MONOCLONAL GAMMOPATHY OF CLINICAL SIGNIFICANCE (MGCS)

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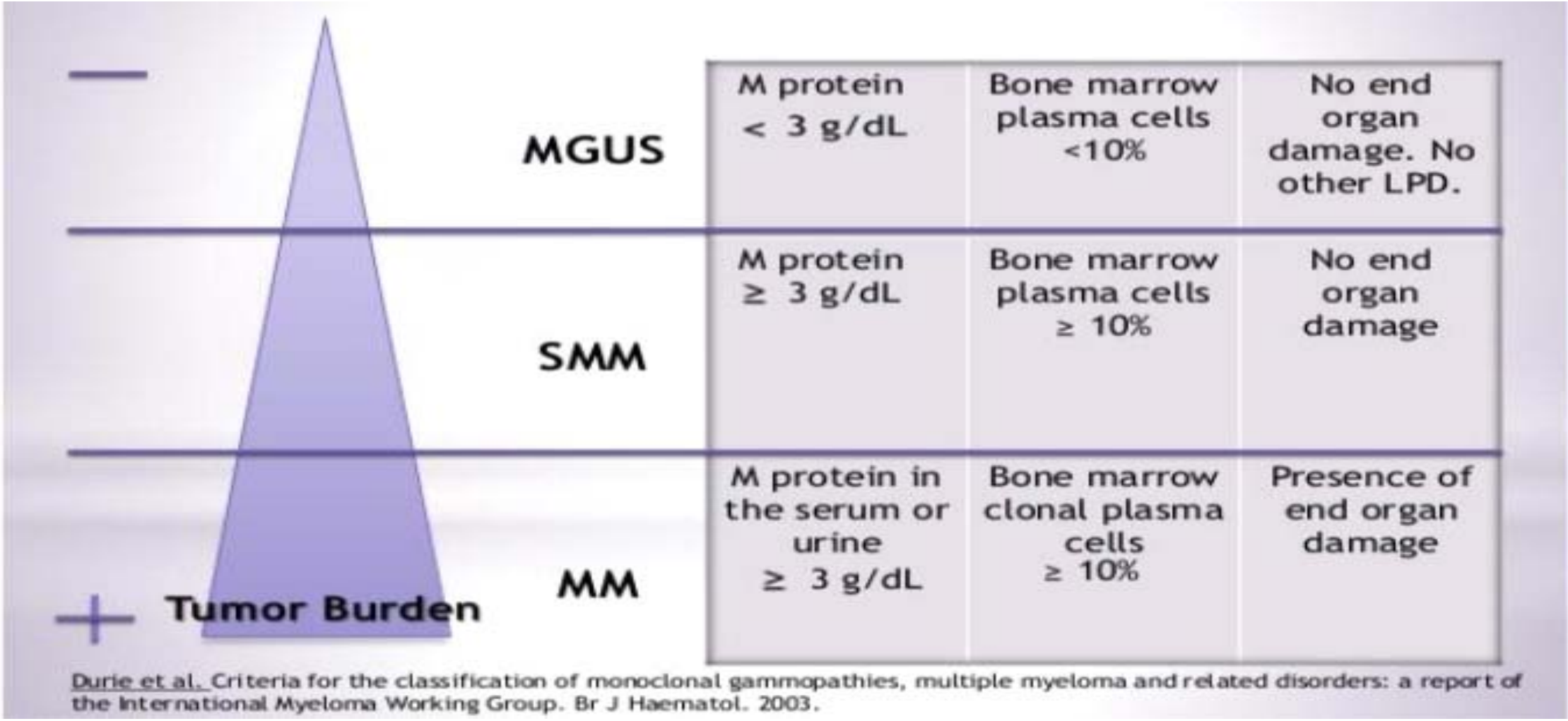
ΜΟΝΟΚΛΩΝΙΚΕΣ ΓΑΜΜΑΠΑΘΕΙΕΣ



MGUS	M protein < 3 g/dL	Bone marrow plasma cells <10%	No end organ damage. No other LPD.
SMM	M protein ≥ 3 g/dL	Bone marrow plasma cells ≥ 10%	No end organ damage
MM	M protein in the serum or urine ≥ 3 g/dL	Bone marrow clonal plasma cells ≥ 10%	Presence of end organ damage

Durie et al. Criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders: a report of the International Myeloma Working Group. Br J Haematol. 2003.

	Mechanism	Main characteristics of monoclonal gammopathy	Main organ(s) involved	Reference
Autoantibody activity				
Type II mixed cryoglobulinemia*	Rheumatoid	IgM	Immune complex–mediated vasculitis; skin +++, kidney, peripheral nerve; may be systemic	28,60
C1 inhibitor deficiency	C1 inhibitor		Angioedema	25
Von Willebrand disease	vW factor		Bleeding	26
Bullous skin diseases	Dermoepidermal junction (collagen VII)		Skin	21
Xanthomatosis	Various lipoproteins	Usually IgG	Cholesterol accumulation in macrophages; skin and tendons; other localizations (necrobiotic xanthogranumathosis)	29,30
Cold agglutinin disease	Red blood cell (Ii)	IgM	Cold-induced skin manifestations + intravascular hemolysis	27
IgM-associated peripheral neuropathy	MAG +++	IgM	Peripheral nerve; ataxic polyneuropathy (anti-MAG)	22,23
	Gangliosides		CANOMAD	
CAP* activation				
C3 glomerulonephritis	Mechanism to be determined; autoantibody activity against CAP regulator protein (factor H) in some cases	IgG	Kidney only	31-34
Atypical hemolytic-uremic syndrome			Systemic	
Cytokine mediated				
POEMS syndrome	VEGF	λ LC (~100%), IgA 50% Vλ1 (#100%) Osteosclerotic bone lesions	Peripheral nerve (100%) and various other manifestations	35,36

Unknown mechanism				
Systemic capillary leak syndrome		IgG, IgA (rare)	Systemic	45
TEMPI syndrome		IgG	Systemic	46,47
Neutrophilic dermatosis†		IgA >80% (except Sweet syndrome)	Skin +++; different types and different associated manifestations	39
Acquired cutis laxa		Usually IgG; association with γ HCDD	Skin +++ Other manifestations (lung, digestive tract)	43
Scleromyxedema		IgG with slow electrophoretic mobility	Skin +++; other localizations	40,41,52
Scleroedema		IgG	Skin only	41
Schnitzler syndrome	Acquired autoinflammatory syndrome by IL-1 deregulation?	IgM	Skin +++; systemic symptoms; osteosclerotic bone lesions	44
Sporadic late-onset nemaline myopathy			Exclusively muscles (skeletal and possibly cardiac)	48

ΑΠΑΡΑΙΤΗΤΕΣ ΕΞΕΤΑΣΕΙΣ

- Επί υποψίας MGRS

1. OMB –Παρουσία μικρού αλλά τοξικού κλώνου
2. Βιοψία νεφρού- διήθηση του νεφρού από μονοκλωνικές εναποθέσεις όμοιες με την μονοκλωνική πρωτείνη του ορού
3. SPEP IFE και FLC ratio

Θεραπεία

- Απαραίτητη η εξάλειψη του παθολογικού κλώνου
- Θεραπεία όπως ΠΜ με σκοπό CR

  Velcade-Endoxan-Dex, Revlimid, Daratumumab, Μεταμόσχευση μυελού-νεφρού

- Πρόγνωση καλύτερη του ΠΜ, όπως στην αμυλοείδωση