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Pathophysiology Review: Mastocytosis — Or Allergies Behaving Badly

- 12 February 2018
- *by:* Cindy Kibbe, CCS, BS (CLS)
- *in:* Coders' Corner
- *note:* no comments

 *by Cindy Kibbe, CCS, BS (CLS)*

Mastocytosis is a rare immune cell disorder that affects some estimated 200,000 people in the United States.[1]

OK, if this is such a rare disease, why should we as coders be concerned? Between treatment from a recent Coding Clinic and a number of new codes and Exclusions in 2018, this lesser-known disease has had quite the make-over.

Let's take a closer look at this intriguing disease and the changes we will see in our coding.

Defining Mastocytosis
According to The Mastocytosis Society, mastocytosis is an abnormal accumulation of mast cells in one or more organ systems.[2] There are three main categories:

- Cutaneous — Increase in mast cells in the skin
- Systemic — Abnormal accumulation of mast cells in organs[3]
- Mast Cell Sarcoma — Tumors composed of abnormal mast cells invading tissues[4]

All three forms can occur in both children and adults.[3]

What Do Normal Mast Cells Do?
Before we can go further into the pathophysiology of mastocytosis, let's do a quick review of how normal mast cells work.

A mast cell is a type of white blood cell (granulocyte) that resides in tissues. It plays a key role in the immune system by releasing different types of granules (cytokines) that act as chemical "alarm signals" to fight disease.

One particularly important granule in mast cells is histamine. Technically, histamine causes smooth muscle contraction and vasodilation.[3] Anyone who has ever experienced hay fever or a peanut allergy is probably very familiar with histamines — it's one of the chemicals in your body that causes all the itching, rashes, and flushing during an allergy attack.

When you are exposed to a particular allergen to which you are sensitive, that allergen binds to receptors on mast cells. Once that happens, mast cells release all those lovely histamine granules and other cytokines. *Voila!* An allergic reaction occurs.

For most of us, this is largely a nuisance. For people with mastocytosis, an allergic response by mast cells can be life-threatening.

Mast Cells in Mastocytosis

Because there are too many mast cells in those with mastocytosis, the most minor of triggers can quickly become an overwhelming hypersensitivity reaction and anaphylaxis. Mastocytosis can also have lingering symptoms such as musculoskeletal pain, abnormal skin pigmentations, and even cancer.

Mast cell triggers can truly be just about anything. Some common ones include:

- Allergens (Food, venoms, drugs, etc.)
- Temperature changes
- Stress
- Mechanical irritation (e.g. a light scratch on the skin)
- Sun exposure
- Infections[2],[1]

Mastocytosis and Coders

In previous years, coders attempting to assign codes to this esoteric but present disease had a heck of a time. There was no way to differentiate mastocytosis from related conditions such as mast cell activation syndrome, or MCAS (which has its own sub-types).

Coding Clinic 4Q16 gave some guidance on using then-newer codes of D89.4 —, Mast Cell Activation Syndrome.[5]

In 2018, we now have a whole slew of new codes covering mastocytosis, MCAS, and mast cell neoplasms. Oh, my!

ICD-10-CM, Chapter 2 – Neoplasms, added several new codes in 2018.[6],[7] Notice the addition of several new codes and more Exclusions.

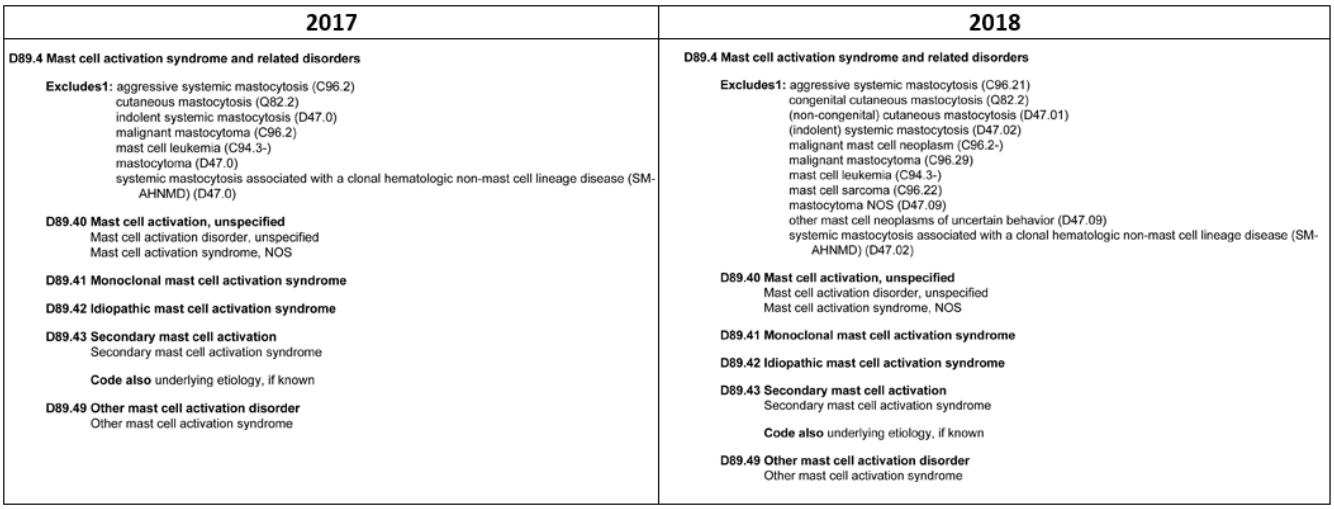
Let's compare:

2017	2018
C96.2 Malignant mast cell tumor Aggressive systemic mastocytosis Mast cell sarcoma **Excludes1:** indolent mastocytosis (D47.0) mast cell leukemia (C94.30) mastocytosis (congenital) (cutaneous) (Q82.2)	**C96.2 Malignant mast cell neoplasm** **Excludes1:** indolent mastocytosis (D47.02) mast cell leukemia (C94.30) mastocytosis (congenital) (cutaneous) (Q82.2) **C96.20 Malignant mast cell neoplasm, unspecified** **C96.21 Aggressive systemic mastocytosis** **C96.22 Mast cell sarcoma** **C96.29 Other malignant mast cell neoplasm**
D47 Other neoplasms of uncertain behavior of lymphoid, hematopoietic and related tissue **D47.0 Histiocytic and mast cell tumors of uncertain behavior** Indolent systemic mastocytosis Mast cell tumor NOS Mastocytoma NOS **Excludes1:** malignant mast cell tumor (C96.2) mastocytosis (congenital) (cutaneous) (Q82.2) **D47.1 Chronic myeloproliferative disease** Chronic neutrophilic leukemia Myeloproliferative disease, unspecified **Excludes1:** atypical chronic myeloid leukemia BCR/ABL-negative (C92.2-) chronic myeloid leukemia BCR/ABL-positive (C92.1-) myelofibrosis NOS (D75.81) myelophthisic anemia (D61.82) myelophthisis (D61.82) secondary myelofibrosis NOS (D75.81)	

	D47 Other neoplasms of uncertain behavior of lymphoid, hematopoietic and related tissue **D47.0 Mast cell neoplasms of uncertain behavior** **Excludes1:** congenital cutaneous mastocytosis (Q82.2-) histiocytic neoplasms of uncertain behavior (D47.Z9) malignant mast cell neoplasm (C96.2-) **D47.01 Cutaneous mastocytosis** Diffuse cutaneous mastocytosis Maculopapular cutaneous mastocytosis Solitary mastocytoma Telangiectasia macularis eruptiva perstans Urticaria pigmentosa **Excludes1:** congenital (diffuse) (maculopapular) cutaneous mastocytosis (Q82.2) congenital urticaria pigmentosa (Q82.2) extracutaneous mastocytoma (D47.09) **D47.02 Systemic mastocytosis** Indolent systemic mastocytosis Isolated bone marrow mastocytosis Smoldering systemic mastocytosis Systemic mastocytosis, with an associated hematological non-mast cell lineage disease (SM-AHNMD) **Code also**, if applicable, any associated hematological non-mast cell lineage disease, such as: acute myeloid leukemia (C92.6-, C92.A-) chronic myelomonocytic leukemia (C93.1-) essential thrombocytosis (D47.3) hypereosinophilic syndrome (D72.1) myelodysplastic syndrome (D46.9) myeloproliferative syndrome (D47.1) non-Hodgkin lymphoma (C82-C85) plasma cell myeloma (C90.0-) polycythemia vera (D45) **Excludes1:** aggressive systemic mastocytosis (C96.21) mast cell leukemia (C94.3-) **D47.09 Other mast cell neoplasms of uncertain behavior** Extracutaneous mastocytoma

(Click on the images above to enlarge.)

Changes were also made in Chapter 3, Blood Diseases in 2018[6],[7] :



2017	2018
D89.4 Mast cell activation syndrome and related disorders **Excludes1:** aggressive systemic mastocytosis (C96.2) cutaneous mastocytosis (Q82.2) indolent systemic mastocytosis (D47.0) malignant mastocytoma (C96.2) mast cell leukemia (C94.3-) mastocytoma (D47.0) systemic mastocytosis associated with a clonal hematologic non-mast cell lineage disease (SM-AHNMD) (D47.0) **D89.40 Mast cell activation, unspecified** Mast cell activation disorder, unspecified Mast cell activation syndrome, NOS **D89.41 Monoclonal mast cell activation syndrome** **D89.42 Idiopathic mast cell activation syndrome** **D89.43 Secondary mast cell activation** Secondary mast cell activation syndrome **Code also** underlying etiology, if known **D89.49 Other mast cell activation disorder** Other mast cell activation syndrome	**D89.4 Mast cell activation syndrome and related disorders** **Excludes1:** aggressive systemic mastocytosis (C96.21) congenital cutaneous mastocytosis (Q82.2) (non-congenital) cutaneous mastocytosis (D47.01) (indolent) systemic mastocytosis (D47.02) malignant mast cell neoplasm (C96.2-) malignant mastocytoma (C96.29) mast cell leukemia (C94.3-) mast cell sarcoma (C96.22) mastocytoma NOS (D47.09) other mast cell neoplasms of uncertain behavior (D47.09) systemic mastocytosis associated with a clonal hematologic non-mast cell lineage disease (SM-AHNMD) (D47.02) **D89.40 Mast cell activation, unspecified** Mast cell activation disorder, unspecified Mast cell activation syndrome, NOS **D89.41 Monoclonal mast cell activation syndrome** **D89.42 Idiopathic mast cell activation syndrome** **D89.43 Secondary mast cell activation** Secondary mast cell activation syndrome **Code also** underlying etiology, if known **D89.49 Other mast cell activation disorder** Other mast cell activation syndrome

(Click on the image above to enlarge.)

While there were no new codes to D89.4 in 2018, several new Excludes codes were added.

As you can see, there has been quite a bit of changes made to this little-known but serious disease in ICD-10. With the push to gain greater specificity in all of our code sets, it is good to review updates to even diseases we might not see in our everyday coding lives.

Cindy Kibbe photo credit: Wendy Wood

Sources:

1. National Institutes of Health
2. The Mastocytosis Society
3. The Cleveland Clinic
4. RareDiseases.org
5. Coding Clinic, Fourth Quarter, 2016
6. 2017 ICD-10-CM
7. 2018 ICD-10-CM

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About the Author

Cindy Kibbe, CCS, BS (CLS)
Cindy Kibbe is a CCS-credentialed coding professional and an award-winning writer. She also has a background as a clinical application analyst, a quality management coordinator, and a registered medical technologist. She is currently seeking a position as an inpatient coder or HIM coordinator.