

Questionnaire: Diagnosis of Mast Cell Activation Disease

The indicated values for those items acknowledged by or found in the patient are summed.
 A total score above 8 but less than 14 indicates a pathological activation of mastcells.
 At a total score of 14 and more, a systemic mast cell mediator release syndrome is clinically verified.

CLINICAL SIGNS

1	The patient complains about recurring or continuing burning and/or crampy abdominal pain of unknown cause, and/or recurring or continuing diarrhoea of unknown cause, and/or frequently intense meteorism/gassiness (independent of the composition of diet), and/or about episodically occurring nausea.		1
2	The symptoms respond to treatment with H1-antihistamines.		1
3	The progression of the symptoms occurred in episodes		1
4	The progression of the symptoms occurred in episodes with symptom-free periods becoming shorter.		1
5	The patient complains about episodically occurring burning and/or choking chest pain attacks, which are often experienced as life-threatening. Electro cardiographic findings are without pathological signs.		1
6	The patient complains about occasional or continuing pain in the urinary bladder and/or pelvis, accompanied by painful desire to void and/or blood in the urine. There is no bacteriuria.		1
7	The patient complains about occasional or continuing paresthesia (burning, pins and needles, numbness) and/or pain which does not respond to treatment with analgesics.		1
8	Gastroscopy and biopsies from the stomach and duodenum are:		0
	a. Without pathological findings.		0
	b. Or show minor signs of inflammation.		1
	c. Or show <i>Helicobacter pylori</i> - and NSAID-negative erosion and/or ulcer.		3
	d. Or show clusters of mast cells and/or a considerable number of spindle-shaped mast cells and/or CD25-positive mast cells.		10
9	Colonoscopy and intestinal biopsies are without:		0
	a. Pathological findings.		0
	b. Or show minor signs of inflammation.		1
	c. Or show melanosis coli (abuse of anthra-cenediones ruled out).		1
	d. Or show clusters of mast cells, and/or a considerable number of spindle-shaped mast cells, and/or CD25-positive mast cells.		10
10	The patient reports the following signs of episodically occurring symptoms of autonomic dysfunction:		1
	a. Tachycardia or palpitation/dysrhythmia.		1
	b. Flush (redness, feeling of heat).		2
	c. Hot flash, sweat.		2
	d. Paroxysmal hypo/hypertension with dizziness to the point of syncope.		2
11	Although there are no pathological findings in routine laboratory parameters and imaging methods, the patient presents with:		1
	a. A pronounced asthenia.		1
	b. Fatigue.		1
	c. Loss in weight.		1
12	During the symptomatic periods of the disorder the patient is:		1
	a. Afflicted with anal pruritus and/or anal eczema.		1
	b. Intestinal adhesions are present without prior history of abdominal surgery		1
SECTION TOTAL			

TRIGGERING FACTORS

13	Deprivation of sleep		1
14	Fasting for 24 hours		1
15	Histamine containing food (e.g. red wine, cheese, tuna)		1
SECTION TOTAL			

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LABORATORY PARAMETERS

16	The patient shows signs of a bleeding diathesis (e.g. abnormal secondary bleeding or bruises after minimal trauma and/or lesions).	1
17	During the symptomatic periods of the disorder the patient showed, at least once, hyperbilirubinemia (up to 2.5 mg/dL), and/or an increase of trans amidases (up to twice their upper limits of normal), and/or diet-independent hypercholesterolemia (up to 7.8 mmol/L).	1
18	There are low titers of autoantibodies without clinical signs in the organs or tissues against which the autoantibodies are directed.	1
19	a. The serum total tryptase was normal.	0
	b. The serum total tryptase was elevated > 11 and < 20 ng/mL.	3
	c. The serum total tryptase was elevated more than 20 ng/mL.	10
20	a. The level of heparin in blood was normal.	0
	b. The level of heparin in blood was elevated > 0.05 anti-Factor Xa units/mL.	3
21	a. The level of N-methylhistamine in a 12-hurine collection was normal.	0
	b. The level of N-methylhistamine in a 12-hurine collection was marginally elevated.	1
	c. The level of N-methylhistamine in a 12-hurine collection was elevated up to tenfold of the reference value.	5
	d. The level of N-methylhistamine in a 12-hurine collection was elevated by more than tenfold of the reference value.	10
SECTION TOTAL		

IMAGING METHODS

22	The patient has splenomegaly and/or hepatomegaly.	1
23	The patient has bone pain with signs of osteoporosis and/or osteopenia and/or osteosclerosis.	1
SECTION TOTAL		

MEDICAL HISTORY

24	The patient shows involvement of the skin in terms of:	
	a. Brown-reddish maculopapulous rash/eruption.	2
	b. Angioedema of the lips, lids of the eye, infraorbital.	2
	c. Pruritus (itching) without rash/eruption and/or disease-related folliculitis.	1
	d. A clear increase in the number of telangiectasias.	1
25	The patient reports sudden attacks of migraine-like headache.	1
26	The patient reports memory loss (ability to remember names or words) and/or concentration difficulty and/or sleep disturbances.	1
27	The patient reports tinnitus attacks and/or ocular discomfort (dry eyes, red eyes, stinging eyes), and/or rhinorrhea/chronic nasal congestion and/or stomatitis. (Score if two or more of these symptoms are present)	1
28	The patient reports non-allergic respiratory ailments, such as asthma, compulsion to clear the throat, titillating/ticklish feeling in the respiratory tract and/or shortness of breath during routine tasks.	1
29	In the past, common viral infections of the upper respiratory tract were frequently complicated by bacterial superinfection.	1
30	The patient can state precisely the date of the first clinical manifestation of the mast cell mediator release syndrome because it appears to them to be associated with an infectious disease.	1
SECTION TOTAL		
GRAND TOTAL		

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