

Clarification of the HEME Multiple Primary Rule M2 EXCEPTION

In the current HEME Manual, published October 2025, Multiple Primary Rule **M2: Single Histology** states to abstract a SINGLE PRIMARY when there is a SINGLE HISTOLOGY. However, there is an **important EXCEPTION to this involving MALT lymphomas.**

Occurrences of multiple MALT lymphomas are NOT ALWAYS SINGLE PRIMARIES. The exception rule states to “Abstract multiple primaries when a **nodal MALT (C770-C779, 9699/3)** occurs **before or after an extranodal MALT (all other sites, 9699/3).** **These are two distinct lymphomas that have the same histology code.”**

Clarification was sought from SEER regarding this exception when a nodal MALT lymphoma presented either before or after an extranodal MALT lymphoma where the involved lymph nodes were considered to be regional nodes for that extranodal lymphoma site OR in situations where the patient was NEVER disease free from either type of MALT lymphoma.

According to SEER (AASR #46864), there are NO EXCEPTIONS to the EXCEPTION rule. Per SEER, “the exception rule does NOT reference either involved nodal locations OR disease-free status because neither is a factor in applying the rule. If you have a nodal BEFORE or AFTER an extranodal MALT, they are TWO primaries per Rule M2.”

M2 EXCEPTION RULE would apply in the following examples:

- Patient was dx'd 2018 w/ extranodal MALT lymphoma of lung (no LAD at time of dx) w/ STABLE disease until 2026 when patient develops hilar/mediastinal LAD found to be c/w MALT lymphoma
- Patient was dx'd 2018 w/ extranodal MALT lymphoma of lung (no LAD at time of dx); NED s/p tx until 2026 when patient develops hilar/mediastinal LAD found to be c/w MALT lymphoma
 - Per M2 Exception rule BOTH scenarios would be TWO separate primaries: 2018 extranodal MALT dx'd BEFORE a 2026 nodal MALT—*disease status not a factor*

However, the M2 EXCEPTION Rule WOULD NOT APPLY in this situation

- Patient dx'd 2018 w/ RUL extranodal MALT lymphoma of lung (no LAD at time of dx); disease STABLE s/p tx until 2024 when a NEW LUL lung nodule and hilar/mediastinal LAD was found on PET. LN FNA IHC was c/w MALT lymphoma. The new LUL lung nodule was never bxd but felt to be MALT lymphoma
 - The M2 Exception rule would NOT apply in this situation because BOTH an extranodal AND a nodal MALT were occurring at the SAME time following an extranodal MALT diagnosis. This would be a SINGLE primary.