|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| **Type** | **Subtype** | Prevalence | Radiographic Appearance | Histologic Morphology | Notable Features | Histology Images | Clinical Images |
| **Intramedullary** |  |  |  |  |  |  |  |
|  | **Conventional** | >80% | * Permeative lesion with varying degrees of osteolysis and osseous matrix
* Cortical destruction, soft tissue extension
* Sunburst periosteal reaction, Codman’s triangle
* Lytic pattern may predominate in older patients13
 | Severe anaplasia, pleomorphism, frequent mitoses, immature osteoid matrix, varying degrees of additional types of extracellular matrix determine the histologic variant1. **Osteoblastic**: 75% of cases12
2. **Chondroblastic**:defined by at least 30% chondroid matrix composition, 16% of cases15
3. **Fibroblastic**: defined by dominant collagen matrix with a storiform pattern, 10% of cases7,12
4. **Others**: includes giant-cell rich, osteoblastoma-like, epithelioid, clear cell, chondroblastoma-like12
 |  | Figure 1 Supplemental Digital Content | Figure 2 Supplemental Digital Content |
|  | **Telangiectatic** | 4-8%14, 16 | * Eccentric with cortical expansion
* Predominantly lytic
* MRI demonstrates a soft tissue mass, areas of hemorrhage, peripheral, septal and nodular enhancement with contrast17
 | Blood filled spaces separated by septae with pleomorphic, malignant cells. Osteoid may be less apparent or absent on biopsy samples | * Diagnosis may be difficult given radiographic resemblance to aneurysmal bone cysts
* Pathologic fracture is common (~30%)14
 | Figure 3 Supplemental Digital Content | Figures 4 and 5 Supplemental Digital Content |
|  | **Small** **Cell** | 1% | * Permeative lesion with variable amounts of intramedullary sclerosis
* Mineralization may be absent/minimal
 | Small, round malignant cells with varying amounts of osteoid production | Distinguished from Ewing sarcoma by the lack of nuclear spindling and the FLI-1 translocation18 |  |  |
|  | **Low Grade Central** | 1-2% | * Mixed sclerotic/lytic lesion with a geographic border
* Cortical erosion and an extraosseous soft tissue mass, seen more readily with MRI
 | Hypocellular spindle tumor with minimal cytologic atypia with a permeative growth pattern within the surrounding normal bone | Overexpression of CDK4 and MDM2 helps differentiate from benign fibro-osseous lesions19 | Figure 6 Supplemental Digital Content | Figure 7 Supplemental Digital Content |
| **Surface** |  |  |  |  |  |  |  |
|  | **Parosteal** | 4%20 | * Well-mineralized cortically based mass
* Medullary involvement is present in 35% of cases20
 | Spindle cells with variable amounts of trabecular bone and cartilage | * Posterior distal femur is the classic location
* Overexpression of CDK4 and MDM221
* Dedifferentiated parosteal osteosarcoma can develop in approximately 25%21
 | Figure 6 Supplemental Digital Content | Figures 8 and 9 Supplemental Digital Content |
|  | **Periosteal** | < 2%22 | * Broad-based tumor on the surface of the diaphysis
* Cortical thickening and scalloping, periosteal reaction, soft tissue mineralization23
 | Areas of atypical cartilage with focal areas of intermediate osteosarcoma arising beneath the periosteum | * Diaphyseal location
* Role of chemotherapy is controversial
* Overall survival superior to conventional osteosarcoma
 | Figure 10 Supplemental Digital Content | Figures 11-13 Supplemental Digital Content |
|  | **High-Grade Surface** | < 1% | * Radiographically may resemble parosteal or periosteal osteosarcoma, but with more aggressive features
* Medullary involvement present in approximately 50% of cases24
 | Surface based lesion that histologically resembles high-grade, intramedullary conventional osteosarcoma with pleomorphic cells producing osteoid. May have histologic variants, including osteoblastic or chondroblastic | Treated like conventional, intramedullary osteosarcoma  |  | Figure 14 Supplemental Digital Content |

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