασμό με την μικρή πιθανότητα να εκδηλωθεί η νόσος πρώτα με νεφρική κρίση ταυτόχρονα με φυσιολογική πιέση μάλλον μας απομακρύνουν απο τη διάγνωση αυτή

Αγγειίτιδες

Συνήθως οι αιματολογικές επιπλοκές που παρατηρούνται στα πλαίσια αγγειίτιδας είναι:

Αναιμία χρονίας νόσου, θρομβοκυττάρωση

ή

Αναιμία/θρομβοπενια σχετιζόμενη με τα φάρμακα (κοτριμοξαζολη,κυτταροτοξικά,) ή επιπλοκές αυτών (σήψη)

- Ωστόσο έχουν περιγραφεί περιστατικά με ΜΑΗΑ

British Journal of Rheumatology. 1996;35:377-379

Αγγειίτιδες

Η απουσία πολυσυστηματικών εκδηλώσεων σε συνδυασμό με αρνητικά ANCA καθιστούν λιγότερη πιθανή της διάγνωση αυτή

- Βέβαια η ANCA(-) wegener , η οζώδης πολυαρτηρίτιδα και η σχετιζόμενη με κρυοσφαιριναιμία αγγειίτιδα δεν μπορούν να αποκλειστούν.

Αγγειίτιδες

Ελεγχος επι ενδείξεων με :

- κρυοσφαιρίνες (δεδομένου και του χαμηλού συμπληρώματος)
- Βιοψία δέρματος-μυός-αγγείου ή πάσχοντος οργάνου
- Αγγειογραφία

Αντιφωσφολιπιδικό σύνδρομο

APS/cAPS

- Σύνδρομο σχετιζόμενο με αρτηριακές-φλεβικές θρομβώσεις,υποτροπιάζουσες αποβολές το πρώτο τρίμηνο και παρουσία αντιφωσφολιπιδικών αντισωμάτων

- Μπορεί όμως να εμφανιστούν και αιματολογικές επιπλοκές όπως θρομβοπενία,AIHA, TTP/HUS, HELLP syndrome και καταστροφικό APS.

Blood Rev. 2008 Jul:22(4):187-94

Αντιφωσφολιπιδικό σύνδρομο

APS/cAPS

- Η μικροαγγειοπαθητική αιμολυτική αναιμία μπορεί να είναι και η πρώτη εκδήλωση του συνδρόμου αυτού

Ann Rheum Dis 2004;63:730–736

- Η παρουσία των apL εχει αναφερθεί και σε περιστατικά με TTP/HUS με ιδιαίτερη δυσκολία όσον αφορά την διάκριση τους και αρκετοί συγγραφείς έχουν αναγνωρίσει πιθανή συσχέτιση σε βαθμό μάλιστα που να προτείνουν τον όρο: Microangiopathic phospholipid syndrome

British Journal of Haematology, 149, 292–306

Ann Rheum Dis. 2007;66(4):429-432

Αντιφωσφολιπιδικό σύνδρομο

APS/cAPS

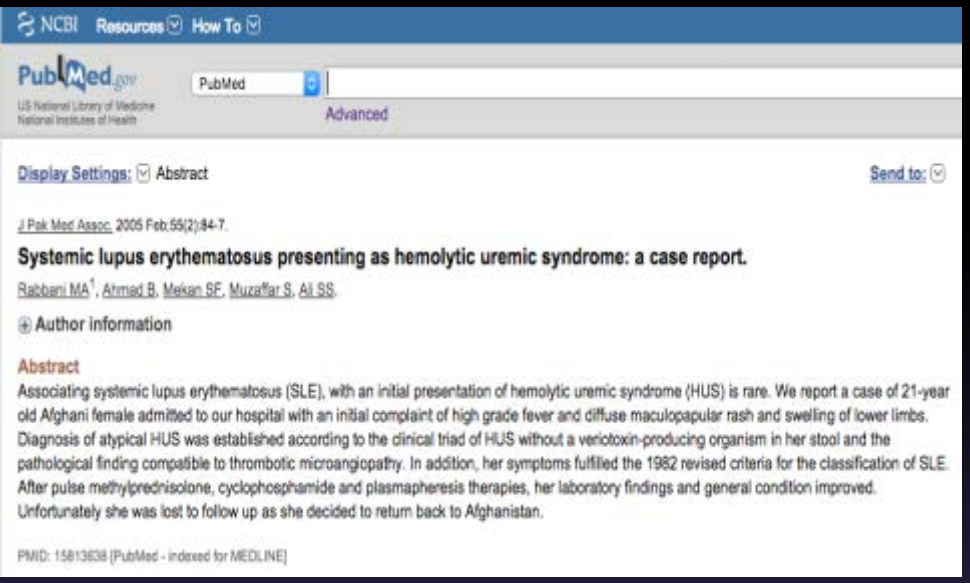
Περαιτέρω έλεγχος με :

- Αντιπηκτικό Λύκου
- Anti-β2GPI
- Αντισώματα έναντι καρδιολιπίνης
- VDRL
- Triplex φλεβών κάτω άκρων
- CTPA ?

ΣΕΛ

- Ο Λύκος ως αίτιο θρομβωτικής μικροαγγειοπάθειας (ΤΜΑ) εχει αναφερθεί σε αρκετά case-reports στην διεθνή βιβλιογραφία με πιθανέστερους μηχανισμούς : την αγγειίτιδα του Λύκου,το αντιφωσφολιπιδικό σύνδρομο (APS, cAPS) και TTP.

ΣΕΛ



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J Pak Med Assoc. 2005 Feb;55(2):84-7.

Systemic lupus erythematosus presenting as hemolytic uremic syndrome: a case report.

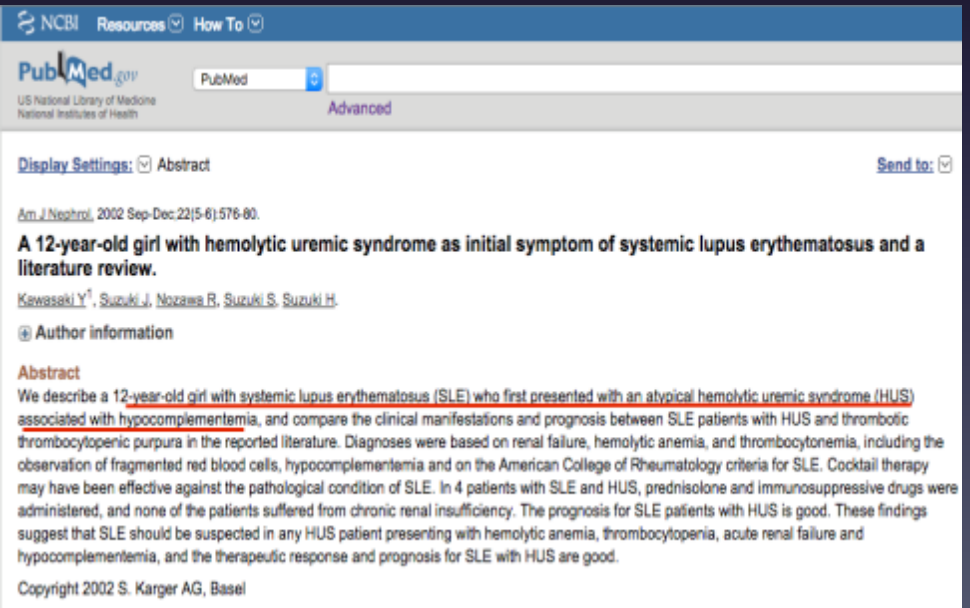
Rabbani MA[1], Ahmed B, Mekan SF, Muzaffar S, Ali SS.

Author information

Abstract

Associating systemic lupus erythematosus (SLE), with an initial presentation of hemolytic uremic syndrome (HUS) is rare. We report a case of 21-year old Afghani female admitted to our hospital with an initial complaint of high grade fever and diffuse maculopapular rash and swelling of lower limbs. Diagnosis of atypical HUS was established according to the clinical triad of HUS without a veriotoxin-producing organism in her stool and the pathological finding compatible to thrombotic microangiopathy. In addition, her symptoms fulfilled the 1982 revised criteria for the classification of SLE. After pulse methylprednisolone, cyclophosphamide and plasmapheresis therapies, her laboratory findings and general condition improved. Unfortunately she was lost to follow up as she decided to return back to Afghanistan.

PMID: 15813638 [PubMed - indexed for MEDLINE]



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Am J Nephrol. 2002 Sep-Dec;22(5-6):576-80.

A 12-year-old girl with hemolytic uremic syndrome as initial symptom of systemic lupus erythematosus and a literature review.

Kawasaki Y[1], Suzuki J, Nozawa R, Suzuki S, Suzuki H.

Author information

Abstract

We describe a 12-year-old girl with systemic lupus erythematosus (SLE) who first presented with an atypical hemolytic uremic syndrome (HUS) associated with hypocomplementemia, and compare the clinical manifestations and prognosis between SLE patients with HUS and thrombotic thrombocytopenic purpura in the reported literature. Diagnoses were based on renal failure, hemolytic anemia, and thrombocytonemia, including the observation of fragmented red blood cells, hypocomplementemia and on the American College of Rheumatology criteria for SLE. Cocktail therapy may have been effective against the pathological condition of SLE. In 4 patients with SLE and HUS, prednisolone and immunosuppressive drugs were administered, and none of the patients suffered from chronic renal insufficiency. The prognosis for SLE patients with HUS is good. These findings suggest that SLE should be suspected in any HUS patient presenting with hemolytic anemia, thrombocytopenia, acute renal failure and hypocomplementemia, and the therapeutic response and prognosis for SLE with HUS are good.

CASE STUDY

CME

Nature Clinical Practice Rheumatology (2007) **3**, 357-362
doi:10.1038/ncprheum0511
Received 26 June 2006 | Accepted 11 April 2007

Thrombotic microangiopathic hemolytic anemia in a patient with SLE: diagnostic difficulties

Ami A Shah, John P Higgins and Eliza F Chakravarty* About the authors

Correspondence *Division of Immunology and Rheumatology, Stanford University School of Medicine, 1000 Welch Road, Suite 203, Palo Alto, CA 94304, USA

Email echakravarty@stanford.edu

SUMMARY

Background A 19-year-old woman with newly diagnosed systemic lupus erythematosus (SLE) presented with hemolytic anemia, thrombocytopenia, hypertension, tonic-clonic seizures, blurry vision, nephrotic syndrome and renal insufficiency.

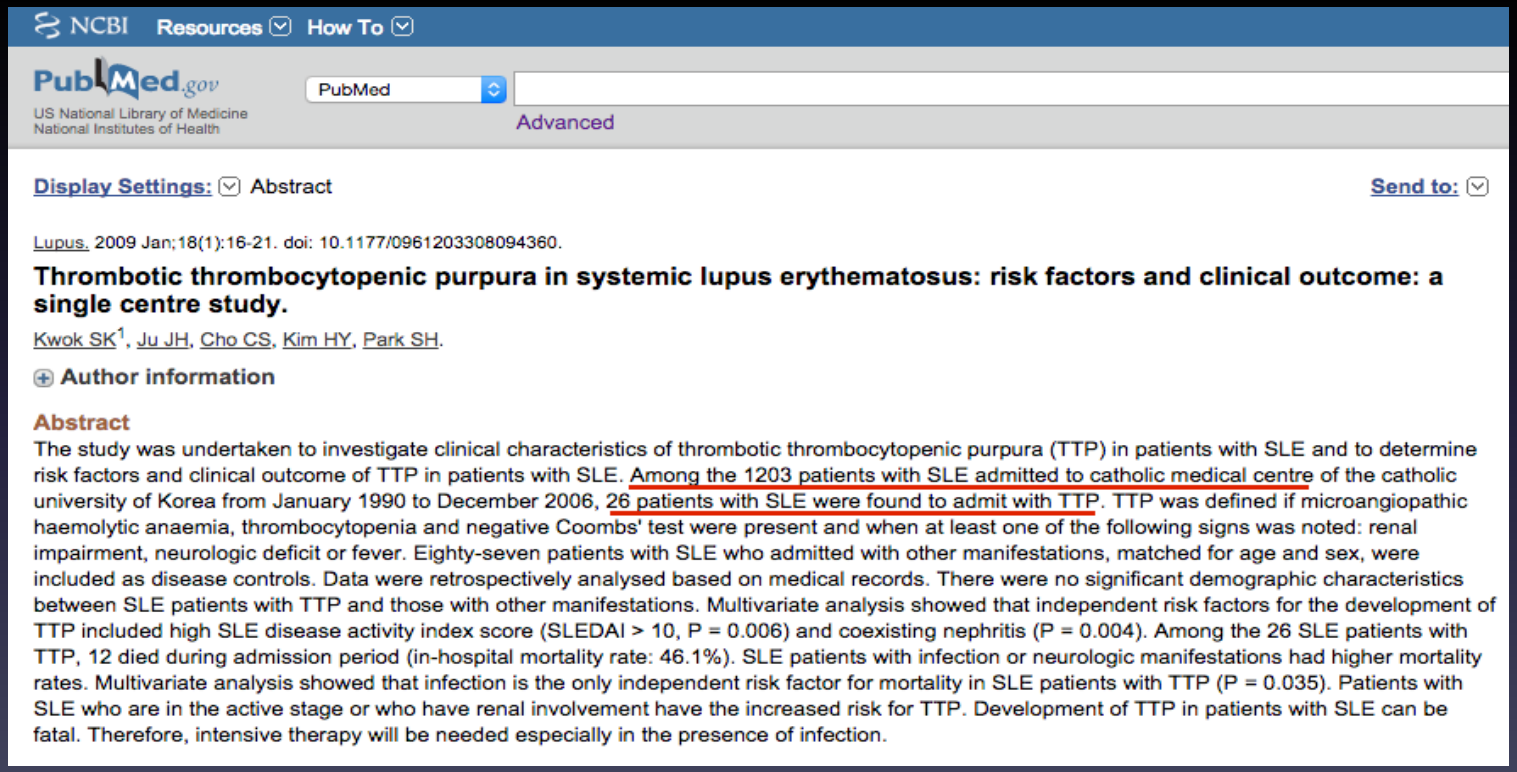
Investigations At a general hospital, the investigations included brain MRI, echocardiography, laboratory tests including measurement of the amount of protein excreted daily, platelet count, levels of lactate dehydrogenase, creatinine and anticardiolipin antibodies, direct Coombs' test, peripheral blood smear, and measurement of blood pressure. At a tertiary institution the investigations included physical examination, electroencephalography, brain MRI, magnetic resonance angiography, repetition of laboratory tests plus measurement of von Willebrand factor-cleaving protease activity, measurement of levels of antibodies to double-stranded DNA and platelets, and renal biopsy.

Diagnosis Thrombotic microangiopathic hemolytic anemia with a possible underlying diagnosis of malignant hypertension, antiphospholipid antibody syndrome, catastrophic antiphospholipid antibody syndrome, thrombotic thrombocytopenic purpura, or active SLE.

Management At the general hospital, therapy included a single dose of intravenous cyclophosphamide 500 mg, eight daily plasma exchange treatments, three daily infusions of methylprednisolone 1 g followed by methylprednisolone 60 mg every 8 h, an infusion of rituximab 657 mg and ultrafiltration via hemodialysis. At the tertiary institution, therapy included an infusion of cyclophosphamide 650 mg, aspirin 81 mg daily, prednisone 40 mg daily, mycophenolate mofetil 750 mg twice daily, and aggressive management of hypertension.

ΣΕΛ

- Πιο συγκεκριμένα θρομβωτική μικροαγγειοπάθεια εχει αναφερθεί σε ποσοστό εως και 2% των ασθενών με Λύκο



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Lupus. 2009 Jan;18(1):16-21. doi: 10.1177/0961203308094360.

Thrombotic thrombocytopenic purpura in systemic lupus erythematosus: risk factors and clinical outcome: a single centre study.

Kwok SK[1], Ju JH, Cho CS, Kim HY, Park SH.

Author information

Abstract

The study was undertaken to investigate clinical characteristics of thrombotic thrombocytopenic purpura (TTP) in patients with SLE and to determine risk factors and clinical outcome of TTP in patients with SLE. Among the 1203 patients with SLE admitted to catholic medical centre of the catholic university of Korea from January 1990 to December 2006, 26 patients with SLE were found to admit with TTP. TTP was defined if microangiopathic haemolytic anaemia, thrombocytopenia and negative Coombs' test were present and when at least one of the following signs was noted: renal impairment, neurologic deficit or fever. Eighty-seven patients with SLE who admitted with other manifestations, matched for age and sex, were included as disease controls. Data were retrospectively analysed based on medical records. There were no significant demographic characteristics between SLE patients with TTP and those with other manifestations. Multivariate analysis showed that independent risk factors for the development of TTP included high SLE disease activity index score (SLEDAI > 10, P = 0.006) and coexisting nephritis (P = 0.004). Among the 26 SLE patients with TTP, 12 died during admission period (in-hospital mortality rate: 46.1%). SLE patients with infection or neurologic manifestations had higher mortality rates. Multivariate analysis showed that infection is the only independent risk factor for mortality in SLE patients with TTP (P = 0.035). Patients with SLE who are in the active stage or who have renal involvement have the increased risk for TTP. Development of TTP in patients with SLE can be fatal. Therefore, intensive therapy will be needed especially in the presence of infection.

Lupus. 2009 Jan:18(1):16-21

ΣΕΛ

- Μάλιστα οι λοιμώξεις αναφέρεται ότι μπορούν να πυροδοτήσουν οχι μόνο flair-up του Λύκου αλλα και ΤΜΑ στους ασθενείς αυτούς.

RHEUMATOLOGY
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Advance Access publication 11 December 2010

Original article

Thrombotic microangiopathy in systemic lupus erythematosus: a cohort study in North Taiwan

Ming-Han Chen[1,2], Ming-Huang Chen[2,3], Wei-Sheng Chen[1,2], Peter Mu-Hsin Chang[2,3], Hui-Ting Lee[1,2], Hsiao-Yi Lin[1,2] and De-Feng Huang[1,2]

Abstract

Objectives. Thrombotic microangiopathy (TMA) co-existing with SLE is rarely reported. This study aimed to investigate the triggering factors, clinical features and outcomes of SLE patients with TMA in Northern Taiwan.

Methods. Twenty-five TMA cases out of 2461 SLE patients admitted to Taipei Veterans General Hospital, between 2000 and 2010, were enrolled.

Results. When TMA occurred, 16 (64.0%) patients had infection; 22 (88.0%) were in an active disease state with a SLEDAI score >10. Among the infection group, 13 (81.3%) had an increase in the SLEDAI score of ⩾4. We found that older age (⩾50 years), low platelets (⩽20 000/nm^3), presence of infection, acute renal failure (ARF) or four or more TMA features were independent risk factors for persistent haematological abnormalities ($P < 0.05$); older age (⩾50 years) and a high reticulocyte index (>2%) were the risk factors for persistent renal function impairment ($P < 0.05$). The overall mortality rate was 52.0% (13 out of 25); older age (⩾40 years), low complement value, presence of infection ($P < 0.001$), two or more infection sources, ARF and four or more TMA features were the statistically significant factors contributing to a higher mortality rate. Patients receiving plasma exchange seven times or more had a significantly higher rate of improvement in renal function and haematological abnormalities.

Conclusions. Our study showed that infection was one of the major triggers for the flare-up of SLE disease activity and occurrence of TMA in SLE. Infection is also a strong risk factor for outcome in SLE patients with TMA. Plasma exchange can be considered as an adjuvant treatment modality.

Key words: Thrombotic microangiopathy, Systemic lupus erythematosus, Infection, Plasma exchange.

ΣΕΛ

Literature-review 105 περιπτώσεων ΤΜΑ σε ασθενείς με ΣΕΛ σύμφωνα με το οποίο:

- Η διάγνωση του ΣΕΛ ετέθη ταυτόχρονα με TTP/HUS στο 45.7% η ακολούθησε αυτήν της ΤΜΑ σε ποστοστό 3.8%
- Το 83.8% είχαν νεφρική συμμετοχή
- Η συνδυασμένη θεραπεία (Πλασμαφαίρεση +κορτικοειδή/κυτταροτοξικοί παράγοντες) πέτυχε υψηλότερα ποσοστά ύφεσης

Clin Rheumatol (2014) 33:419–427

ΣΕΛ

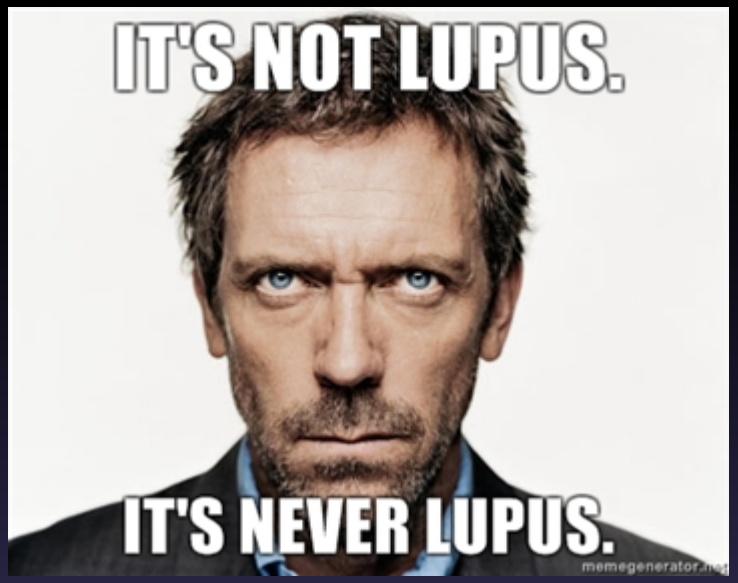
Συμπερασματικά:

1) TTP/HUS μπορει να συμβεί στο 2% των ασθενών με Λύκο
2) Λοιμώξεις μπορούν να αποτελέσουν triggering-factor για TTP-HUS στους ασθενείς αυτούς
3) Η διάγνωση του Λύκου μπορεί να έπεται ή να τεθεί κατα την διάγνωση TTP-HUS σε ποσοστό 50%
4) Η νεφρική συμμετοχή είναι πολύ συχνή
5) Συνδυαστική θεραπεία ειναι πιο αποτελεσματική

ΣΕΛ

Ο ασθενής παρουσιάζει παρατεινόμενο εμπύρετο και :

- Θετικά ANA 1:640
- Χαμηλό συμπλήρωμα C3,C4
- Παράταση aPTT
- Πλευριτική και ασκιτική συλλογή
- Νεφρική ανεπάρκεια
- Λεμφοπενία/θρομβοπενία


IT'S NOT LUPUS.
IT'S NEVER LUPUS.
memegenerator.net

ΣΕΛ

Περαιτέρω έλεγχος με :

- Anti-dsDNA
- Anti-SM
- Coombs
- Αντιφωσφολιπιδικα αντισώματα
- Βιοψία νεφρού?

TTP/HUS

Η διάκριση των δύο αυτών νοσημάτων βασιζόμενη σε κλινικά κριτήρια ενδέχεται να είναι αδύνατη καθώς το HUS μπορεί να έχει εξωνεφρικές εκδηλώσεις και αντίστοιχα η TTP να προσβάλλει τους νεφρούς.

Παθοφυσιολογικά η TTP χαρακτηρίζεται απο μειωμένη δραστικότητα της μεταλλοπρωτεάσης ADAMTS13 η οποία διασπά τα πολυμερή Von Willebrand

Ενώ στο HUS η κυρίαρχη διαταραχή είναι η άκριτη ενεργοποίηση της εναλλακτικής οδού του συμπληρώματος με τελικό αποτέλεσμα την δημιουργία του MAC στις μεμβράνες των κυττάρων

TTP/HUS

Στο περιστατικό μας αφού αποκλείσουμε τα συγγενή αίτια σκεφτόμαστε :

- Ιδιοπαθές TTP/HUS
- Λοιμώξεις (shiga toxin- e.coli, HIV, Strep.pneumoniae, other infections)
- Φαρμακα (quinine,χημειοθεραπευτικα)
- Αυτοάνοσα νοσήματα (ΣΕΛ,αντιφωσφολιπιδικο σύνδρομο)
- κακοήθειες

TTP/HUS

Transfusion. 2014 Feb;54(2):389-97. doi: 10.1111/trf.12263. Epub 2013 May 27.

High prevalence of infectious events in thrombotic thrombocytopenic purpura and genetic relationship with toll-like receptor 9 polymorphisms: experience of the French Thrombotic Microangiopathies Reference Center.

Morgand M[1], Buffet M, Busson M, Loiseau P, Malot S, Amokrane K, Fortier C, London J, Bonmarchand G, Wynckel A, Provôt F, Poullin P, Vanhille P, Presne C, Bordessoule D, Girault S, Delmas Y, Hamidou M, Mousson C, Vigneau C, Lautrette A, Pourrat J, Galicier L, Azoulay E, Pène F, Mira JP, Rondeau E, Ojeda-Uribe M, Charron D, Maury E, Guidet B, Veyradier A, Tamouza R, Coppo P; Thrombotic Microangiopathies Reference Center.

⊕ Collaborators (44)

⊕ Author information

Abstract

BACKGROUND: Infectious events have been reported as major environmental triggers of thrombotic thrombocytopenic purpura (TTP). We detail here the potential association between infections and TTP.

STUDY DESIGN AND METHODS: We recruited randomly and prospectively a cohort of 280 consecutive TTP patients during a 9-year period. Features of infection were systematically recorded.

RESULTS: Features consistent with an infectious event were observed in 114 patients (41%) at time of TTP diagnosis. Infectious agents were documented in 34 cases and were mainly Gram-negative bacilli. At time of diagnosis infected patients more frequently had fever ($p < 0.001$). Infections at diagnosis did not impact prognosis and outcome. Thirty-six percent of patients experienced an infectious event during hospitalization, which resulted in more exacerbation of TTP ($p = 0.02$). Infections were not overrepresented during treatment in patients who received steroids and/or rituximab. Further genetic analysis of toll-like receptor (TLR)-9 functionally relevant polymorphisms revealed that TLR-9 +2848 G and TLR-9 +1174 A genotypes were more frequent in TTP patients than in controls ($p = 0.04$ and $p = 0.026$, respectively) and more particularly in patients negative for the Class II human leukocyte antigen system susceptibility allele DRB1*11 ($p = 0.001$ and $p = 0.002$, respectively). Haplotypes estimation showed that 1174A-2848G haplotype was significantly more frequent in TTP ($p = 0.004$), suggesting a primary role for this haplotype variation in conferring a predisposition for acquired TTP.

CONCLUSION: Infections should be considered as an aggravating factor during the course of TTP. Particular polymorphisms in TLR-9 gene may represent risk factors for TTP.

Πιθανές διαγνώσεις

1) TTP/HUS δευτεροπαθές σε έδαφος κακοήθειας,λοίμωξης ή αυτοάνοσου νοσήματος

2) ΣΕΛ

3) Κακοήθεια