Test yourself in mast cell activation disease

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Choose only one answer:

1. The appearance of blistering when rubbing a lesion of urticaria pigmentosa is termed:
a. Dermatographism.
b. Darier's sign.
c. Darlymple's sign.
d. Nikolsky sign.

2. All of the following are clinical manifestations of systemic mastocytosis except:
a. Anaphylactic shock.
b. Flushing & headache.
c. Diarrhea & dyspepsia.
d. Nasal congestion & bronchospasm.
e. Hypertensive encephalopathy

3. The term mast cell activation syndrome should be applied when:
a. Clinical signs of severe recurrent or chronic systemic mast cell activation are present.
b. An increase in serum tryptase to at least 20% above baseline + additional 2 ng/mL measured during a clinical episode.
c. The symptoms respond to therapy with mast cell stabilizing agents
d. All of the above.

4. The first-line treatment in patients with mast cell activation disease suffering from osteoporosis is:
a. Vitamin D alone.
b. Vitamin D plus calcium application.
c. Biphosphonates.
d. Calcium application alone.

5. Differential diagnoses of mast cell activation syndrome include:
a. Myocardial infarction.
b. Adrenal insufficiency.
c. Epilepsy.
d. Hereditary or acquired angioedema.
e. All of the above.

6. Mast cell activation disease denotes a collection of disorders, including:
a. Mastocytosis and mast cell leukemia.
b. Mastocytosis, mast cell activation syndrome and mast cell leukemia.
c. Mastocytosis and mast cell activation syndrome.
d. Cutaneous mastocytosis, mast cell activation syndrome and mast cell leukemia.

7. WHO criteria for establishing the diagnosis of systemic mastocytosis is:
a. Multifocal dense infiltrates of mast cells more than 15 mast cells in aggregates in bone marrow biopsies.
b. Multifocal dense infiltrates of mast cells more than 15 mast cells in aggregates in bone marrow biopsies in association with serum tryptase > 20 ng/ml.
c. Multifocal dense infiltrates of mast cells more than 15 mast cells in aggregates in bone marrow biopsies in association with serum tryptase = 20 ng/ml.
d. S. tryptase > 20 ng/ml and c-kit mutation in tyrosine kinase at codon 816 in mast cells in extracutaneous organs are enough to diagnose systemic mastocytosis.
e. None of the above.

8. Clinical complaints as a result of a pathologically increased mast cell activity is termed:
a. Mast cell mediator release syndrome.
b. Mast cell activation syndrome.
c. Mast cell activation disease.
d. Systemic mastocytosis.

9. The term cutaneous mastocytosis means:
a. Urticaria pigmentosa.
b. Diffuse cutaneous mastocytosis.
c. Mastocytoma of the skin.
d. Telangiectasia macularis eruptive perstans (TMEP).
e. All of the above.

(Answers on page 98)