NATIONAL INSTITUTES OF HEALTH
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SURVEILLANCE, EPIDEMIOLOGY AND END RESULTS (SEER) PROGRAM

2007 Multiple Primary and Histology Coding Rules

"Beyond the Basics" Breeze Sessions

Lung

June 15, 2007

Slide 1

Welcome to the 2007 Multiple Primary and Histology Coding Rules, "Beyond the Basics" presentations.

Slide 2

In this session we will do training on some key points in the Lung Multiple Primary and Histology Coding Rules.

Slide 3

We will do an overview of:
- lung histology
- the Lung Histology Chart
- combination codes
- particular problems people have encountered using the Lung Multiple Primary Rules
- questions that have arisen in using the Lung Histology Coding Rules

We are also doing one practice case in this session. Additional cases are posted on the Website with answers and rationale. You can go to the Website and do the cases at your leisure. There will be no recorded Practicum for this "Beyond the Basics" Breeze Session.

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In talking about the histology for Lung, one of the problems people have with the rules relates to cases with a Fine Needle Aspiration (FNA). In a FNA there is only a small amount of tissue, which makes it difficult for the pathologist to make a specific histologic diagnosis; the histologies are usually non-specific in a FNA sample. Pathologists need more tissue to make a more definitive diagnosis.

With biopsies and resections there are two specific issues with lung tumors. One issue is that lung tumors tend to have large areas of necrotic cells. Dead cells, or necrotic cells, do not provide much information so if the majority of the tumor is necrotic you will see a non-specific diagnosis. If there are areas in a lung tumor that are necrotic, in the microscopic description you will likely see information about a non-specific histology from the necrotic areas and you may also see more specific information which the pathologist found from the non-necrotic areas. This is common with lung tumors.

Many lung tumors are undifferentiated or have large areas that are undifferentiated. In other words, those tumors have large areas that are bizarre cells that no longer resemble the parent cell. The pathology from undifferentiated tumors will usually be non-specific. Again, you may have an entire tumor that is undifferentiated and you will have a non-specific histologic type for that tumor. Or, you may have an area of the tumor that is undifferentiated and in the microscopic description you might see terms that are non-specific from the undifferentiated area as well as more specific information found from cells in another more differentiated area of the tumor.

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Another problem we see with lung cancers is the use of the words "mass, nodule and lesion." For casefinding and for staging, there are very specific rules that tell you when to code a mass, a nodule and a lesion and when those should not be coded. For the purposes of these rules, we use those terms interchangeably. On the whole, the term "mass" more frequently describes cancer and the words "nodule" or "lesion" are pretty interchangeable and may or may not describe cancer. Again, for the purposes of these rules only, we use the words "mass, nodule and lesion" interchangeably.

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To give you an overall perspective on lung cancer, of all the nodules and lesions in a lung, 50-60% are benign. The chest x-ray is a poor indicator of whether or not a lesion is benign or malignant. The PET scan is the best indicator. You will find rules in the Lung (Site-specific) Rules that tell you the order in which to use information from clinical tests when necessary. Please pay close attention to that information because those rules organize these clinical tests in a hierarchical order telling you which tests will provide the most definitive diagnosis.

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Next, we are going to look at the Lung Cancer Histology "Tree." If you look at the "tree" in your MP/H Rules Manual you will see that we are starting at the top of the tree at the right hand branch. The purpose of this review is to provide an understanding of how the specificity of these histologies progresses from the top to the bottom of the tree. This review will also explain the rationale behind adding Chart 1 to these rules; Chart 1 does differ in some respects from the numeric code order in the ICD-O-3. Finally, this review will teach you how to use these Charts both in the Lung rules and in the other site-

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specific MP/H rules. We will teach you the terminology used in the rules such as "branches" on the Charts.

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If you look at the right hand side of the tree, the first entry is "Malignant neoplasm, NOS." Malignant neoplasm, NOS means there are malignant cells, i.e. they have the capability of invading or extending. This is a general term with little specific information. You will see this diagnosis for example, when a bronchial brushing is done. This is the least specific diagnosis.

Notice that when we say there are malignant cells, we do not know if they are carcinoma, sarcoma, melanoma or any other specific type of neoplasm.

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Now please look at the second entry on the Chart: "Carcinoma, NOS; Carcinoma, undifferentiated NOS; and Carcinoma, anaplastic, NOS." Those are codes 8010, 8020 and 8021. The only thing we knew before was that we had cells with the capability of invading or extending. Now with this tier of terms on the Chart we know those cells are epithelial and we know that this excludes sarcoma, lymphoma and melanoma; it is a carcinoma. This is, therefore, a more definitive diagnosis than "Malignant neoplasm, NOS."

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The next entry on the tree is "Non-small cell carcinoma." Non-small cell carcinoma is actually a default code. It is not an actual histology. Many pathologists try to distinguish small cell cancers and non-small cell cancers due to the differences in treatment. The clinical physician has to know this information. The only thing this diagnosis (i.e. non-small cell carcinoma) tells you is that it is carcinoma and that it is not small cell carcinoma. So a default code was added to the ICD-O-3 for non-small cell cancer. The fact is that the pathologist cannot determine from the sample provided whether this is a squamous cell, adenocarcinoma or large cell. The type of cancer is not known; this is a generic default code used when there is inadequate tumor tissue to decide upon a more specific histology.

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Beneath that code of non-small cell on the tree we see "Sarcomatoid carcinoma." Sarcomatoid carcinoma is frequently misunderstood. It means there is a sarcoma-like differentiation. It is **not** a sarcoma; this is a carcinoma.

For sarcomatoid, the pathologist has a little more information:
- there is a sarcoma-like differentiation
- a sarcomatoid carcinoma contains spindle cells and/or giant cells.

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When you see a pathology report that talks about sarcomatoid carcinoma it is not unusual to see a reference to spindle cell and/or to giant cell but you don't code it to spindle cell carcinoma or to giant cell carcinoma; these are features of sarcomatoid carcinoma.

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We are next going to talk about pleomorphic carcinoma, which you see under sarcomatoid on the tree. Pleomorphic carcinoma is similar to sarcomatoid. It, too, is a carcinoma and contains spindle cells and/or giant cells as part of its make-up. The difference between pleomorphic and sarcomatoid is that pleomorphic is poorly differentiated. Poorly differentiated cancers have a worse prognosis. Therefore, if you have the term sarcomatoid and the term pleomorphic, you want to code to the pleomorphic carcinoma because it is poorly differentiated by definition.

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The next entry on the tree under "Pleomorphic" is "Large Cell Carcinoma, NOS," which is another term that is commonly misunderstood. Large cell carcinoma is a specific histologic type. It is undifferentiated and therefore it falls under the poorly differentiated pleomorphic carcinoma. A feature of large cell carcinoma is that you cannot identify either squamous or glandular (adeno) differentiation in this type; that is important to notice.

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Under "Large Cell Carcinoma" on the tree you will see "Adenocarcinoma, NOS." The histology "Adenocarcinoma, NOS" has more information than the terms on the tree branches above it. With adenocarcinoma we know this is an epithelial carcinoma with either glandular differentiation or mucin production. As you can see, we are getting more information as we go further down the Chart.

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On the right side of the tree branch horizontally across from Adenocarcinoma, you will see "Squamous Cell Carcinoma, NOS." Squamous Cell Carcinoma again is an epithelial cell carcinoma that has keratinization and/or intercellular bridges. Notice that Adenocarcinoma and Squamous Cell Carcinoma are on a horizontal line on the Chart; that's because the tree branches right here. When we started with Large Cell we did not know if we had glandular differentiation or squamous differentiation. Now we branch into those two very different histologic types. This is a branch. One branch starts the Adenocarcinoma and all of the more specific Adenocarcinoma types; the other branch is Squamous Cell and all of the more specific Squamous Cell types. When we talk about a branch of the tree, we are talking about the fact that when we got more specific information it did not go in a straight line as it did going from Pleomorphic to Large Cell. We found two cancers that were histologically very different. The Rules will say: Are these cancers on the same branch? The answer is, "No. The branch comes over and down

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for adenocarcinoma. It comes over and down for squamous cell carcinoma. They are different branches of the histologic tree." This is how you use the word "branch" in the Histology Coding Rules.

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I want to remind you that when coding lung cancers the most common histologies you will see are:
- squamous cell (epidermoid)
- adenocarcinoma
- large cell carcinoma

That's why we gave you Chart 2 which gives you specific instructions on how to code cancers when you don't have the unusual histologies shown on Chart 1. Don't be afraid to use Chart 2. Many times Chart 2 will have all the histologies you need to code the particular case on which you are working.

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In looking at the Chart on the Non-Small Cell side (on the right side of the tree) we see a specific entry for Large Cell Neuroendocrine Carcinoma--8013. This particular neuroendocrine carcinoma has a histology code of 8013 because it is part of the Large Cell group but it descends from Non-Small Cell. Note that when we go to the Small Cell part of Chart 1 (on the left side of the Chart) we also see Neuroendocrine Carcinoma, NOS, code 8246. This is a different histology code and it represents an extremely different histology. You need to be aware that there are two neuroendocrine entries: one is a Large Cell Neuroendocrine Carcinoma (8013) directly under Large Cell Carcinoma, NOS (8012); the other is Neuroendocrine CA, NOS (8246) which is on Chart 1 horizontally across from Non-Small Cell CA (8046). One clue is when you look at the pathology report it says neuroendocrine you will see terms in the microscopic description talking about a Large Cell or a Pleomorphic or perhaps even a Non-Small Cell Carcinoma; in this example you will know from Chart 1 that this is a Large Cell Neuroendocrine Carcinoma and you won't miscode it.

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Now we will "hop" to a different branch of the tree.

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We will begin on the Small-Cell box on Lung Chart 1. You will see "Neuroendocrine Carcinoma, NOS" on the box just above "Small Cell Carcinoma, NOS." Note that a low Grade Neuroendocrine Carcinoma is a Carcinoid. Please be aware that if you have a pathology report that says, "Neuroendocrine Carcinoma, Grade I" it should be coded to a Carcinoid. Also be aware that this only applies to **low Grade Neuroendocrine tumors**; this does **not** apply to all Neuroendocrine Carcinomas.

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Under Neuroendocrine Carcinoma, NOS (8246) we have two main entries: Carcinoid, NOS and Small Cell, NOS. These entries vastly differ from each other. Small Cell Carcinoma has a poor prognosis while Carcinoid has an excellent prognosis. When coding these neuroendocrine carcinomas it is important to try to differentiate between the Small Cell Carcinomas and the Carcinoids.

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Neuroendocrine CA, NOS on the left side of Chart 1 are the Carcinoid branch of Neuroendocrine tumors. Some specific types of Neuroendocrine on this branch of the tree are "Carcinoid, NOS" (the subtype of this is "Atypical Carcinoid"); "Combined Small Cell" and "Small Cell Carcinoma, NOS"—all on the same horizontal line on Chart 1 under Neuroendocrine CA, NOS. These are three very distinct divisions of Neuroendocrine tumors on the small cell side of the Chart.

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We will next discuss "Table 1: Combination/Mixed Codes for Lung Histologies."

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The Combination/Mixed Codes Table is important for lung and we want to make sure everyone knows how to use this Table. We have received many questions about this Table. We will go through some examples of Table entries to illustrate how to use the Table.

You will see a Table entry such as "Small Cell Carcinoma AND one histology in Column 2." There is a Note: "**Diagnosis must be Small Cell CA (NOS), not a subtype**." That is a very important Note; pay attention to it. The diagnosis must specifically say "Small Cell Carcinoma;" it cannot say, for example, "Fusiform." You need to know that you must have that exact histology when you see a **Note** such as this one. When you do not see the **Note** you can use any of the subtypes.

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The entry on this slide is a bit different from the previous example because it simply says: "Squamous Cell Carcinoma AND one of the histologies in Column 2." This is an example that does not have a Note saying to only use Squamous Cell Carcinoma. This means you can use Squamous Cell Carcinoma and any of the Squamous Cell Carcinomas shown on the Histology Tree AND the combinations in Column 2 in order to use code 8074—"Squamous cell carcinoma, spindle cell or Squamous cell, sarcomatoid."

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This slide is a third look at how to use Table 1. The required terminology is "Squamous Cell Carcinoma AND Large Cell Non-keratinizing." That means that absolutely nothing else can be used; those terms are "required" in order to use code 8072, Squamous cell carcinoma, large cell, non-keratinizing.

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Another section of Table 1 says you can have "at least two histologies in Column 2." That means you could have Clear Cell Adenocarcinoma and Solid Adenocarcinoma or you could have Solid and Papillary and Clear Cell; in other words, as many as you wish. You must have at least two of the histologies but you may have two or three or four of them. You can then use code 8255. Warning: you **cannot** use code 8255 for any combination of histologies. For lung cancers you must have a combination of at least any two of the histologies in Column 2. You cannot use 8255 for any other combinations of histologies.

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We will talk about a couple of the Lung Multiple Primary Rules about which people have had questions.

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The first question concerns rules M1 and M12. The wording says the case has at least one tumor in one lung and at least two tumors in the contralateral lung. This situation historically has caused problems for registrars; physicians usually biopsy only one of the lesions. Clinically, the physician looks at such a case as a fatal disease. The physician does not think it useful to biopsy more than one tumor in these cases. Clinicians say they treat patients based on the histology of that one tumor. Quality patient care says the patient should not be subjected to a more exhaustive diagnostic workup because there would be no benefit to the patient.

The Commission on Cancer physician consultants to the MP/H Rules Committee and the Physician Advisors to the MP/H Rules Committee advised the Committee that these cases should be abstracted as a single primary using the histology from the single biopsy. They advised us that registrars should not worry about whether or not the un-biopsied tumors are benign or metastatic or a different histology than the biopsied tumor. These cases should be coded as a single primary. They will be treated as a single primary clinically. By adopting this guidance all in the field will treat these cases in the same manner and there will be consistency in coding them as a single primary.

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Rule M6 says that one tumor in each lung is coded as multiple primaries unless they are **proven** to be metastatic. This rule tells you that if there is **one tumor in the right lung and one tumor in the left lung** you treat them as **two primaries** unless you actually have a biopsy that says one of those tumors is metastatic. Again, the Physician Consultants to the MP/H Rules Committee said patients in most of these cases would be treated as though they had multiple primaries. They may have, for example, a wedge biopsy in one lung; they may have a wedge biopsy of the other lung removing both tumors. This is a treatable disease that is most correctly abstracted as two primaries. Each lung should be staged appropriately.

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We will talk a little about some points in using the Lung Histology Coding Rules.

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The first point concerns using rules H1 or H8. Those rules say if there is no pathology report you code from the Medical Record (MR). A hierarchy is provided in the rules showing the priority order of documents from which to code. Those rules are good; they tell you where to go to get the information about the histology but they don't tell you how to code the histology. Once you have used rule H1 or rule H8 you need to know how to code the histology. The same problem occurs in using rules H2 or H9; they tell you if your only information is from a metastatic site, for example, biopsy of a supreclavicular lymph node for a lung primary, then you would code the histology from the metastatic site. But neither of these two rules tells you how to code the histology.

When you use these rules you need to go back through the Histology Coding Rules and use the appropriate Module either Single Tumor Module or Multiple Tumors Module and make another pass through the rules to choose the correct histology code. This is a new concept. We will be writing more specific instructions when we do the revisions to the MP/H Rules. In the meantime, we are teaching you that when you use rules H1, H8, H2 or H9 those rules tell you where to get the information about the histology. You then go back through the Histology Coding Rules a second time using the appropriate module for either single or multiple tumors to determine which histology you should code.

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Finally, we are going to do a Practice Case. The case reads:

Final Diagnosis:
Lung (right lower lobectomy): 8.0 cm moderately differentiated **acinar type adenocarcinoma**, bronchial and vascular margins of resection negative for tumor, peribronchial lymph nodes negative for tumor, separate 1.0 cm **well differentiated neuroendocrine carcinoma (carcinoid tumor)**.

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First, you must answer the question: Is this a single or multiple tumor(s)? The pathology report clearly states that there are two tumors so you go to the Multiple Tumors Module and start with Rule M3.

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You would stop at Rule M11 that says if the tumors have ICD-O-3 histology codes that differ at the first, second or third character they are multiple primaries. In this case the histology codes differ at the second and third characters: 8550 and 8240. The instructions from M11 tell you that this is a multiple primary.

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Let's code the histology. The histology for primary #1 is Acinar type adenocarcinoma. This is a single tumor and histology so we will go to the Single Tumor Module and start with Rule H1.

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You will stop at Rule H3 or H5, which tells you to use Chart 1. You will decide that Acinar carcinoma is a specific type of Adenocarcinoma and you will use code 8550/3.

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For primary #2 the diagnosis is Neuroendocrine (Carcinoid). This is a single tumor. You will use the Final Diagnosis to code the histology. You will go to the Single Tumor Module starting with rule H1.

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You will stop at Rule H5 and you will refer to Chart 1. On Chart 1 you will see that a Carcinoid is a descendent of Neuroendocrine and you will code 8240/3.

This is the end of the Practice Case. Ten Practice Cases are posted on the Website with the recorded Breeze Sessions. You are welcome to code those ten cases. You will find the answers and rationale posted with each case.

Thank you for attending this Beyond the Basics Breeze Session on key points in the Lung Multiple Primary and Histology Coding Rules.