TUMOR HAND OUT

Evaluation of a bone lesion

Recommended references:

Metastatic bone disease: diagnosis, evaluation, and treatment. Biermann JS, Holt GE, Lewis VO, Schwartz HS, Yaszemski MJ. J Bone Joint Surg Am. 2009 Jun;91(6):1518-30. Review. No abstract available. PMID: 19487533

Evaluation of the patient with carcinoma of unknown origin metastatic to bone. Rougraff BT. Clin Orthop Relat Res. 2003 Oct;(415 Suppl):S105-9. Review. PMID:

Evaluation of a soft tissue masses

Soft tissue sarcomas that will metastasize to lymph nodes are few and can by memorized by the pneumonic **S C A R E**

Synovial cell sarcoma

Clear cell sarcoma

Angiosarcoma

Rhabdyomyosarcoma

Epithelioid sarcoma

While the "typical" soft tissue sarcoma of the extremity is located deep and in proximal musculature of adults, there are some notable exceptions.

Synovial cell sarcoma is a peculiar disease in that will present in younger patients, distal to the elbow or knee, and may have a prolonged period without growth – often resulting in a delay in diagnosis.

Clear cell sarcoma can arise from tendons or aponeuroses; it is rare for other sarcomas to do so.

Rhabdomyosarcoma and infantile fibrosarcoma are two sarcomas that will affect children.

Pathologists may use Fluorescent in situ hybridization (FISH) to identify translocations to better determine the histologic diagnosis. These translocations and their fusion products are often tested on exams. The following table is a representation of commonly tested diseases, translocations, and genes involved.

Histologic Subtype	Translocation	Genes Involved
Alveolar rhabdomyosarcoma	t(2;13) (q35,q14)	PAX3, FKHR
	t(1;13) (p36;q14)	PAX7, FKHR
Alveolar soft part sarcoma	t(X;17) (p11;q25)	TFE2, ASPL
Clear cell sarcoma	t(12;22) (q13;q12)	ATF1, ETF
Dermatofibrosarcoma protuberans	t(17;22) (q22;q13)	COL1A1, PDGFB1
Desmoplastic small round cell tumor	t(11;22) (p13;q12)	WT1, EWS
Ewing's sarcoma/primitive neuroectodermal tumor	t(11;22) (q24;q12)	FLI1, EWS
	t(21;22) (q22;q12)	ERG ,EWS
	t(7;22) (p22;q12)	ETV1, EWS
	t(2;22) (q33;q12)	FEV, EWS
	t(17;22) (q12;q12)	E1AF, EWS
Extraskeletal myxoid chondrosarcoma	t(9;22) (q21–31;q12.2	CHN, EWS
	t(9;17) (q22;q11)	CHN, RBP56
Myxoid/round cell liposarcoma	t(12;16) (q13;p11)	CHOP, TLS
	t(12;22) (q13;q11–q12)	CHOP, EWS
Synovial sarcoma	t(X;18) (p11.2;q11.2)	SSX1 or SSX2, SS18 (SYT)

Adapted from Clark MA, Fisher C, Judson I, Thomas JM. Soft-tissue sarcomas in adults. *N Engl J Med.* 2005; 353: 701–711.

The stage of any cancer is meant to elucidate the extent of the disease which can in turn guide prognosis and treatment.

CT scan of the chest is part of the staging of every soft tissue sarcoma given the proclivity of sarcoma to metastasize to lung.

CT of the abdomen and pelvis and nuclear medicine bone scan are also used the staging in myxoid liposarcoma given the ability of this disease to metastasize to fat. Bone marrow and the retroperitoneum are two "fat-rich" areas where metastases can be identified.

The below table demonstrates the staging system employed by the American Joint Committee on Cancer (AJCC) and the International Union Against Cancer (UICC) for soft tissue sarcoma:

TABLE 1: AJCC version 7 staging for soft tissue sarcomas

Primary Tumor (T)

TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
T1	Tumor 5 cm or less in greatest dimension
T1a	Superficial tumor
T1b	Deep tumor
T2	Tumor more than 5 cm in greatest dimension
T2a	Superficial tumor
T2b	Deep tumor

Note: Superficial tumor is located exclusively above the superficial fascia without invasion of the fascia; deep tumor is located either exclusively beneath the superficial fascia, superficial to the fascia with invasion of or through the fascia, or both superficial yet beneath the fascia.

Regional Lymph Nodes (N)

NX	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1	Regional lymph node metastasis

Note: Presence of positive nodes (N1) in M0 tumors is considered Stage III.

Distant Metastasis (M)

M0	No distant metastasis
M1	Distant metastasis

ANATOMIC STAGE/PROGNOSTIC GROUPS

Stage IA	T1a	N0	M0	G1, GX
	T1b	N0	M0	G1, GX
Stage IB	T2a	N0	M0	G1, GX
	T2b	N0	M0	G1, GX
Stage IIA	T1a	N0	M0	G2, G3
	T1b	N0	M0	G2, G3
Stage IIB	T2a	N0	M0	G2
	T2b	N0	M0	G2
Stage III	T2a	N0	M0	G3
	T2b	N0	M0	G3
	Any T	N1	M0	Any G
Stage IV	Any T	Any N	M1	Any G

From Edge SB, Byrd DR, Compton CC, et al (eds): AJCC Cancer Staging Manual, 7th ed. New York, Springer, 2010.

A Few Notes on Primary Bone Tumors

Osteosarcoma is the most common malignancy of bone in children and adolescents.

It will often present with a pain or pathologic fracture at sites of marked skeletal growth (most notably the distal femur, proximal humerus, proximal tibia). Staging consists of CT scan of the chest and bone scan to look for pulmonary and skeletal metastasis, respectively. After the lungs, the skeleton is the second most common site of osteosarcoma metastasis.

Table 1: Ennekings system of classification of Malignant musculoskeletal tumors

Stage	Grade	Site	Metastasis
IA	Low	Intracompartmental	None
IB	Low	Extracompartmental	None
IA	High	Intracompartmental	None
IIB	High	Extracompartmental	None
III	Any	Any	Regional or distant metastasis

Table 2: American Joint Committee on Cancer system of staging bone cancers

Stage	Grade	Size	Metastasis
I-A	Low	<8 cm	None
I-B	Low	>8 cm	None
II-A	High	<8 cm	None
II-B	High	>8 cm	None
III	Any	Any	Skip metastasis
IV-A	Any	Any	Pulmonary metastasis
IV-B	Any	Any	Nonpulmonary metastasis

Shah M, Anchan C. Evaluation of Osteogenic Sarcoma. Journal of Bone and Soft Tissue Tumors Jan-Apr 2016;2(1):8-12.

Cartilage lesions are common with very few being malignant. Enchondromas are often incidental findings, most notably in the proximal humerus, in patients with shoulder pain from mechanical etiologies. Biopsy is generally not indicated. Use of injections can be helpful as “tumor pain” will not

respond to intraarticular or bursal injection. Radiographic surveillance for any change in the appearance of the lesion is indicated with loss of calcification, new bony erosions, or development of a soft tissue mass being the most concerning findings for sarcomatous degeneration.

Intra-osseous cartilage tumors in the pelvis are chondrosarcoma even if relatively under whelming histologically given the aggressive natural history of pelvic chondrosarcoma even in the setting of a low histologic grade. Treatment of chondrosarcoma is surgical as it does they generally do not respond to chemotherapy or radiation. Chondrosarcoma is the most common primary malignancy of bone in adults.