20160076

References

Source 1: **WHO Class CNS Tumors**

pgs: **33**

Question

MP/H Rules/Histology--Brain and CNS: What is the histology code for a tumor originating in the cerebellum and extending into the fourth ventricle described as a glioblastoma with primitive neuroectodermal tumor component (WHO Grade IV)?

Answer

The WHO Classification of CNS tumours lists glioblastoma with primitive neuroectodermal tumor component as a subtype of glioblastoma and assigns 9440/3. Also referred to as glioblastoma with a primitive neuronal component.

Date Finalized

12/14/2016

20160074

References
Source 1: **WHO Class Breast Tumors**
pgs: **62**

Question
MP/H Rules/Histology--Breast: How should histology be coded for a breast primary with resection final diagnosis of "Ductal carcinoma with neuroendocrine features?" See Discussion.

Discussion
Should the histology for "Ductal carcinoma with neuroendocrine features" be coded to 8500 (Ductal carcinoma, NOS) or 8574 (Adenocarcinoma with neuroendocrine differentiation)?

Answer
Code the histology to 8574/3 for Ductal carcinoma with neuroendocrine features.

Ductal carcinoma is also called "invasive breast carcinoma of no special type." WHO classifies Invasive breast carcinoma with neuroendocrine differentiation as 8574/3.

Date Finalized
12/14/2016

20160071

References
Source 1: **SEER Summary Stage 2000**
pgs: **172-175**

Question
SEER Summary Stage 2000--Melanoma: Can Clark's level classification still used to Summary Stage melanoma? It was previously used by AJCC TNM, but was not included in the 7th edition. I see it is still listed in the CAP protocols for melanoma.

Answer
Clark's level can be used to assign in situ, localized or regional summary stage.

If there is a discrepancy between the Clark's level and the pathologic description of extent, use the higher Summary Stage code.

Date Finalized
12/14/2016

20160070

References
Source 1: **WHO Class H & N Tumors**
pgs: **19**
Notes: **2005**

Question
Primary site/MP/H Rules/Histology: What is the appropriate site and histology code for a tumor described as a "Large mass in suprasellar cistern encroaching into sphenoid & ethmoid sinuses", with the pathology described as "INI-1 deficient sinonasal undifferentiated carcinoma"? Of note, this patient has a history of a pituitary adenoma, resected overseas a few months prior to this diagnosis.

Answer
The primary site is unclear. The lesion is intracranial, but this may not be the primary site. In the absence of any additional information, assign C390, 8020/3. According to WHO, sinonasal undifferentiated carcinoma can involve the nasal cavity, maxillary antrum, and/or ethmoid sinus.

SMARCB1 (INI-1) is a tumor-suppressor gene located on chromosome 22q11.2. Tumors that showed loss of expression were SMARCB1-deficient tumors which are characterized by nests, sheets, and cords of cells without any histologic evidence of specific (e.g., squamous or glandular) differentiation.

Date Finalized
12/14/2016

20160069

References
Source 1: **WHO Class Female Reproductive Organs**
pgs: **142-145; 232-233**
Notes: **2014**

Question
Reportability/MP/H Rules/Histology: Do we interpret Endometrial stromal sarcoma / Undifferentiated stromal sarcoma as low grade / high grade still in order to code the correct histology?

And for VIN, if a pathology report comes across as undifferentiated VIN, do we report it as VIN 3, even if that terminology isn't there?

Answer
There are three reportable endometrial stromal tumors: Low-grade endometrial stromal sarcoma (LGESS), 8931/3; High-grade endometrial stromal sarcoma, 8930/3; and Undifferentiated uterine sarcoma, 8805/3.

VIN 1 and VIN 2 are **not** reportable.

VIN 3 is a synonym for High-grade squamous intraepithelial lesion, 8077/2. It is reportable.

WHO includes Differentiated-type vulvar intraepithelial neoplasia, 8071/2. It is reportable. According to WHO, the differentiation refers to abnormal keratinocyte differentiation, which is why it is classified as 8071.

WHO does not list "undifferentiated VIN."

Date Finalized
12/14/2016

20160068

References: none listed

Question
Reportability--Brain and CNS: Are sphenoid wing meningiomas reportable? See discussion.

Discussion
It's my understanding that true intraosseous meningiomas are very rare. It's also my understanding that cranial meninges DO cover the sphenoid wing, so I'm wondering if it's possible to have a meningioma of the sphenoid wing on imaging that arises from the meninges NOT the bone. Is that the deciding factor on reportability? It's been suggested to me that meninges cells do lie within the bone, but again if a meningioma is described as being located at the sphenoid wing on imaging, without bone involvement - and no surgery is performed - I do not understand why it is specifically excluded as non-reportable.

Answer
Note: This answer **updates previous answers** which have been removed from the SEER Inquiry System.

Intraosseous meningiomas are not reportable. You are correct, these are rare meningiomas originating in bone. The term "sphenoid wing meningioma" is sometimes used for an intraosseous meningioma of the sphenoid bone. Yes, it's possible to have a meningioma of the sphenoid wing on imaging that arises from the meninges NOT the bone. Read the available information carefully. When the site of origin is described as "along the sphenoid wing" or "overlying the sphenoid wing" report the meningioma. These descriptions indicate that the meningioma originates from the meninges covering bone rather than the bone itself. Meningioma arising in bone is rare enough, that when present, we would expect it to be clearly stated as such. In the absence of a statement indicating origin in bone, the meningioma is most likely arising from meninges covering the bone.

Date Finalized
11/29/2016

20160067

References
Source 1: **2007 MP/H Rules**
Notes: **Other Sites**

Question
MP/H Rules/Histology--Skin: What histology code and MP/H Rule apply to a skin primary with the final diagnosis, Basaloid carcinoma with squamous and neuroendocrine differentiation? See Discussion.

Discussion
The patient had an upper arm shave biopsy with final diagnosis of basaloid carcinoma with squamous and neuroendocrine differentiation. The pathologist also comments: Based on the morphologic features and immunophenotype, a basaloid carcinoma with squamous and neuroendocrine differentiation derived from non-Merkel cell origin is favored. However, we cannot completely exclude a combined Merkel Cell Carcinoma with both squamous and neuroendocrine differentiation with negative Merkel cell polyomavirus. Given the difference in biological behaviors of these two differential diagnoses, close clinical follow-up is recommended. Further resection was negative for residual malignancy.

Would SINQ 20150033 apply, thus resulting in final histology of carcinoma with neuroendocrine carcinoma (8574/3)?

Answer
Assign 8574/3 according to Other Sites rule H17 for basaloid carcinoma with squamous and neuroendocrine differentiation.

There is no combination code that includes basal cell, squamous, and neuroendocrine. We can combine basal cell with squamous, 8094/3, or carcinoma with neuroendocrine differentiation, 8574/3. Rule H17 directs us to assign the higher code, 8574/3.

Date Finalized
12/14/2016

20160066

References
Source 1: **Subject Matter Expert**
Source 2: **ICD-O-3**

Question
MP/H Rules/Histology--Breast: What histology code and MP/H Rule applies to the Histologic Type of "invasive ductal carcinoma with metaplastic stroma" for a single breast tumor? See Discussion.

Discussion
The patient had a partial mastectomy with final diagnosis of invasive ductal carcinoma with metaplastic stroma. Knowing that metaplastic breast carcinoma has a worse prognosis than other types of breast cancer, is metaplastic stroma a synonym for metaplastic carcinoma when used in this context?

Answer
Code to metaplastic carcinoma, 8575/3. According to our expert pathologist consultant, "The term 'metaplastic stroma' implies that at least a portion of the carcinoma has undergone a 'metaplastic' change from epithelial in appearance to 'stromal' in appearance. I assume this is what CAP means by 'Invasive mammary carcinoma with matrix production,' which the WHO equates to metaplastic carcinoma."

Date Finalized
12/14/2016

20160065

References
Source 1: **WHO Class Lung Tumors**
Notes: **3rd and 4th editions**

Question
MP/H Rules/Histology--Lung: What histology code and MP/H Rule applies to the Histologic Type described as adenocarcinoma, mixed invasive mucinous and non-mucinous which involves multiple lung tumors present in a single lobe? See Discussion.

Discussion
The patient had a lower lobectomy with final diagnosis of adenocarcinoma with the following features: Tumor Focality: Multiple separate tumor nodules in same lobe; Tumor Size: 2.6 cm, 0.7 cm, 0.3 cm and 0.1 cm in greatest dimension; Histologic Type: Adenocarcinoma, mixed invasive mucinous and non-mucinous adenocarcinoma; Histologic Grade: Moderately differentiated.

Answer
Assign histology code 8254/3.

The 2007 MP/H Lung rules do not include coding guidelines for mixed mucinous and non-mucinous tumors. Lung Table 1 (in the Terms and Definitions, pages 37-38, http://seer.cancer.gov/tools/mphrules/mphrules_definitions.pdf) is very specific about which histologies can be coded to mixed adenocarcinoma (8255/3). Mucinous is not included per the note at the end of Table 1. Per WHO 3rd and 4th Ed Tumors of the Lung, mixed mucinous and non-mucinous tumors of the lung are classified as 8254/3. Mixed invasive mucinous and non-mucinous adenocarcinoma is a synonym for BAC, mucinous and non-mucinous.

Date Finalized
10/31/2016

20160064

References

Source 1: **WHO Classification of Tumors of the Urinary System and Male Genital Organs**
pgs: **164-165**
Notes: **4th edition**

Question

Behavior--Prostate: What is the correct behavior of intraductal carcinoma from a prostate biopsy with a Gleason score 4+4=8. While highly aggressive, but not suggestive of invasion, coding behavior as /2 seems inappropriate.

Answer

WHO classifies intraductal carcinoma of the prostate 8500/2. According to WHO, "the hallmark of intraductal carcinoma of the prostate is a proliferation of prostate carcinoma cells that is within and may significantly expand the native prostatic ducts and acini, with the basal cell layer at least partially preserved." Further, differentiation between intraductal carcinoma and infiltrating high-grade carcinoma of the prostate may require basal cell stains. Under Prognosis, WHO states: "…intraductal carcinoma of the prostate on prostate biopsies is often associated with high-grade cancer (with a mean Gleason score of 8) …."

So while it may seem counter-intuitive, assign behavior code /2 when the diagnosis is intraductal carcinoma of the prostate.

Date Finalized

10/31/2016

20160061

References
Source 1: **Subject matter expert**

Question
Reportability/Behavior--Small intestine: Is a carcinoid tumor, described as benign, reportable? See Discussion.

Discussion
A segmental resection pathology report states "benign mucosal endocrine proliferation consistent with a 0.3 cm duodenal carcinoid tumor." The diagnosis comment further states, "the separate small endocrine lesion is histologically benign, consistent with a 3 mm carcinoid tumor." This seems to be an example of a description of a microcarcinoid tumor referenced in SINQ 20160011. However, in this new case the pathologist specifically states the tumor is benign.

The WHO definition of microcarcinoid indicates this is a precursor lesion, which seems to indicate it is not malignant. However, SEER's previous answer stated we should report these tumors because the ICD-O-3 definition of carcinoid is 8240/3. Do you think that the mention of the term "benign" in the pathology report is actually related to the size of this lesion? Is the reference to benign mucosal endocrine proliferation referring to the WHO classification (making the case reportable as stated in SINQ 20160011), or is this a situation in which we should apply the Matrix Rule and the case is nonreportable?

Answer
This carcinoid tumor, described as benign, is not reportable. According to our expert pathologist consultant, this case is not reportable because the pathologist uses "benign" to describe the mucosal endocrine proliferation and based on that, the neuroendocrine cell proliferation is hyperplasia/benign - not reportable.

Date Finalized
10/31/2016

20160057

References
Source 1: **2007 MP/H Rules**

Question
MP/H Rules/Histology--Prostate: What is the histology code for a prostate case whose histology reads "adenoca with mixed ductal and acinar variants?

Answer
Assign 8523/3.

The 2013 revision to ICD-O-3 has a new code for mixed acinar ductal carcinoma; however, this new code will not be implemented in the U.S. until 2018 or later. Page 7 of the Guidelines for ICD-O-3 Update Implementation document released by NAACCR 1/1/2014 instructs us to use 8523/3 in the meantime.

Date Finalized
11/23/2016

20160056

References
Source 1: **WHO Class Male Genital Tumors**

Question
MP/H Rules/Histology--Testis: How should histology be coded for a testicular primary with a combination of teratoma, yolk sac tumor and embryonal carcinoma? See discussion.

Discussion
Patient had a radical orchiectomy with the final diagnosis of "Mixed germ cell tumor with the following features -- histologic type: Mixed germ cell tumor (teratoma 50%, yolk sac tumor 25%, and embryonal 25%)."

Answer
Assign 9085/3. Code this combination of teratoma, yolk sac tumor, and embryonal tumor in the testis to mixed germ cell tumor (9085/3) based on the WHO Classification of Tumors of the Male Genital Organs.

Date Finalized
10/06/2016

20160052

References
Source 1: **SEER Summary Stage 2000**

Question
Summary Stage 2000--Lymphoma: How is SEER SS2000 coded for an ocular adnexal lymphoma when it extends from the primary site to adjacent sites that are still orbital structures? See Discussion.

Discussion
In this case, the lymphoma arose in the posterior orbit and the primary site was coded as C696 (orbit, NOS). The mass directly extended to at least one "adjacent" site, the lacrimal gland. Should SS2000 be coded to 1 (localized) or 5 (regional, NOS) when an ocular adnexal lymphoma arises in the posterior orbit and extends to involve the lacrimal gland? Although both the posterior orbit and the lacrimal gland are parts of the orbit, they have separate ICD-O-3 topography codes. Should extension to multiple sites within the orbit be classified as localized disease?

The issue is what constitutes "adjacent" structures for a tumor that arises in the orbit. In an article published by the Indian Journal of Ophthalmology it states, "According to the Ann-Arbor staging system, lymphoma confined to the orbit is designated as Stage I, involvement of adjacent structures (sinuses, tonsil and nose) makes it Stage II." Does SEER agree with this definition of "adjacent" structures? Or are the lacrimal gland, ciliary body, retina, conjunctiva and/or choroid "adjacent" structures for a lymphoma stated to arise in the posterior orbit?

Answer
Assign SEER SS2000 code 5 (Regional, NOS) for a lymphoma of orbit extending to lacrimal gland. In SEER SS2000, this is Stage IIE: Direct extension to adjacent organs or tissues.

Date Finalized
12/14/2016