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■ ORIGINAL ARTICLE ■

A Proposal for Creating a Guideline for Cancer Registration of the Fibromatosis, PEComa Group, Malignant Lymphoma *In Situ* and Dendritic Cell Tumors (III)

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Background: Understanding the biologic behavior of a tumor is a prerequisite for tumor registration code assignment. The aim of this report was to propose appropriate behavior codes of the International Classification of Disease Oncology 3 (ICD-O3) to rare, yet pathologically interesting hematopoietic and soft tissue tumors. **Methods:** The Study Group for Hematopathology, the Bone and Soft Tissue Pathology Study Group, and the Cancer Registration Committee prepared the questionnaire containing provisional behavior codes of selected diseases. **Results:** *In situ* lesions of mantle cell and follicular lymphomas, dendritic cell tumors, and neoplasms with perivascular epithelioid cell differentiation (PEComa), not otherwise specified were classified as malignant (-/3). The fibromatosis group, with the exception of lipofibromatosis, was proposed as benign (-/0). Lipofibromatosis and several diseases that belong to the PEComa group were proposed as uncertain malignant potential (-/1). For the hematologic and soft tissue tumors, 274 and 288 members of the Korean Society of Pathologists, respectively, provided opinions through questionnaire, and most responders showed agreement with the provisional behavior code proposed. **Conclusions:** The determination of behavior codes for the rare diseases described in this study, especially those of the PEComa group or malignant lymphoma, could be viewed as impractical and premature, but this study provides the basis for future research on this topic.

Key Words: ICD-O3; Behavior code; Hematologic malignancy; Soft tissue neoplasms

Standardization of the pathologic terminology and a universal understanding of the biologic behavior of diseases are fundamental for cancer registration coding.[1] International Classification of Disease for Oncology 3 (ICD-O3) provides guidelines

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for tumor or cancer registries for coding the site (topography) and the histology (morphology) of neoplasms, which are usually obtained from a pathologic report. It makes a multi-axial classification including the site, the morphology, the behavior, and the grading of neoplasms possible. The topography axis uses the ICD-10 classification of malignant neoplasms for all types of tumors. The morphology axis provides five-digit codes ranging from M-8000/0 to M-9989/3. The first four digits indicate the specific histological term. The fifth digit after the slash (/) is the behavior code, which indicates whether a tumor is malignant, benign, *in situ*, or uncertain whether benign or malignant (Table 1).[1] The Cancer Registration Committee of the Korean Society of Pathologists (KSP) planned to assign the biologic behavior codes of ICD-O3 for the tumors that are currently disputed regarding the name of diagnosis and behavior code. The committee published the guidelines for gastrointestinal tumors in 2008,[2] and for microinvasive tumors of the ovary and breast in 2012.[3]

The Study Group for Hematopathology and the Bone and Soft Tissue Pathology Study Group of KSP participated in this work in 2009 and 2010, respectively. In the field of hematopathology, the publication of a new edition of the World Health Organization (WHO) classification in 2008, not only introduced new disease entities, but also altered some preexisting behavior codes. In addition, some ambiguous behavior codes from a previous edition still exist in this new edition.[4,5] The WHO classification of bone and soft tissue also contains several diseases to which behavior codes in ICD-O3 were not assigned.[6] These diseases share a common characteristic in that they are rare diseases and their biologic behavior and characteristics are uncertain as to whether they are benign or malignant, or neoplastic or not.[6]

Although some tumor types are extremely rare, they have pathologically interesting aspects and should not be ignored. Therefore, it is necessary to consider the biologic behavior of these diseases. With this background, the Study Group for Hematopathology and the Bone and Soft Tissue Pathology Study Group worked independently to propose appropriate behavior codes for their relevant diseases. The purpose of this report is (1) to introduce the works performed by the Study Group for Hematopathology and the Bone and Soft Tissue Pathology Study Group, which includes the selection of diseases and the final analysis of data obtained from the questionnaire, and (2) to propose appropriate behavior codes for selected tumors. Based on this work, we believe that an understanding of *in situ* lesions in malignant lymphoma and histiocytic lesions would be increased in the field of hematopathology. In the field of soft tissue tumors, the diseases selected were mostly benign and rare, but reconsideration and an understanding of the biologic behavior of these rarely seen diseases may provide a baseline and valuable standards for further study in the field of bone and soft tissue pathology.

Table 1. The ICD-O3 system[1]

Two axes of classification of neoplastic diseases	Behavioral codes
Topography (see ICD-10)	/0 benign
Morphology	/1 uncertain (whether benign or malignant) or low malignant potential
8000-8009 Not otherwise specified	/2 *in situ*
8010-8790 Epithelial	/3 malignant
8800-9370 Connective tissue	
9380-9589 Nervous system	
9590-9989 Hematologic	

ICD-O3, International Classification of Disease Oncology 3.

MATERIALS AND METHODS

Selection of tumors

After several conferences and workshops, each study group selected the tumors and determined the provisional behavior codes. The Study Group for Hematopathology selected *in situ* lesions of malignant lymphomas, and histiocytic lesions for discussion. *In situ* lesions included mantle cell lymphoma *in situ* and follicular lymphoma *in situ*, and histiocytic lesions included follicular dendritic cell tumors, interdigitating dendritic cell tumors, and Langerhans cell histiocytosis.

The Bone and Soft Tissue Pathology Study Group initially selected diseases that do not have an ICD-O3 code among the bone and soft tissue tumors included in the WHO classification, including:[6] fibromatosis colli, juvenile hyaline fibromatosis, inclusion body fibromatosis, superficial fibromatosis, lipofibromatosis, hyalinizing spindle cell tumor, neoplasm with perivascular epithelioid cell differentiation (PEComa), and clear cell myomelanocytic tumor among the soft tissue tumors, and aneurysmal bone cyst, fibrous dysplasia, osteofibrous dysplasia, and Erdheim-Chester disease among bone tumors. PEComa were further divided into PEComa, not otherwise specified (PEComa NOS), angiomyolipoma (AML), lymphangioleiomyomatosis (LAM), clear cell "sugar" tumor of the lung (CCST), and clear cell myomelanocytic tumor of the falciform ligament/ligamentum teres (CCMMT). Their biologic behaviors were discussed separately. Among the selected tumors, superficial fibromatosis was excluded from this study due to its obvious benign biologic behavior. Hyalinizing spindle cell tumor was also excluded because it is a variant of low grade fibromyxoid sarcoma, and thus should be regarded as a malignant disease. Under the heading

of AML, we excluded renal AML, because its biologic behavior is well known to be benign even though sarcomatous change is rarely reported.[7] Aneurysmal bone cyst, fibrous dysplasia/osteofibrous dysplasia, and Erdheim-Chester disease were initially included in the discussion, but finally excluded due to well-known benignancy or rarity.

A proposal for provisional behavior codes

The Study Group for Hematopathology determined the codes presented both in the preexisting ICD-O3 and the WHO classification 2008 for the selected tumors so that other pathologists could compare the results, because ICD-O3 does not provide a behavior code for follicular lymphoma *in situ* and mantle cell lymphoma *in situ*, but does provide behavior code -/1 for the histiocytic and dendritic cell neoplasms. This is in contrast to the WHO classification 2008, which considers them as malignant tumors.[5]

In the field of bone and soft tissue tumors, three pathologists classified these tumors on the basis of previous reports and made provisional behavior codes. They prepared materials for the charged parts, and the presentation, the discussion, and the convergence of opinions were conducted during the workshop held within the Bone and Soft Tissue Study Group. Finally, provisional behavior codes were proposed for the selected tumors. Because reports regarding these diseases were insufficient in Korea, we referred to several foreign literature databases and WHO classifications to determine provisional behavior codes.

Preparation of questionnaire and analysis of data

The Cancer Registration Committee held additional discussions regarding provisional behavior codes provided by the Study Group for Hematopathology and the Bone and Soft Tissue Pathology Study Group. The final lists of tumors were determined and the provisional biologic behavior codes for the corresponding diseases were established. The questionnaire was made considering these decisions and distributed to the members of KSP during the Academic Congress of Pathology and Cytopathology. Data were collected and then analyzed.

RESULTS

Provisional behavior codes settled in the workshop

The Study Group for Hematopathology suggested the behavior code -/3 (malignant) for mantle cell lymphoma *in situ* and follicular lymphoma *in situ*. For the histiocytic and dendritic cell neoplasms, the majority of the study group members agreed on the malignant behavior (Table 2).

For the bone and soft tissue tumors, the tumors were categorized as fibromatosis or PEComa in order to avoid the complexities of the classification. Among the fibromatosis category, fibromatosis colli, juvenile hyaline fibromatosis, and inclusion body fibromatosis were described as benign, and given the behavior code -/0. Lipofibromatosis was regarded as a borderline tumor and given behavior code -/1. Among the PEComa cate-

Table 3. Questionnaire for the provision of the behavior codes of soft tissue tumors

Diagnosis	ICD-O3 behavior code	Provision of Bone and Soft Tissue Study Group	ICD-O3 behavior code (opinion of members)[a]	Note
Fibromatosis				
Fibromatosis coli	No	/0		
Juvenile hyaline fibromatosis	No	/0		
Inclusion body fibromatosis	No	/0		
Lipofibromatosis	No	/1		
PEComa				
PEComa, NOS	No	/3		
PEComa, AML[b]	No	/1		
PEComa, LAM	No	/1		
PEComa, CCST	No	/1		
PEComa, CCMMT	No	/0		

ICD-O3, International Classification of Disease Oncology 3; PEComa, perivascular epithelioid cell differentiation; NOS, not otherwise specified; AML, angiomyolipoma; LAM, lymphangioleiomyomatosis; CCST, clear cell "sugar" tumor of the lung; CCMMT, clear cell myomelanocytic tumor of the falciform ligament/ligamentum teres.
[a]"Please write your opinion as /0, /1, or /3 in the blank"; [b]The renal AML is excluded.

Table 2. Questionnaire for the provision of behavior codes of hematologic malignancies

Diagnosis	ICD-O3 behavior code	WHO 4th edition	ICD-O3 behavior code (opinion of members)	Note
Follicular lymphoma *in situ*	No	/3		New disease entity in WHO 2008
Mantle cell lymphoma *in situ*	No	/3		New disease entity in WHO 2008
Follicular dendritic cell tumor	/1	/3		
Interdigitating dendritic cell tumor	/1	/3		
Langerhans cell histiocytosis	/1	/3		

ICD-O3, International Classification of Disease Oncology 3; WHO, World Health Organization.

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Table 4. Results of the questionnaire for hematologic malignancies

Diagnosis	ICD-O3 behavior code (/1)	ICD-O3 behavior code (/2)	ICD-O3 behavior code (/3)	ICD-O3 behavior code (hold off)
Follicular lymphoma *in situ*	43	16	207	8
Mantle cell lymphoma *in situ*	39	16	211	8
Follicular dendritic cell tumor	29	3	241	1
Interdigitating dendritic cell tumor	47	3	222	2
Langerhans cell histiocytosis	63	5	205	1

ICD-O3, International Classification of Disease Oncology 3.

Table 5. Results of the questionnaire for soft tissue tumors

Diagnosis	ICD-O3 behavior code (/0)	ICD-O3 behavior code (/1)	ICD-O3 behavior code (/2)	ICD-O3 behavior code (/3)	ICD-O3 behavior code (no response)
Fibromatosis coli	245	34	1	0	8
Juvenile hyaline fibromatosis	254	25	1	0	8
Inclusion body fibromatosis	257	20	3	0	8
Lipofibromatosis	42	232	3	1	10
PEComa, NOS	0	38	4	232	14
PEComa, AML[a]	10	257	4	6	11
PEComa, LAM	4	260	4	11	9
PEComa, CCST	2	257	5	10	14
PEComa, CCMMT	210	51	5	9	13

ICD-O3, International Classification of Disease Oncology 3; PEComa, perivascular epithelioid cell differentiation; NOS, not otherwise specified; AML, angiomyolipoma; LAM, lymphangioleiomyomatosis; CCST, clear cell "sugar" tumor of the lung; CCMMT, clear cell myomelanocytic tumor of the falciform ligament/ligamentum teres.
[a]The renal AML is excluded.

gory, CCMMT was suggested to be a benign disease and the behavior code -/0 was applied. AML, LAM, and CCST were given behavior code -/1, and PEComa NOS was given behavior code -/3 (Table 3).

Results of the questionnaire

A total of 274 members of the KSP provided their opinions by completing the questionnaire. Of the responders, the following percentages indicated a behavior code of -/3 (malignant) for the respective tumor types: 75.5% (207/274) for follicular lymphoma *in situ*, 77% (211/274) for mantle cell lymphoma *in situ*, 88.0% (241/274) for follicular dendritic cell tumor, 81.0% (222/274) for interdigitating dendritic cell tumor, and 74.8% (205/274) for Langerhans cell histiocytosis. The remaining members listed behavior code -/1 (uncertain malignant potential or borderline tumor) for these tumor types, with some members providing code -/2 (carcinoma *in situ*) and a few others suggesting that we postpone coding (Table 4).

Regarding responses to the questionnaire on soft tissue tumors, 288 members of the KSP provided opinions. For the fibromatosis category, most responders gave behavior code -/0 (benign tumor) for fibromatosis colli, juvenile hyaline fibromatosis, and inclusion body fibromatosis, and -/1 (uncertain malignant potential or borderline tumor) for lipofibromatosis as the provision of the Bone and Soft Tissue Study Group. For the PEComa category, most responders assigned PEComa NOS to -/3 (malignant neoplasm), AML, LAM, and CCST to -/1, and CCMMT to -/0 (Table 5).

DISCUSSION

The Study Group for Hematopathology focused on *in situ* lesions of malignant lymphomas and histiocytic lesions. Mantle cell lymphoma *in situ* and follicular lymphoma *in situ* are defined morphologically when tumor cells are restricted to the mantle zone and intrafollicular area, respectively. However when the cells of these tumors circulate in the body, they should be classified as malignant diseases.[5,8-10] Because the term "*in situ*" can cause confusion not only to pathologists, but also to clinicians, the Study Group for Hematopathology decided to include these lesions in order to clearly indicate that "*in situ* lesions" are malignant.

There were some disputes in the study group regarding histiocytic neoplasms. In the WHO classification 2008, histiocytic and dendritic cell neoplasms are classified as malignant disease with behavior code /3 of ICD-O3.[5] Even before this edition, it has been suggested that Langerhans cell histiocytosis should be considered a malignant tumor because of its risk of multisystem

involvement and refractoriness to treatment,[11] although it is treated as a tumor of uncertain malignant potential.[4] Dendritic cell neoplasms also have morphological obscurity between borderline and malignant tumors, but they were eventually defined as malignant disease without distinction, which was in contrast to the previous edition of the WHO classification. Therefore the study group decided to include these histiocytic and dendritic cell neoplasms in this study because it may be meaningful to compare behavior codes between ICD-O3 and the WHO classification 2008. The study group selected mantle cell lymphoma *in situ*, follicular lymphoma *in situ*, follicular dendritic cell tumor, interdigitating dendritic cell tumor, and Langerhans cell histiocytosis for this study.

In the WHO classification 2001, *in situ* lesion of follicular lymphoma was not mentioned.[4] It was only introduced in the WHO classification 2008 under the name intrafollicular neoplasia "*in situ*" follicular lymphoma, and defined morphologically as architecturally normal appearing lymph nodes. Other lymphoid tissues harboring one or more follicles that overexpress Bcl-2 in centrocytes and centroblasts with or without a monomorphic cytologic appearance was described to be suggestive of follicular lymphoma.[5] The meaning of this tumor has not been completely established yet, but it may represent the tissue counterpart of circulating clonal B-cells possessing *BCL-2* rearrangement,[5,9] or the earliest evidence of true follicular lymphoma that subsequently progress to overt follicular lymphoma.[5,10,12] Mantle cell lymphoma "*in situ*" is even mentioned in the WHO classification 2001, but the term "*in situ*" is only found in the WHO classification 2008.[4,5] By the definition, this tumor is confined to the inner mantle zone of lymphoid tissue.[5,8]

Histiocytic and dendritic cell tumors are classified as borderline tumors and are distinguished from their malignant counterpart sarcoma in the WHO classification 2001. They became evenly classified as malignant tumors without distinction of tumor or sarcoma in the WHO classification 2008.[4,5] Follicular dendritic cell tumor is a rare malignant tumor originating from follicular dendritic cells. It can arise in any organ, but most commonly occurs in the cervical lymph node. They show an indolent clinical course but have frequent local recurrence and distant metastasis.[5,13] Interdigitating dendritic cell tumor is also a very rare disease, and usually occurs as a solitary lesion in the lymph node, although extranodal lesions have been reported.[5,14] Langerhans cell histiocytosis usually occurs as solitary lesion in children. Typical of multisystem lesions, it is refractory to treatment and associated with high mortality.[5,15]

The Study Group for Hematopathology did not propose different behavior codes, but rather decided to provide both codes that are presented in the preexisting ICD-O3 and the WHO classification 2008. The biologic codes presented in the WHO classification 2001 are the same as the codes of ICD-O3. The questionnaire form produced by the Study Group for Hematopathology was presented to the members of the KSP at academic congresses. The results of the questionnaire indicated that, members of the KSP generally agreed on the biological behavior of follicular lymphoma *in situ*, mantle cell lymphoma *in situ*, follicular dendritic cell tumor, interdigitating dendritic cell tumor, and Langerhans cell histiocytosis as malignant disease, and therefore, the Study Group for Hematopathology and the Cancer Registration Committee could propose behavior code -/3 for these tumors. The study group also suggested that the diagnostic names of Langerhans cell histiocytosis, unifocal Langerhans cell histiocytosis, and multifocal Langerhans cell histiocytosis should all be recapitulated under the name of Langerhans cell histiocytosis with biologic behavior code -/3. Howerver, the final acceptance of these provisional codes may require additional studies that include long-term follow-up data, greater accumulation of knowledge, and agreement among clinicians.

Among the bone and soft tissue tumors included in the WHO classification,[6] the study group selected tumors that do not have an ICD-O3 code; however, the biologic behaviors of these tumors are well known among pathologists even though ICD-O3 codes were not granted. Of these, the following diseases were selected: fibromatosis colli, juvenile hyaline fibromatosis, inclusion body fibromatosis, superficial fibromatosis, lipofibromatosis, hyalinizing spindle cell tumor, PEComa, and clear cell myomelanocytic tumor.

Among the tumors of bone and soft tissue presented in the WHO classification, many tumors have behavior code -/1, such as, desmoid-type fibromatosis, inflammatory myofibroblastic tumor, and myopericytoma. These tumors have some characteristics in common, such as local recurrence, somewhat poor clinical outcome, and infrequent distant metastasis.[6] Therefore, these characteristics were considered in this study when determining the biologic behavior of the diseases.

Fibromatosis colli is a benign, site-specific lesion that occurs in the distal sternocleidomastoid muscle of infants.[6] It occurs in 4% of live births, and the majority of affected infants are diagnosed before 6 months of age. The site of involvement typically comprises the lower one third of the sternocleidomastoid muscle. It is managed in a non-surgical manner, but surgical intervention may be required in some patient.[16,17] We considered this lesion to be a benign disease and therefore suggest a behav-

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ior code of -/0. Juvenile hyaline fibromatosis is an extremely rare and apparently non-neoplastic disorder that typically presents in infancy, and is characterized by the accumulation of extracellular "hyaline material" within skin, somatic soft tissues, and the skeleton, resulting in tumor-like masses.[6] The hyaline material is produced by an aberrant population of fibroblasts. This tumor has a progressive nature and forms superficial and deep nodules with resulting deformity and dysfunction. It most frequently occurs in skin of the face and neck, gum, periarticular soft tissues, and several bone tissues. Surgery is the treatment of choice, but the recurrence rate is high.[18,19] There are reports that squamous cell carcinoma can arise from juvenile hyaline fibromatosis.[20,21] We considered this disease to be locally invasive with malignant potential, and therefore behavior code -/1 may be appropriate. However, definitive evidence of malignancy or metastasis was not found, and therefore we suggest biologic code -/0. Inclusion body fibromatosis is a rare benign proliferation of fibroblastic and myofibroblastic cells that typically occurs on the digits of young children. It is named for the intracytoplasmic inclusions that are detected in a minority of the lesional cells.[6] Treatment is surgical excision, but the local recurrence rate is high.[22,23] Thus, based on this characteristic, the biologic behavior may be /1, but because definitive evidence of malignancy or metastasis was not found, we also suggest behavior code -/0 for this disease as well. Lipofibromatosis is a rare pediatric tumor and a histologically distinctive fibrofatty tumour of childhood, which was previously designated as infantile fibromatosis of non-desmoid type with a predilection for distal extremities. The tumor has a high rate of non-destructive local recurrence, but exhibits no metastatic potential.[24,25] Because of its local recurrence, we suggest biologic behavior code -/1.

The PEComa category showed good agreement among participants and attendees of the workshop. PEComa is a neoplasm of perivascular epithelioid cells, and includes several diseases within the PEComa family.[6] However, their biologic behaviors have not been established because of the rarity of the disease and uncertain criteria for malignancy. The subset of PEComa tumors with malignant behavior do show a high mitotic index, necrosis, marked cytologic atypia, and an infiltrative growth pattern.[26] AML in this study represented non-renal AML. AML usually occurs in the kidney, but is also found in the liver, lung, uterus, skin and oral cavity in rare situations.[27] One report has indicated that LAM has a poor prognosis due to respiratory failure.[28] CCST is considered benign, but tumors larger than 2 cm that are symptomatic and focally necrotic should be regarded as potentially malignant neoplasms.[29] CCMMT has predilection for children and young adults, and it is known as a relatively benign disease.[30] As presented above, AML, LAM, and CCST could be considered as behavior code -/1, and CCMMT as -/0. PEComa NOS, which does not show AML, LAM, CCST, or CCMMT characteristics, is considered a subset of PEComa with malignant behavior,[26] and therefore we suggest behavior code -/3 for this tumor type.

We could not find studies that were similar to this study in literature databases from around the world, so comparison between our data and others was not possible. Malignant lymphomas are generally thought of as malignant diseases by physicians. However, new disease entities and a gradual increase of our understanding of rare diseases have resulted in a need for the continuous reconsideration of the biologic behavior of theses tumors. In Korea, the incidence of tumors of lymphoid tissue is increasing, and therefore the results of this study should be considered in the future as a comparator for newly reported studies.

Tumors of the bone and soft tissue undergo continual changes in the concept, classification, nomination, and biologic behavior. Bone and soft tissue tumors have a relatively low incidence, and therefore experience regarding these tumors is limited, especially for the diseases selected in this study. Although the majority of diseases entities in the study were benign or had uncertain biologic behavior and were not pertinent to the purpose of cancer registration, we think believe it is of great value to discuss and provide the behavior code for these rare disease entities. Therefore, this study has provided behavior codes for these selected tumors, but as future studies provide a more detailed understanding of these tumors, a further refinement may be necessary. In addition, tumor types not included in our questionnaire will be assessed in the future.

Conflicts of Interest

No potential conflict of interest relevant to this article was reported.

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