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Are LCIS, GIST and Pseudomyxoma Peritonei Cases Reportable?

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Determining reportability and knowing how to code histology/behavior for many cases can be fairly straightforward. For example, it is clear that Adenocarcinoma, NOS for any site is reportable. We probably all know how to code the Histology and Behavior fields for this histology. While this can offer a source of confidence when processing and coding such cases, there are several pitfalls we've been known to stumble into when it comes to a few other histologies.

There are a handful of histology and behavior combinations that have been known to confuse even the most seasoned registrars among us. These cases have been mistaken as benign when they are malignant or have been thought to be malignant when they are, in fact, benign. When the most current histology rules are not correctly applied, the result is either overreporting or underreporting incidence cases. When we aren't sure whether a particular histology is reportable, we need to take the time to look it up. In this article I'm going to review three of the most common histology/behavior combinations that have proven difficult for us to handle consistently.

Lobular Carcinoma In Situ (LCIS)

The first histology to be reviewed is LCIS. Confusion regarding the reportability of this disease process may have originated as a result of the AJCC TNM Staging 8th Edition change of staging for LCIS from Tis to not being staged. LCIS was included in prior editions of the AJCC staging manuals, but it is removed from the 8th Edition because LCIS is stated to be a benign condition and is not treated as a carcinoma. This, in conjunction with many other 2018 reportability changes, caused many of us to question whether we are still reporting LCIS cases. One might inadvertently assume that if the histology was staged in the past, but no longer is, that it is also no longer reportable.

However, as SINQ-20170080 and the CAnswer Forum (STORE/FORDS/National Cancer Data Base, Call for Data, Submission Specifications, "LCIS 2018" 10/08/2018) remind us, staging does not determine reportability. This is true for several other histologies as well. For example, in situ tumors of the thyroids and leukemia cases are also not staged but they are reportable.

To add to the confusion regarding the reportability of LCIS are the following notations found on the American Cancer Society's (ACS) website:

- Lists LCIS under the site's "Non-Cancerous Breast Conditions" section

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- Refers to LCIS as lobular neoplasia
- Indicates LCIS is different from DCIS and is not cancer.
- States LCIS is a benign (noncancerous) condition that merely puts a patient at risk to develop invasive cancer
- Specifies that LCIS is not a true cancer or pre-cancer, so often no treatment is recommended
- According to the WHO classifications of tumors, the editors of the ICD-O-3 and SEER, the term lobular neoplasia is not considered reportable unless it is actually stated to be LCIS.

Further muddying the waters, the term lobular neoplasia refers to a spectrum of disease featuring atypical lobular hyperplasia at one end of the spectrum and lobular carcinoma in situ (LCIS) at the other end. On the nonreportable, benign/borderline end of the lobular neoplasia spectrum there is LN I-II/LIN I-II as well as atypical lobular hyperplasia (ALH). On the other end of the spectrum, we have LCIS and lobular intraepithelial neoplasia III (LN III/LIN III). These histologies almost exclusively arise in breast tissue, indicate in situ disease with a behavior grade of 2, and are reportable.

So, do we report LCIS of the breast or not? While it is true that, as stated on the ACS website, LCIS doesn't typically develop into an invasive malignant process; it is still an in situ cancer. There is at least a little good news about LCIS; it is always reportable to SEER, NPCR and CoC through at least diagnosis year 2018. It is important to also note that while LCIS is coded to 8520/2, for 2018 there is a new code for pleomorphic LCIS which is 8519/2.

Gastrointestinal Stromal Tumor (GIST)

While LCIS has the benefit of always being reportable, gastrointestinal stromal tumors (GISTs) present a different challenge. These tumors can be benign, borderline, or malignant. The challenge is figuring out what the behavior is. As with all cases, it is important to review reportable terms. This is especially true for GISTs, as they are often described as being at a "high risk of malignant behavior." This is not equivalent to a statement of malignancy and the use of such terminology is not enough to merit reporting the case.

The easiest way to determine reportability is to look for a pathologist statement of malignancy. If the GIST is malignant, it is definitely reportable. However, as SINQ-20140088 explains, pathologists may be hesitant to use the term "malignant." This is made more confusing if a pathologist stages the GIST without stating the behavior of the tumor. As we learned with LCIS, staging does not determine reportability and a pathologist may opt to stage tumors that are not malignant.

So, even when there is staging, if there is no statement of malignancy, we must look elsewhere to determine reportability. The easiest thing to check for is the presence of metastatic deposits in either lymph nodes or distant sites or if the tumor is described as having presented in multiple foci. If a cancer has spread or presents in this manner, it is unquestioningly malignant per the SEER Manual and the STORE Manual.

When there is no information on metastasis on the pathology report, we need to review the medical record to see if we can find a clinical statement of malignancy made by the physician. A documented clinical statement of a malignant GIST may also be used to accession a GIST as malignant (see SINQ 20150027) when the pathologist does not comment on the tumor's behavior but the patient's physician does. The

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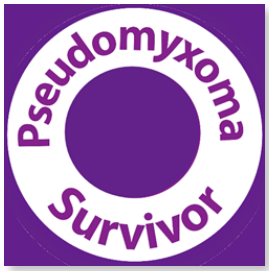
entire clinical and pathological picture may be assessed by the physician and determined to represent a reportable malignant GIST that may or may not be treated with systemic treatment.

At a hospital, if your pathologists don't refer to any GIST as malignant in the pathology report, but in discussion with them, your registry outlines the terminology and tumor or specific case characteristics that they believe are equivalent to a diagnosis of malignancy, the registry may accession these as malignant GISTs provided these guidelines developed with the pathologist are documented in the registry's policies/ procedures. Documentation of this fact is required on the patient abstract prior to submission to the central registry to ensure the behavior and reportability of the case is not changed by the CSS staff.

Pseudomyxoma Peritonei



The last histology I'll discuss is pseudomyxoma peritonei. In the ICD-O-3, pseudomyxoma peritonei is listed with a behavior code of 6 (malignant metastatic or secondary site). However, per SINQ-20031171, this behavior being associated with this histology does not necessarily make these cases reportable. This statement will make more sense after I review what pseudomyxoma peritonei is.

The SEER Glossary defines pseudomyxoma peritonei as a build-up of mucus in the peritoneal cavity. The mucus may come from ruptured ovarian cysts, the appendix or from other abdominal tissues, and mucus-secreting cells may attach to the peritoneal lining and continue to secrete mucus. While the primary may be a malignant tumor, it can also be a non-malignant process, such as a ruptured benign polyp or a low grade appendiceal mucinous neoplasm (LAMN). As such, there must be documentation that either the primary tumor or the implants are malignant before accessioning the case. More often than not, this confirmation will be done histologically. Any statement of pseudomyxoma peritonei that is not explored pathologically should be met with skepticism and reviewed carefully to confirm whether it is actually malignant.

Luckily, the 2018 Solid Tumor Rules have ushered in a two-tiered system created by WHO in 2010 that classifies pseudomyxoma peritonei into one of two groups: High-grade or low-grade. A high-grade pseudomyxoma peritonei is considered malignant (and therefore reportable) while a low-grade pseudomyxoma peritonei is benign. For cases diagnosed prior to 2018, or cases in which the pseudomyxoma isn't classified according to the new WHO guidelines, those forms of pseudomyxoma peritonei are only reportable when the:

Underlying tumor that gave rise to the implants has a reportable histology (behavior of /3)
Primary tumor was previously removed and the pathologist cannot assess it, the patient subsequently developed pseudomyxoma peritonei and the implants must be stated to be malignant or invasive into the underlying structures (behavior of /3)

Conclusion

These three examples are not the only tricky histologies we come across as cancer registrars. For more than a few histologies, it can be challenging to determine reportability and the correct code to reflect the cell type and the behavior of the tumor.

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There are a number of other histology changes going into effect beginning with cases diagnosed in 2018. The NAACCR website has the ICD-O-3 Implementation Guidelines available for quick reference; go to https://www.naaccr.org/implementation-guidelines/#ICDO3. It is important to reference these guidelines if we want to be certain we are including and excluding cases properly for our registries.

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