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Nodal vs Extranodal - Coding Primary Site for Lymphoma Primaries

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When coding the primary site for lymphoma primaries, we tend to use the default primary site code C77.9 (Lymph nodes NOS) more often than we should. Sometimes we forget that almost any organ can be affected by lymphoma. I'm sure we've all stared at some lymphoma cases trying to decide how to code the primary site. Is it a case with disseminated lymph node disease that also happens to involve an extranodal site (primary site is coded to C77.0 – C77.9) or is it a case that actually originated in an extranodal site (primary site would not be C77.0 – C77.9)? Trying to make this coding distinction can be challenging sometimes for all of us.

There are many different types and subtypes of lymphoma. The two main categories are Hodgkin disease and non-Hodgkin lymphoma. While the majority of these lymphomas originate in the lymph nodes, it is estimated that 25-35% of the non-Hodgkin lymphomas are classified as extranodal in most countries. On the other hand, Hodgkin disease is primarily nodal in origin. The only way to code primary site accurately for lymphomas is to use the Heme Database, the PH Rules in the Heme Manual plus imaging, physical exam, chart notes and pathology findings.

Major primary sites for lymphoma

Lymphoma affects the body's lymphatic system, which is part of the immune system responsible for helping to fight infections and some other diseases. It also facilitates movement of fluids through the body. Lymphomas can originate in many parts of the body. The majority of lymphomas arise in lymph nodes or lymphatic sites such as spleen, Waldeyer's ring (C02.4, C09.0-C099, C11.1, and C14.2) or thymus. Lymphomas arising in lymph nodes or these select lymphatic sites are known as "nodal" lymphomas.

Coding primary site for lymphoma

The primary site for lymphoma is coded using the Heme database, scans, clinical documentation, pathology reports and PH Rules in the Heme Manual. The pathology report is not the default for determining primary site. The applicable primary site(s) varies depending on the histology. Although there is no hierarchy for coding primary site, the Heme Database should always be the first resource used in determining the primary site in case there is a specific primary site that applies given the presenting histology. However, the Heme DB will not provide the final primary site decision for every case. When the Heme DB doesn't provide a specific site, the PH rules should be used.

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Of note, secondary involvement of distant lymph nodes (for an extranodal lymphoma), bone marrow, liver, spleen or CNS are included in the EOD staging field only. This excludes rare primary lymphoid neoplasms of spleen, multifocal lung involvement, liver or CNS. Secondary involvement of distant site(s) is disregarded for the purpose of coding primary site. For lymphoid neoplasms, this secondary or distant involvement is similar to metastasis for solid tumors and does not alter the primary site assigned by the physician or determined using the PH Rules.

TIPS on Coding Primary Site

TIP If it is clear that a specific lymph node chain is the primary site, code the primary site to that lymph node chain. For example, assume the only area of involvement is the Parotid lymph nodes. The primary site will be C77.0 (Lymph nodes of head, face and neck) because the parotid lymph nodes are located in this region. Code C77.0 can represent that either more than one lymph node in the parotid chain is involved or more than one chain in the same region is involved. As long as all involved nodes are in the same region, that ICD-O-3 site code may be used. When a mass is identified as retroperitoneal, mesenteric, inguinal, mediastinal, or pelvic and is consistent with lymphoma, code to the specific lymph node site: retroperitoneal or mesenteric (C77.2), inguinal (C77.4), mediastinal (C77.1), or pelvic (C77.5).

TIP For C77.8 to apply, the lymph nodes involved must be in different regions (i.e., have different ICD-O site codes) when you look them up in the ICD-O-3 or in Heme Manual, Appendix C (Lymph Node/ Lymph Node Chain Reference Table) and the physician does not indicate in which node/region the lymphoma arose. For example, if the physician does not indicate where the lymphoma arose, but both axillary (C77.3) lymph nodes and inguinal (C77.4) lymph nodes are involved, the primary site is coded to C77.8 because axillary and inguinal have different topography codes.

TIP In the following situations, use code C77.9 for the primary site:

- If it is not clear what the primary site is, the lymphoma is known to have risen in the lymph nodes and there is no documentation of which lymph nodes are involved.
- If the medical record only states that the patient has lymphoma, NOS without indicating specific details (i.e., history only cases).
- If the only documentation we have in the medical record is a lymph node biopsy that confirms lymphoma pathologically, the primary site is coded to C77.9 rather than C80.9. In this situation, we don't know whether there are other areas of involvement yet. However, we know that at least one lymph node was biopsied and involved, so the site code of C80.9 (unknown primary) would not be appropriate in this situation. While it is true we don't know exactly what the primary site is (one chain, multiple chains, lymph nodes or extranodal sites, etc.), we are instructed to use C77.9 (lymph nodes, NOS) in this situation.

TIP If there are multiple areas of involvement, the clinician may choose to biopsy a particular lymph node because it is the most accessible. Do not automatically code primary site to the lymph node

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biopsied, unless it is the only chain involved or it is stated to be the primary site, of course. That was the first TIP we discussed.

TIP If there is involvement of an organ (extranodal site) only or an extranodal PLUS involvement of the organ's regional lymph nodes only, code the primary site to the organ (extranodal site). Examples:

- Code primary site to C16.9 (stomach, NOS) if there is no involvement other than the stomach.
- Code primary site to C18.7 (sigmoid colon) if only the sigmoid colon and the pericolic lymph nodes are involved.

TIP If it is suspected that the lymphoma is extranodal, or there is involvement in multiple organs without information to identify the primary site, or the physician does not document the site, code primary site to C80.9 (Unknown primary site). However, keep in mind that if lymph nodes are known to be involved, the primary site cannot be C809. Rule PH27 (Code primary site to unknown primary site (C809)) confirms there must be no evidence of neoplasm in the lymph nodes, and that, "If lymph nodes are involved, see Rule PH22," which indicates we are to code primary site to C77.9.

- For example, if diffuse large B-cell lymphoma is found in the femur and in the soft tissue of the anterior chest wall but all CT scans are negative for lymphadenopathy, and the physician did not identify the primary site, we are to code the Primary Site field to C80.9 (Unknown primary site). Diffuse large B cell lymphoma can be either nodal or extranodal. This case is likely extranodal because there is no evidence of lymph node involvement. Given there are multiple areas of extranodal involvement and the extranodal site of origin is unknown, code the Primary Site to C809.

  The rule, that allows us to use the C80.9 site code for lymphomas, took effect with the implementation of the ICD-O-3 and with cases diagnosed on or after 1/1/2001.

Conclusion

The incidence of extranodal disease is rising in the United States and is suspected to be due to various factors such as human immunodeficiency virus (HIV), indolent viral infections and increasing use of immunosuppressive therapy. There are also studies evaluating whether there are distinct race-specific patterns in histology- and site-specific incidence of extranodal non-Hodgkin lymphoma associated with racial differences in risk factor exposure and/or genetic predisposition. Given the interest in the rise in incidence of extranodal lymphomas, it is very important to identify and correctly code the primary site for lymphomas in order for researchers to be able to more accurately identify the differences in nodal and extranodal presentations of lymphoma and to determine reasons why this is occurring.

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Coding Clinical Size for Breast Primaries

Jessica Liang

Prior to 2018, the AJCC Staging system for breast was primarily based on tumor size, lymph node status and whether the cancer spread to other parts of the body beyond the breast. The updated staging system for breast cancer now also includes an assessment of tumor grade, hormone-receptor status, HER2 status, and possibly Oncotype DX test results. While the addition of more cancer characteristics has made determining the stage of a breast cancer more accurate, tumor size continues to be not only an important part of staging these cases, but it is highly related to a patient's prognosis (chance for survival).

Coding clinical size can be confusing. This is especially true for breast given the number of ways a clinician or radiologist describe the neoplasm. Per the SEER Program Coding and Staging Manual 2018, the clinical size data item "records the size of a solid primary tumor before any treatment" (SEER Program Coding and Staging Manual 2018 Section IV Description of This Neoplasm, Tumor Size--Clinical). For breast primaries, the clinical size will generally be recorded from imaging (whichever imaging study documents the largest tumor size, unless the physician specifies the imaging study that is considered most accurate), followed by the size noted on a physical exam. However, confusion can arise when we have to consider how the clinician or radiologist refers to the malignancy because only certain terminology is considered acceptable in reporting the tumor's size.

We should only be coding size when a suspected malignancy is identified, and the terminology used is unambiguous. On imaging, the suspected malignancy can be described as a "mass," "tumor" or "neoplasm." When the radiologist describes the size of the area of interest with one of these unambiguous terms, we may accept it as the clinical tumor size.

It is also acceptable to code a size when the abnormality seen on a scan is specifically referred to as a cancer. For instance, a radiologist may refer to the abnormality as a "2 mm cancer" or a "2 mm focus of cancer." Size is able to be coded from this information because the radiologist has unambiguously described the area of interest. Overall, such terminology is more reliable, because it is not ambiguous as to whether it is considered cancerous.

Calcifications seen on imaging may be benign or associated with malignancy. Calcifications (NOS) are not a reliable measurement of the tumor because they are not exclusively associated with malignancy. The size of calcifications on imaging should not be used to code the clinical tumor size unless the radiologist or clinician specifically indicates the calcifications are malignant.

In addition, we should not be coding clinical size based on the aggregate or a span of separately sized tumors identified on imaging. Again, this would not be an accurate representation of the tumor size. We should only be capturing the size of the largest described tumor on imaging. In addition, the SEER Manual instructs us to only code the size of the largest tumor when tumors are multi-focal.

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Accurately coding the clinical size can be confusing if we don't know what terminology is ambiguous. Table 1 specifies the acceptable terms that can be used to code the clinical tumor size and unacceptable ones that are not to be used to code this field. Being familiar with the descriptive terms allowed to be used to code this field will improve the accuracy of this data item.

Table 1
Acceptable/Unacceptable Terms Used to Code Tumor Size

Acceptable Terms	Common Unacceptable Terms Used To Code Size in Error
Cancer; carcinoma; certain terms attributed as "cancer"	Calcifications
Lesion	Cyst
Mass	Enhancement, NOS
Malignancy	Focus, without mention of "cancer"
Neoplasm	Span/aggregate of tumors
Tumor	Ulcer

In summary, the key to accurately coding clinical size for breast cancer cases is for us to remember not only the allowable reports from the medical record that can be used to determine size and the priority order associated with those reports, but to learn the acceptable and unacceptable terms often used in those reports to reference the abnormal clinical findings and whether or not those descriptions can be used to code the clinical size for breast cancer cases.

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