Primary NHL (Non Hodgkins Lymohoma) of Bone.

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Primary NHL of bone
Extra nodal Lymphoma
Reticulum Cell Sarcoma

The common type is
DLBCL Diffuse Large B Cell Lymphoma

B Cell Lymphoma
Also get T Cell Lymphoma
DLBCL more common

Incidence
7 to 10 % of Primary bone tumors
1 to 2 % of Adult NHL

Age
45 to 60 years
In children mean is 12 years and in Adolescents

Sex
Equal
Males 60 %

Association
EBV
HIV
Human Herpes Virus 6

Treatment
Radiotherapy
Chemotherapy
CHOP R
Cyclophosphoamide , Doxorubicin ,Vincristine , Prednisone
Rituximab

Clinical
Pain
Swelling , Mass
Pain not relieved by rest
Limp
Night Pain
Pathological fracture
Spinal cord compression
B symptoms. - fever , night sweats , loss of weight

Monoostotic
Polyostotic

Bones
Long bones Femur
Vertebrae, pelvis. Mandible, Ribs, sternum. Clavicle, palate ,scapula ,skull (Axial skeleton)

Site
Metaphysis + Diaphysis
Medullary cavity

Imaging
Moth eaten , permeative, lytic
Entire bone or large areas of the Bone or wide areas
No internal matrix
No periosteal reaction
Erodes the bone
Lytic ,Blastic, mixed
Soft tissue extension
Muscle extension
Pathological fracture
Unifocal , multifocal
One bone or multiple bones
Monoostotic, polyostotic
Large portion of the bone
Cortex destroyed
Without periosteal reaction or new bone formation
MRI signal abnormalities in bone Marrow

DD on imaging
Benign Reactive lesions , osteomyelitis
Malignant - Hodgkins Lymphoma, Sarcoma, Neuroblastoma, Ewings , LCH

Blood tests
Anemia , ESR ,CRP
LDH increased
Alkaline phosphatase increased
B2. MIcroglobulin increased
Calcium increased

Platelet count increased

CBC
PS
Serum Chemistry
LDH
BM Aspiration
BM Biopsy
Core Needle
Incisional
Excision
Chest X-rays
Plain X-rays
CT
MRI
PET CT
Nuclear imaging

IHC
DLBCL
Germinal Center Phenotype
CD 10 +
BCL 6 +
MUM 1 -

Post Germinal (Non Germinal) Center phenotype
CD 10 -
BCL 6 -
MUM 1 +

CD 45 (LCA) Positive

In DLBCL
CD 20 +

Genetics / Molecular
Rearrangement of
BCL 2
c Myc
Dual Rearrangement

Gross
White
Yellow
Tan
Fish flesh
Infiltrative
Ill circumscribed

Microscopic
Sheets
Clusters
Small to Large lymphocytes
Vesicular nucleus
Polymorphic Infiltration
Atypical lymphocytes
Diffuse growth
Small lymphocytes admired with Large lymphocytes
Marked swindling, fibrosis

Other Lymphomas in the bone
Follicular Lymphoma
Peripheral T Cell L
ALCL
CLL
Marginal Zone L
Mantle Cell L
Burkitts
Lymphoblastic

In children
Burkitts
Lymphoblastic
DLBCL

Primary Bone Lymphoma
Presence of Lymphoma isolated to one bone without distant spread for 6 months after diagnosis

Primary Bone Lymphoma
Group 1 Solitary
Group 2 Multifocal
Group 3 Distant LN metastasis
Group 4 Visceral Disease

Relapse
Other bones
LNs
Soft Tissue
Lungs
BM

DD
Ewings
Neuroblastoma
Round Cell , Blue Cell , Small Cell
Prognosis depends on
Age , sex , stage ,Type of bone , number of bones , Histopathology, LDH level, spinal cord compression

Worse prognosis
Below 9 years
Female pt
Non large Cell histology
BM involvement on biopsy
Increased Calcium
LDH
Number of Extra nodal sites

Old term
Reticulum Cell Sarcoma

Flow Cytometry
FISH
PCR
IHC
NGS

Electron Microscopy

CD 99 negative
CK negative
S 100 negative

Molecular,
Cytogenetic
Clonally Rearranged Ig Gene
(BCL 2. ,BCL 6. Myc
80 % show chromosonal abnormality

HIV
EBV
Herpes Virus 6

Primary Lymphoma of bone with Spindle Cell Morphology
Common sites are
Nasal
Ocular
Skin
Soft tissue

Lymphoma cells produce cytokines TNF , PDGF , TGF B
These growth factors lead to the proliferation of fibroblasts
The fibroblasts impinge on the lymphocytes
So spindle shared. , spindling

CD 45 +
CD 20 +
CD 79a +
PAX 5 +
DLBCL

Decal careful
Destroys the tissue

Infiltrative pattern in bone
Permeates marrow, fat and intact structures

Crush artifacts

Reticulin stain
Network of reticulin fibres around each cell or clusters of cells

Reactive fibrosis
Fine , in between the cells
Spindle cells, storiform pattern , permeative pattern.
Lymphocytes in the background - Lymphoma
Multiple Skeletal sites ,- Lymphoma

Imp feature
Polymorphism of infiltrate
In Ewing's the cells are uniform

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