Kötü Huylu (Malign) Kemik Tümörleri

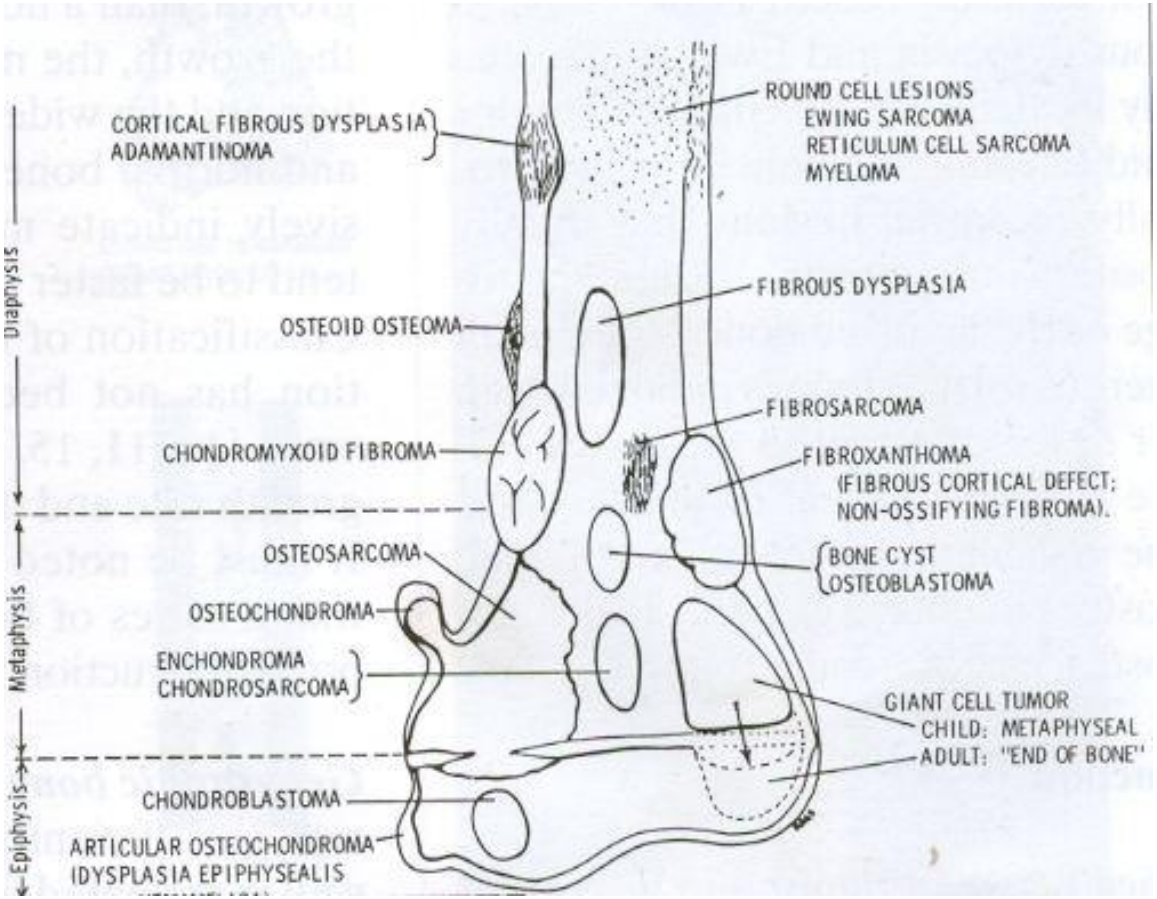
Doç. Dr. Seyit Ali Gümüştaş

Malign Kemik Tümörleri-Kinik

- **Ağrı**
  - Önceleri aralıklı iken sürekli hale gelen, istirahatte geçmeyen ve ağrı kesicilere sınırlı cevap veren
  - Çocuklarda neden (Travma, Enfeksiyon, Romatizmal, AVN, Tümör vs) ortaya konulmalı
  - Büyüme ağrısı yoktur
- **Şişlik**

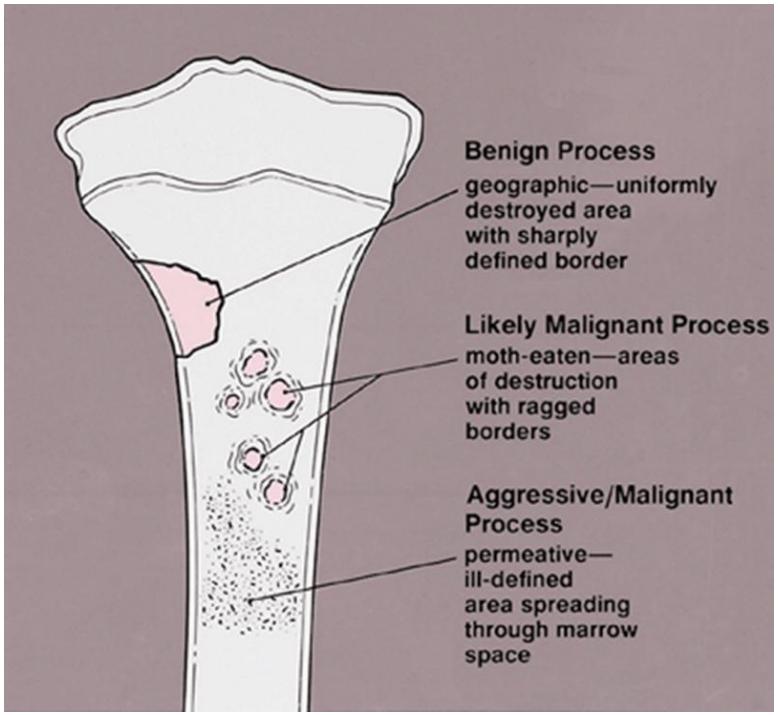
Malign Kemik Tümörleri-Radyoloji

- **Yerleşim Yeri**

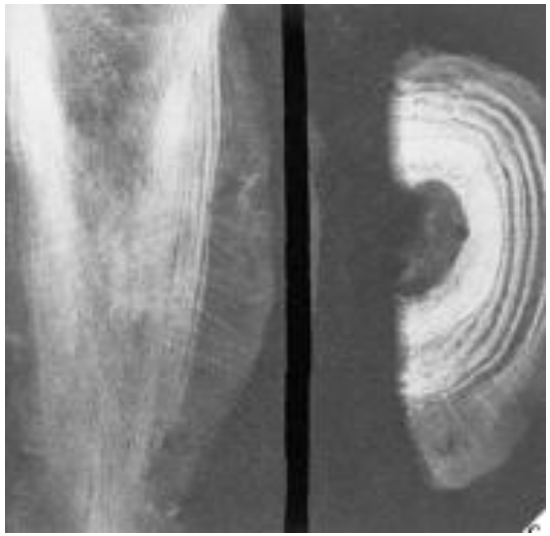


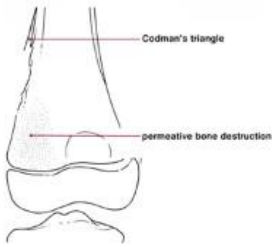
Destrüksiyon Tipi (Lodwick)

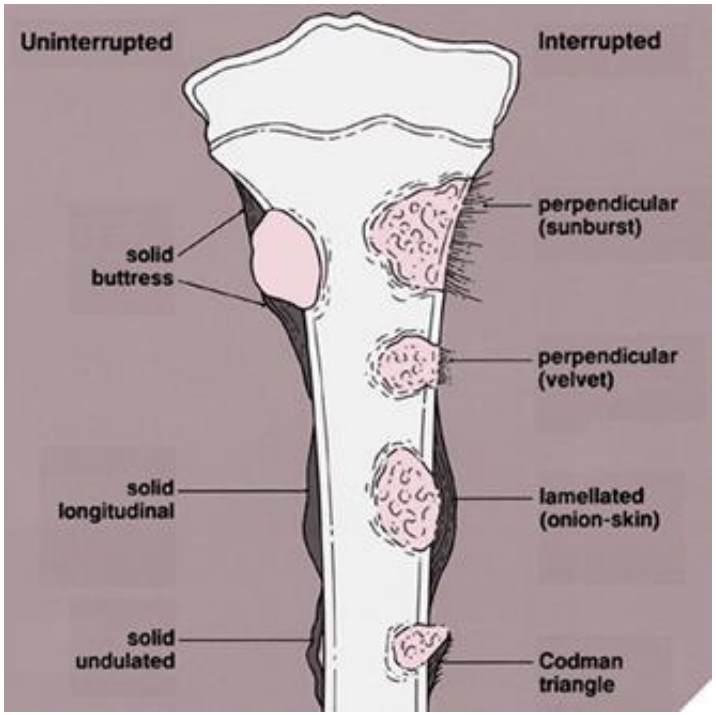
- **Coğrafik** patern (tip I)
- **Güve yeniği** patern (tip II)
- **Permeatif** patern (tip III)



Periost Reaksiyonu

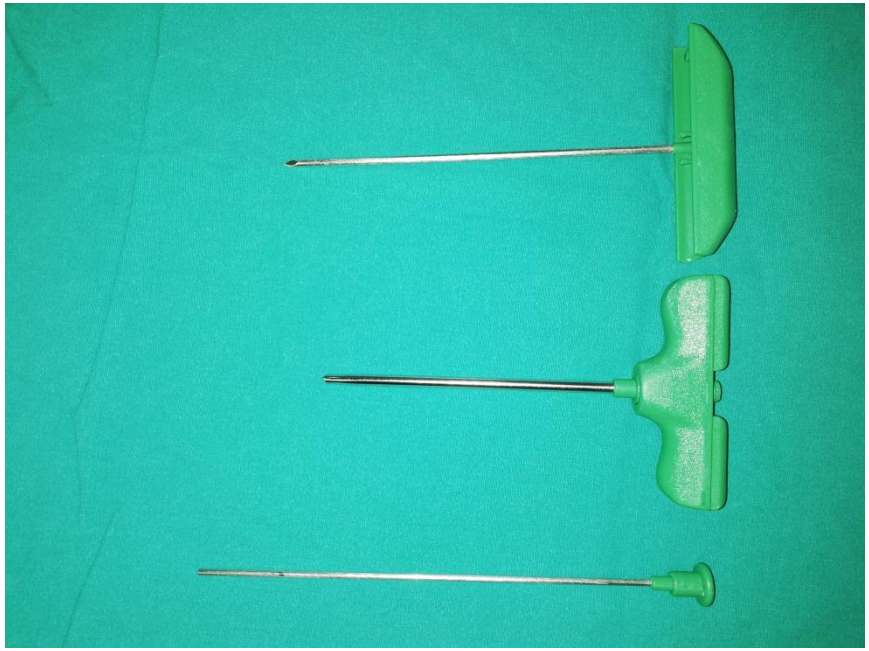






Osteosarkom-Tanı

- Biyopsi: Core-needle/Jamshidi



Tarama-Evreleme

- Akciğer tomografisi
- Tüm vücut kemik sintigrafisi (Tc99m)

American joint committee on cancer system bone sarcoma classification (AJCC Classification)

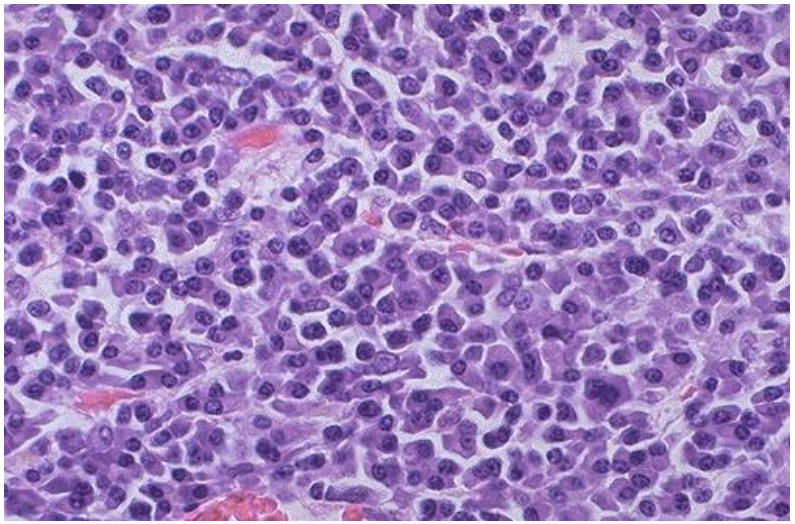
The AJCC system for bone sarcomas is based on tumor grade, size, and presence and location of metastases.

Stage	Grade	Size	Metastases
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 metastases
IV-B	Any	Any	Nonpulmonary metastases

Malign Kemik Tümörleri

- **Primer**
  - Multıpl Myelom
  - Osteosarkom
  - Ewing Sarkom
  - Kondrosarkom
  - Kordoma
  - Adamantinoma
- **Sekonder**
  - Postradyasyon
- **Metastatik**
  - Ekstremite
  - Omurga

Multıpl Myelom



- Plazma hücrelerinin neoplastik monoklonal proliferasyonu
- Kemiğin en sık primer malign tümörü

Multıpl Myelom-Klinik

- Yaygın kemik ağrısı
- Patolojik kırık
- Sistemik bulgu (Kilo kaybı, yorgunluk, ateş, HSM, LAP)

Multıpl Myelom-Laboratuar

- Hiperkalsemi
- Anemi
- Böbrek yetmezliği (Kötü prognoz)
- Sedim yüksekliği
- Serum ve idrar elektroforez (Monoklonal gamopati)

Multıpl Myelom-Tanı

- Bir major bir minör yada üç minör
- **Major kriterler**
  - Plazmositoma (Doku biyopsisi)
  - Kemik iliğinde >%30 plazma hücreleri
  - Serum IgG>3.5 g/dL, IgA>2g/dL, yada İdrar urin>1g/24 saat, yada Bence Jones proteini
- **Minör kriterler**
  - Kemik iliğinde %10-30 plazma hücreleri
  - Serum ve idrar protein değerleri major kriterlerden az
  - Litik kemik lezyonları
  - <Normal IgG değeri

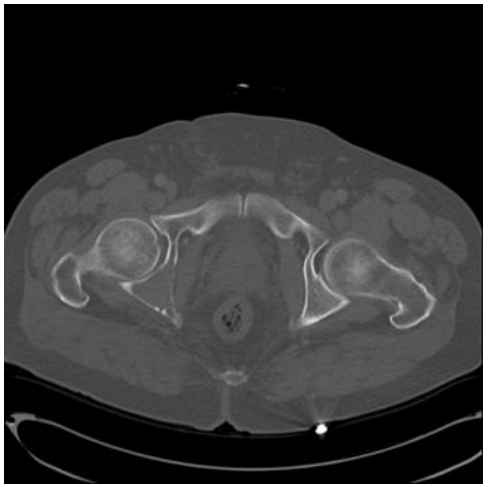
Multıpl Myelom-Görüntüleme

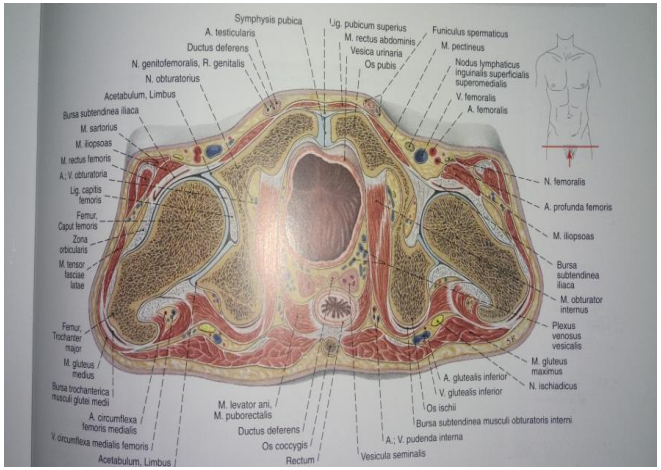
- Kafatası ve omurga tutulumu sık
- Litik lezyon = **Zımba deliği** (‘’punched-out’’)
- Çevreleyen skleroz yok
- Diffüz osteopeni
- **Negatif sintigrafi** (düşük osteoblastik aktivite)
- MRI erken tanı için kullanılır

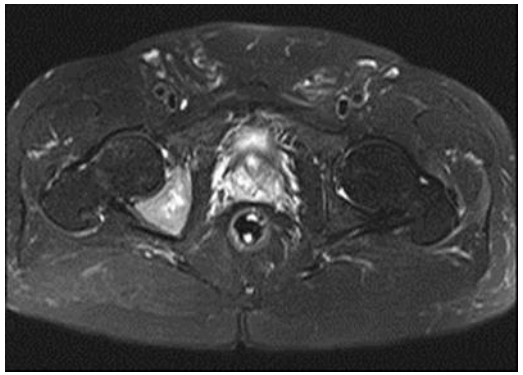
Multıpl Myelom-Tedavi

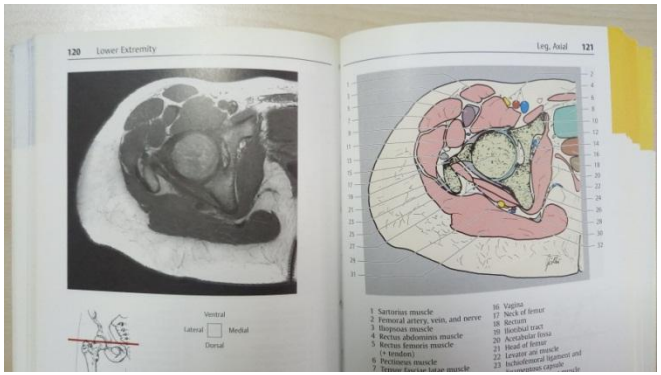
- **Standart:** Yüksek doz KT + Otolog kök hücre transplantasyonu
- **Kortikosteroid**
- **RT:** Ağrıları hafifletmek ve cerrahiden kaçınmak için
- **Bifosfonat/Denosumab:** Kemik lezyon sayısını azaltır, ağrıyı hafifletir, serum kalsiyum seviyesini azaltır
- **Cerrahi:** Patolojik kırık yada kırık riski
- **Kifoplasti/Vertebroplasti:** Vertebra kompresyon fraktürü

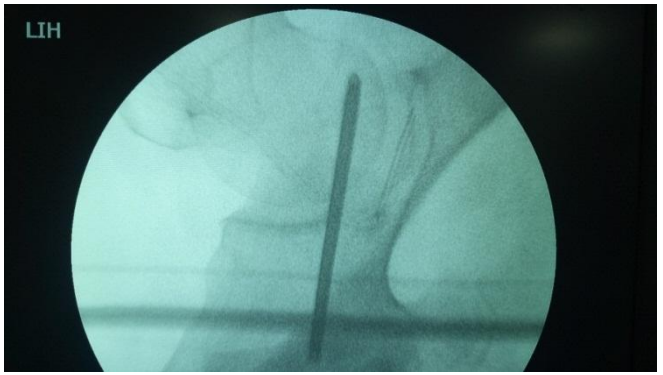
Plazma Hücreli Neoplazm



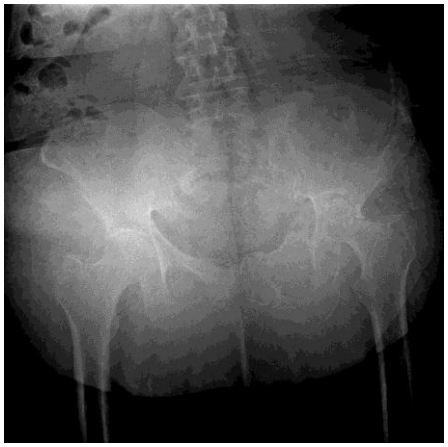


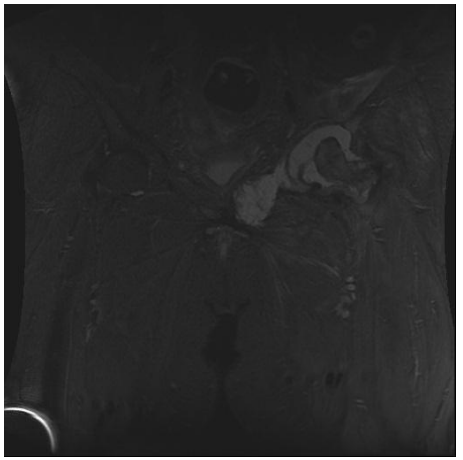
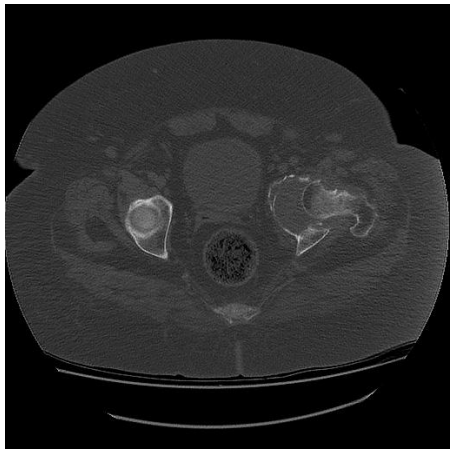


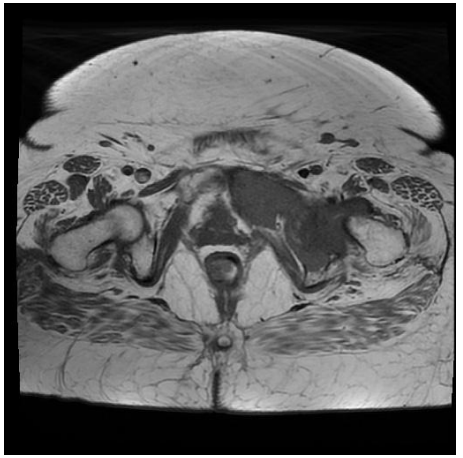




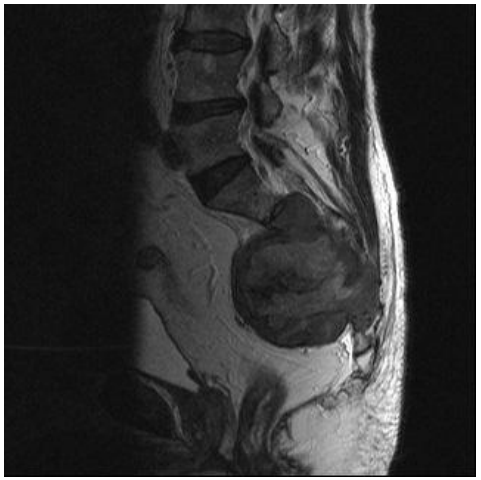
Asetabulum Plazma Hücreli Neoplazm

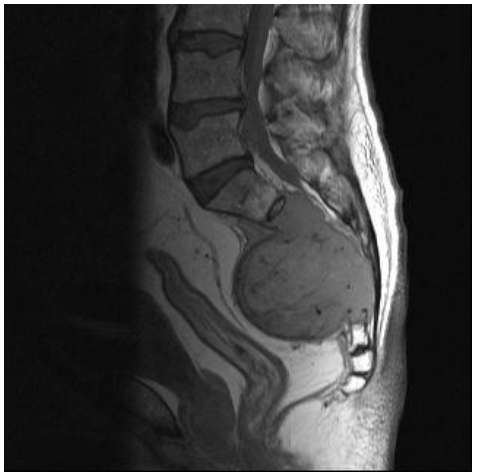


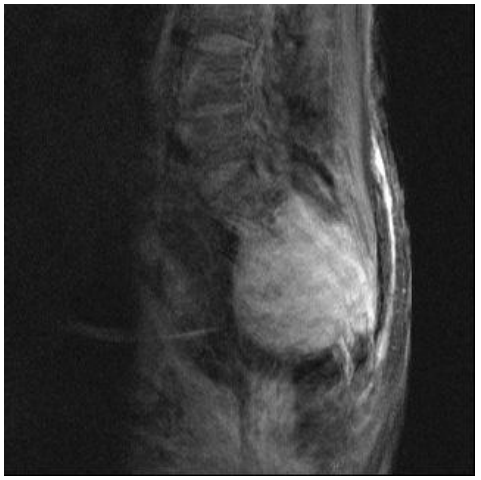





Sakral Plazma Hücreli Neoplazm

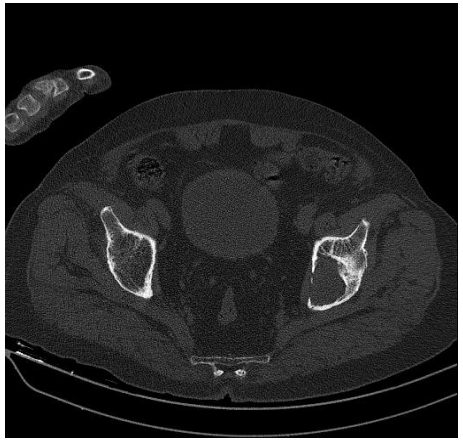


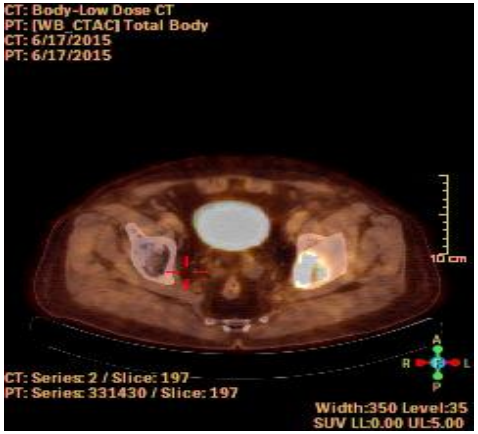




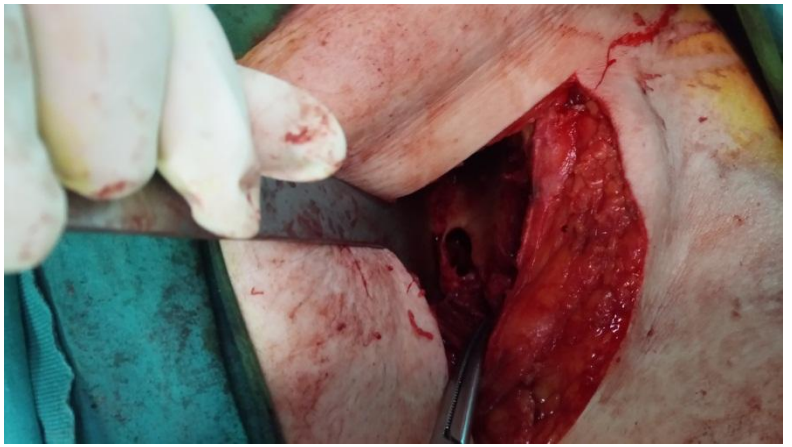
Multıpl Myelom
Patolojik Humerus ve Vertebra Fraktürü

Multıpl Myelom



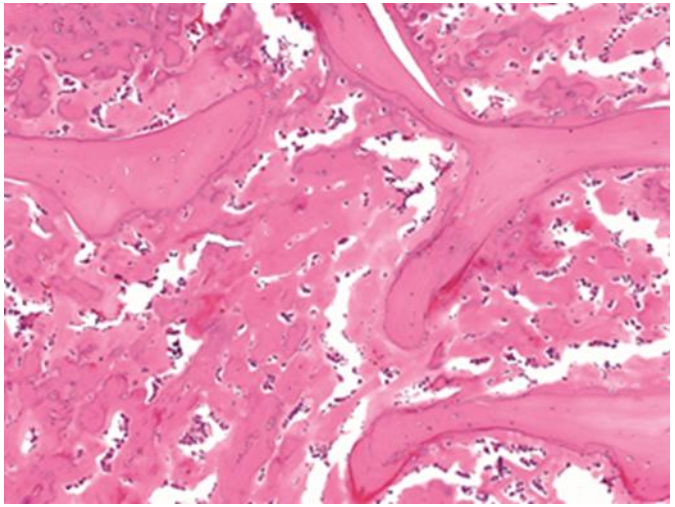


Proksimal Humerus ve Asetabulum Fraktürü



Osteosarkom

Osteosarkom



- Adelosan dönem
- En sık **diz çevresi** sonra proksimal humerus
- Travma hikayesi tarifleyebilir

Osteosarkom-Görüntüleme

- **Metafiz** yerleşimli
- **Sunburst/Codman üçgeni** periost reaksiyonu
- **Skip lezyon** varlığı ekarte edilmeli (Sintigrafi, MRI)

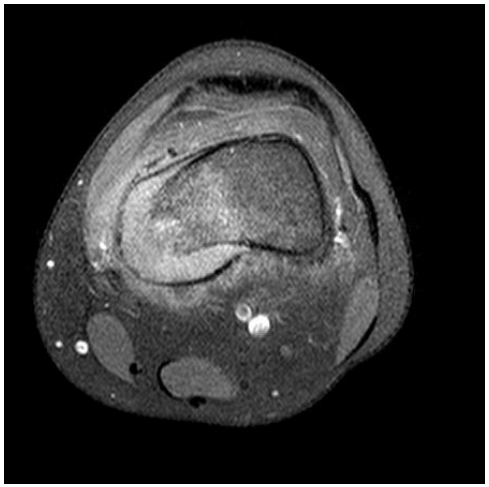
Osteosarkom-Laboratuar

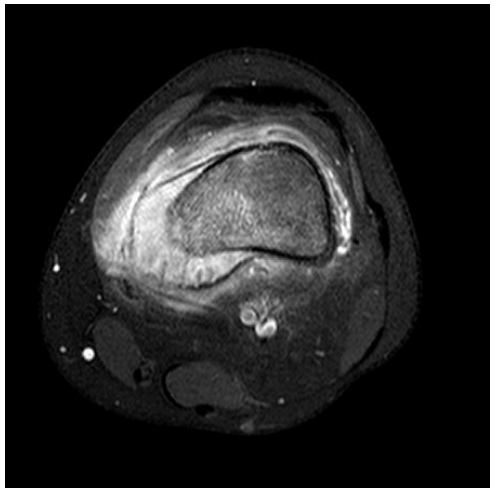
- Hemogram, Sedim, CRP (Osteomiyelit ayırıcı tanı için)
- **ALP** yüksekliği (Osteosarkom)
- LDH (Ewing sarkom)

Osteosarkom-Tiplendirme

- **Primer**
  - Klasik intramedüller
  - Parosteal
  - Periosteal
  - Telenjektatik
- **Sekonder**
  - Paget
  - Post-radyasyon

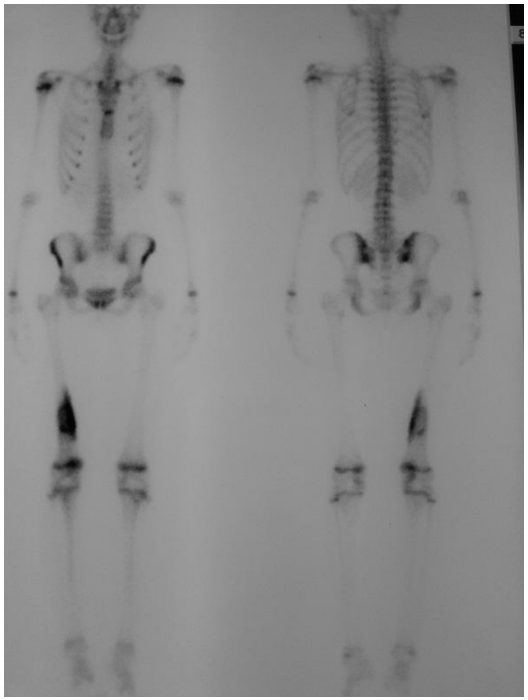
Distal Femur Klasik Osteosarkom



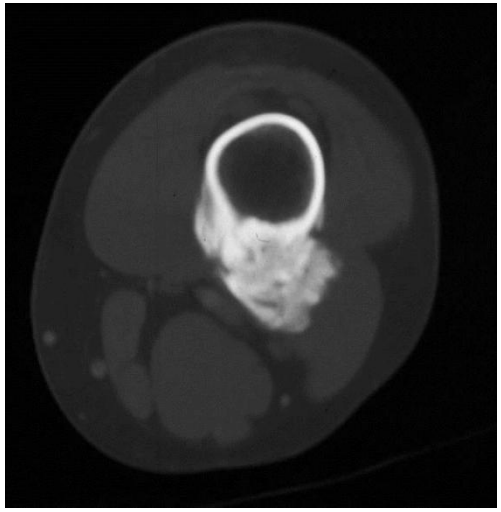


Sağ uyluk ağrısı

Tlenjektatik osteosarkom



Parosteal Osteosarkom-Osteokondrom



Periosteal Osteosarkom

Osteosarkom-Tedavi

- **Neoadjuvan KT**

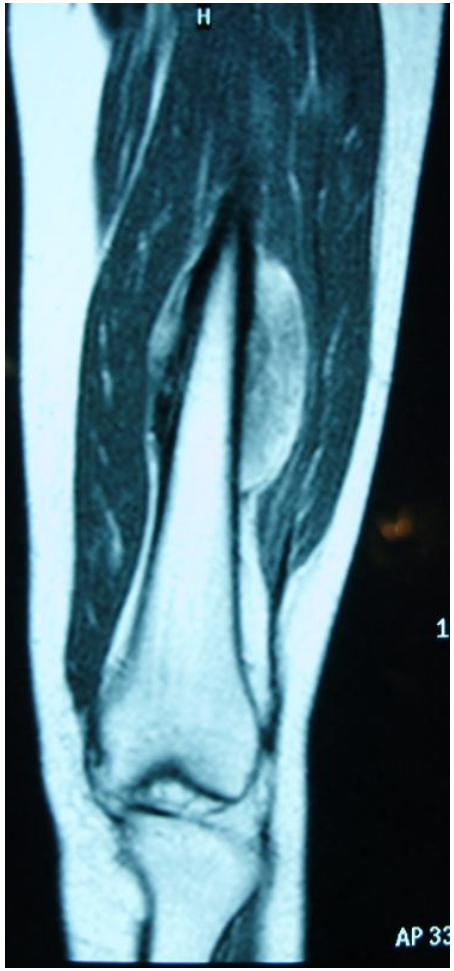
➢ Adriamycin (Doxorubicin), Cisplatinum, Methotrexate, İfosfamid

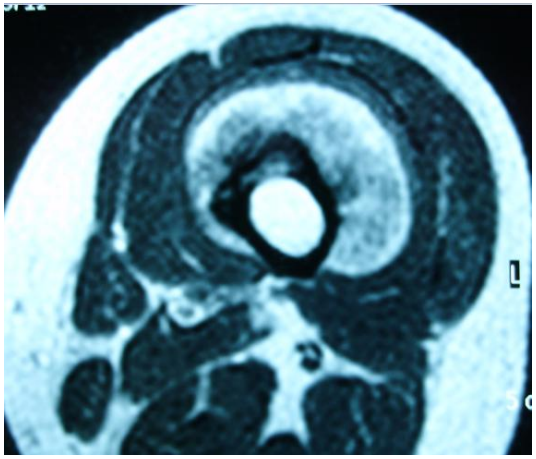
- **Cerrahi** (Geniş rezeksiyon ve rekonstrüksiyon)
- **Adjuvan KT**
- **Radyoterapi** standart tedavi protokolünde yok.

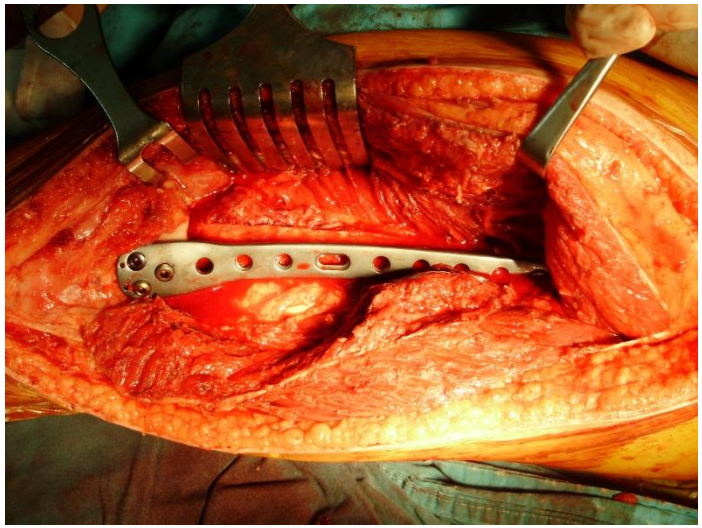
Osteosarkom Kötü Prognoz Kriterleri

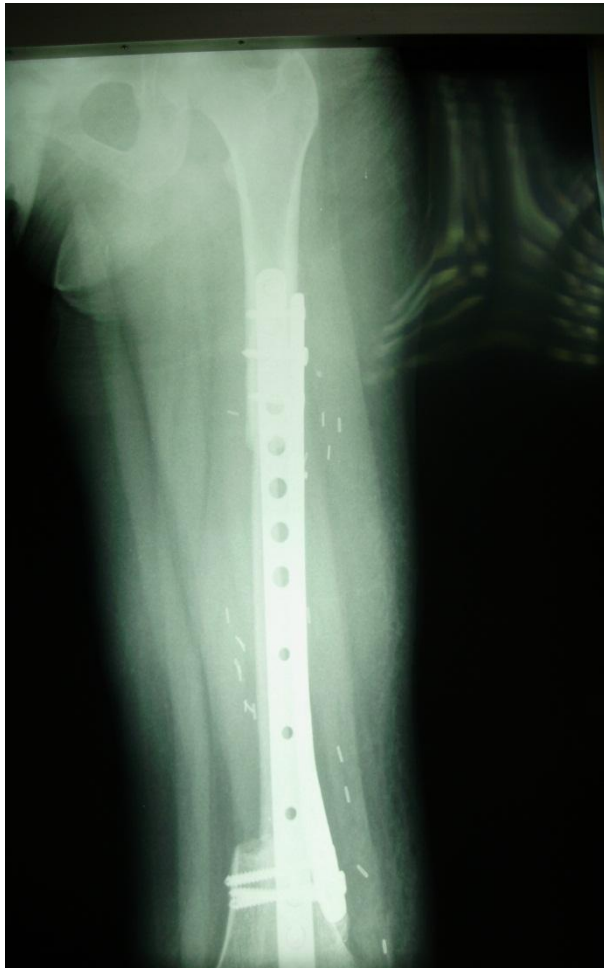
- **Metastaz varlığı** (Skip lezyon dahil)
- **Kötü KT yanıt**. (<%90 nekroz)
- **Pelvis,omurga yerleşimi**
- **Patolojik kırık** varlığı

Parosteal osteosarkom

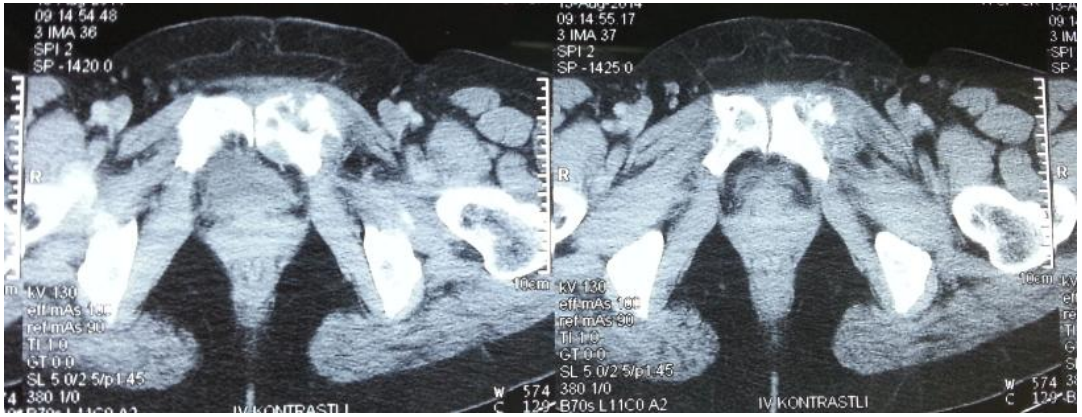


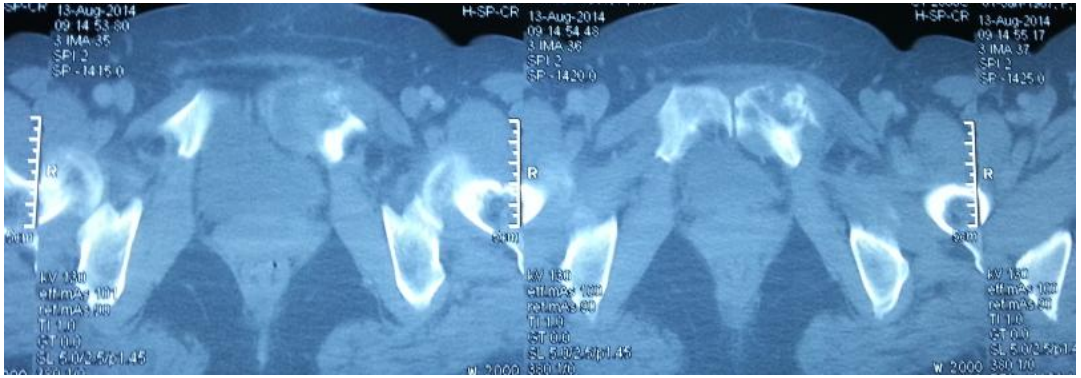




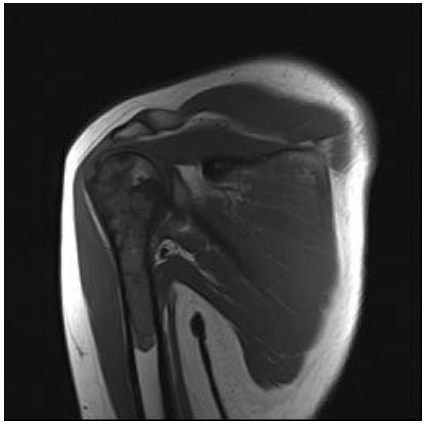


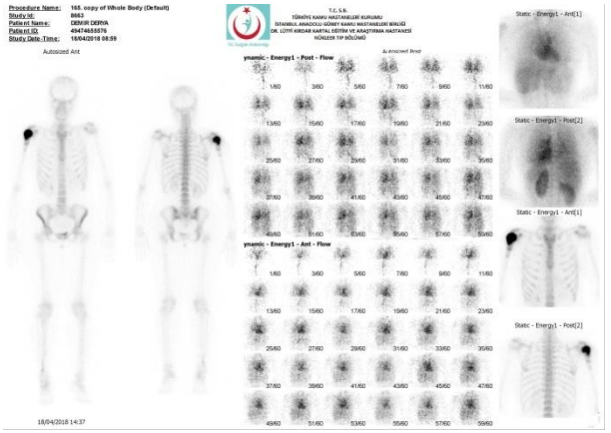
Sol pubik kol osteosarkom



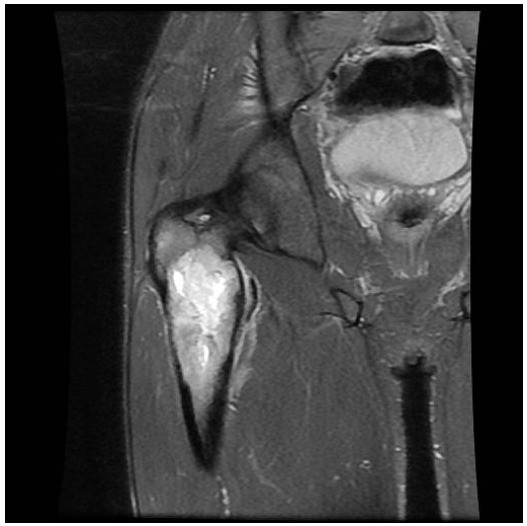
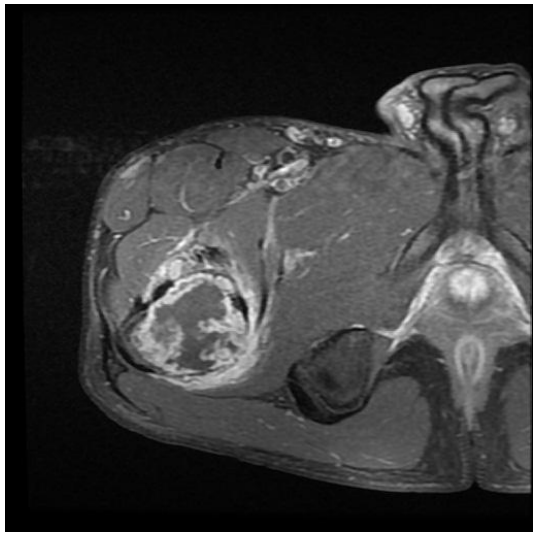


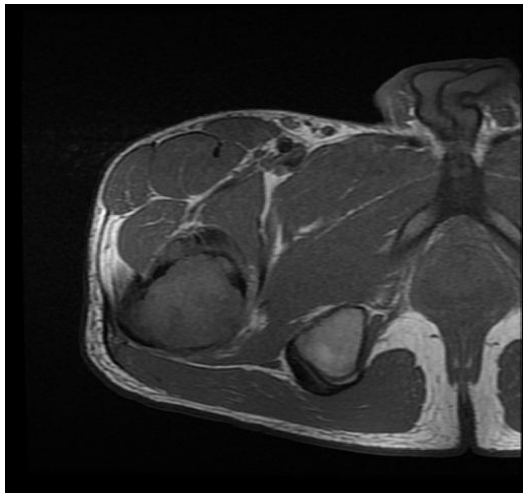
Kondroblastik Osteosarkom



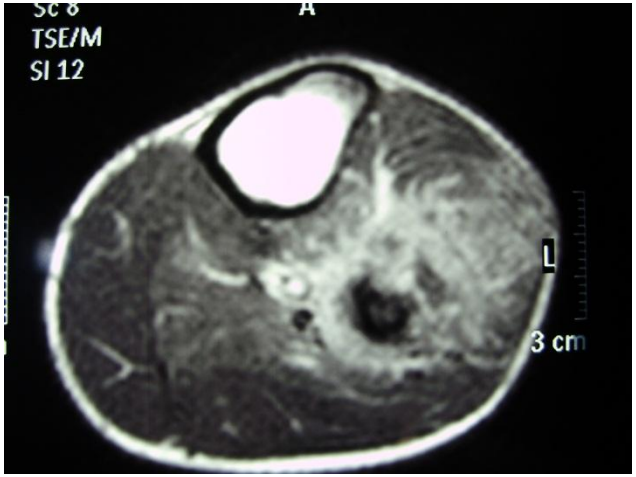


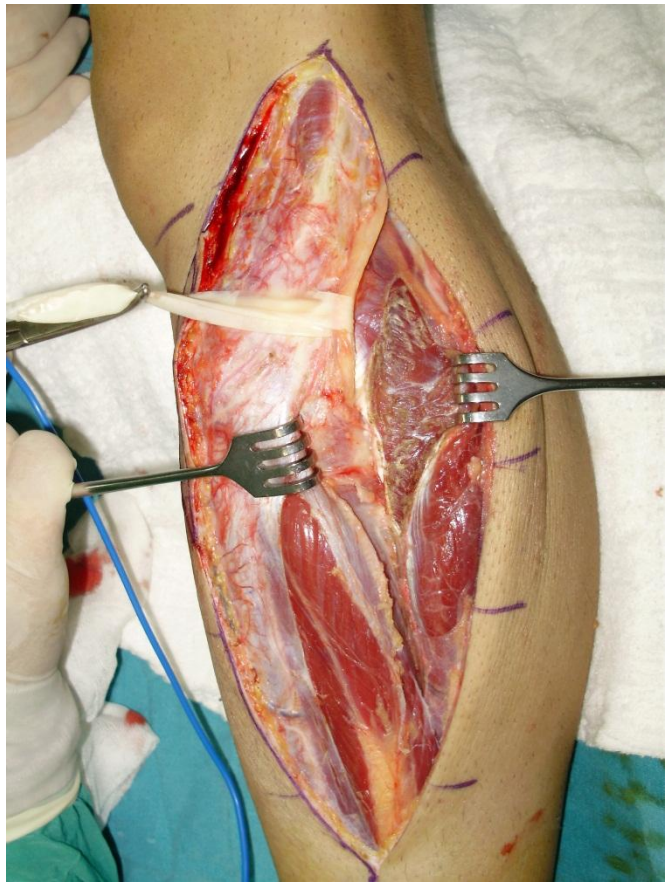
Kondroblastik osteosarkom
Patolojik Fraktür

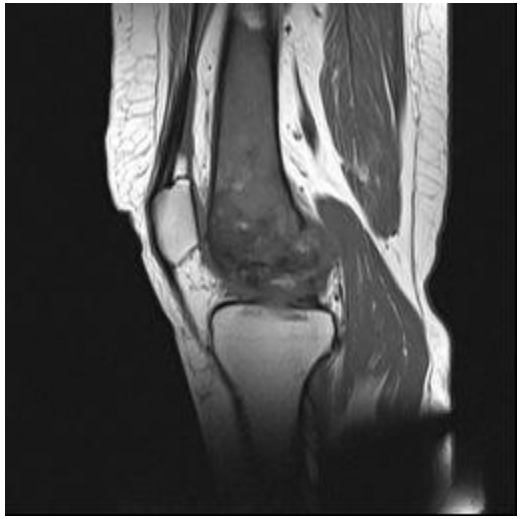



Proksimal fibula osteosarkom

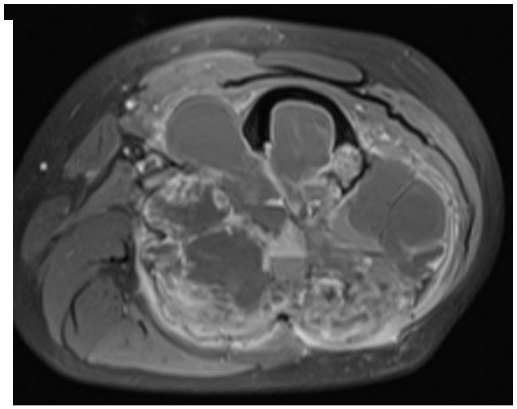


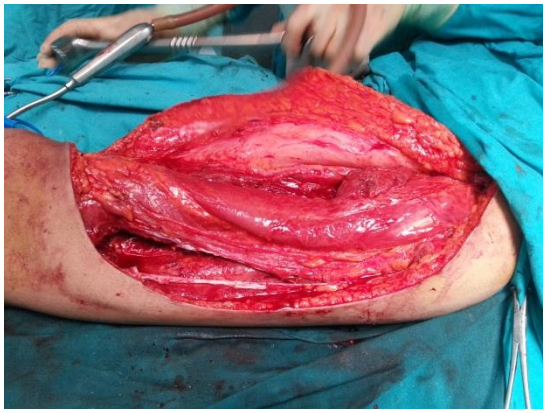


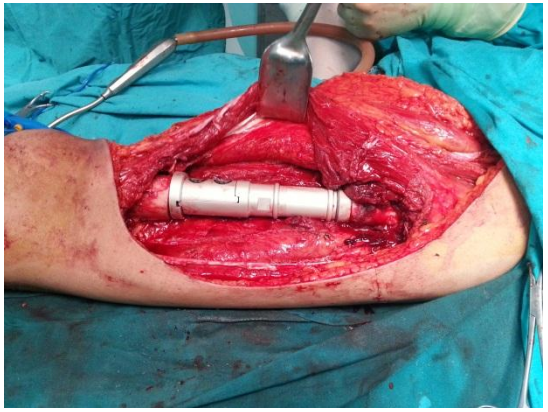
Distal Femur Osteosarkom

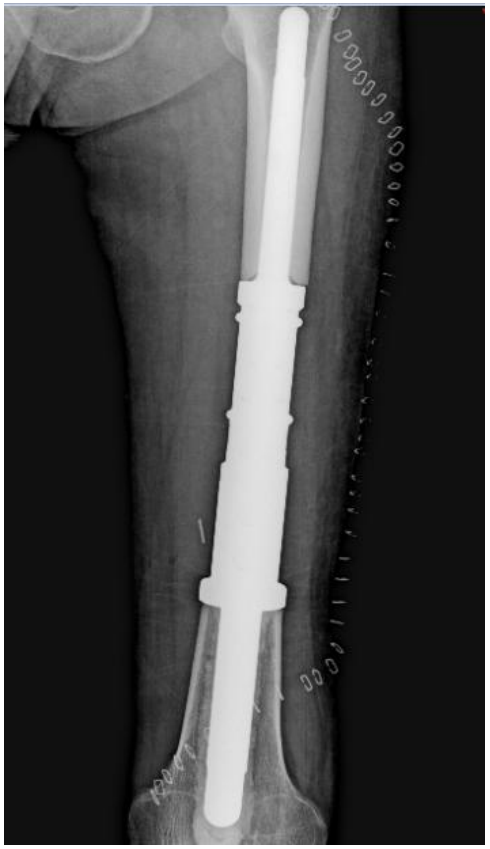


Telenjektatik osteosarkom

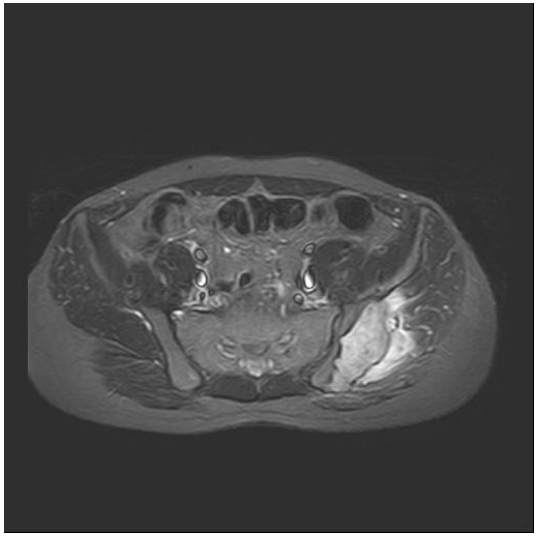


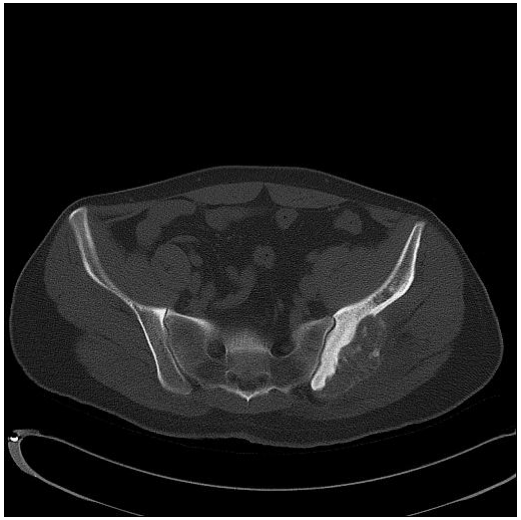


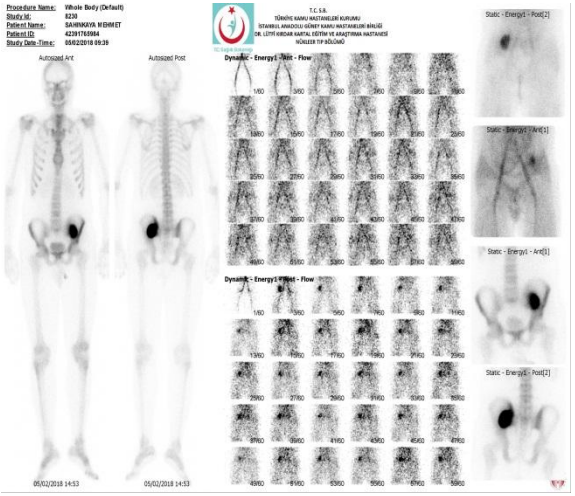




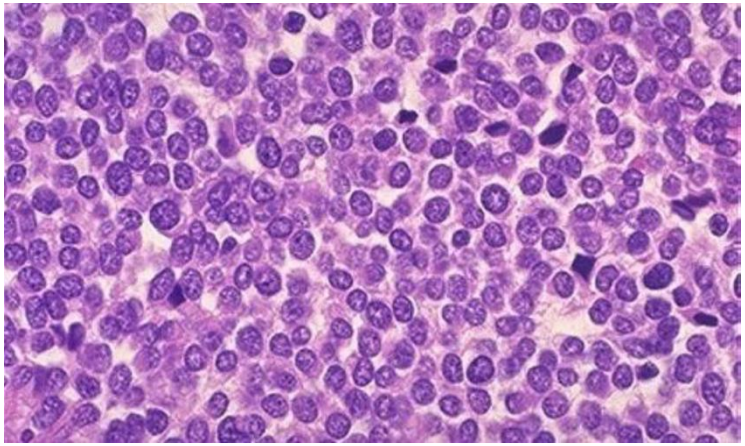
Sol İliak Kanat Osteosarkom







Ewing Sarkom



- Primitive neuroectodermal tumor (PNET)

- Küçük yuvarlak mavi hücreli

Ewing Sarkom-Klinik

- <20 yaş (%80)
- Pelvis, uzun kemik diafiz, scapula
- Sedim, LDH ve WBC yüksekliği
- %25 tanı anında metastaz

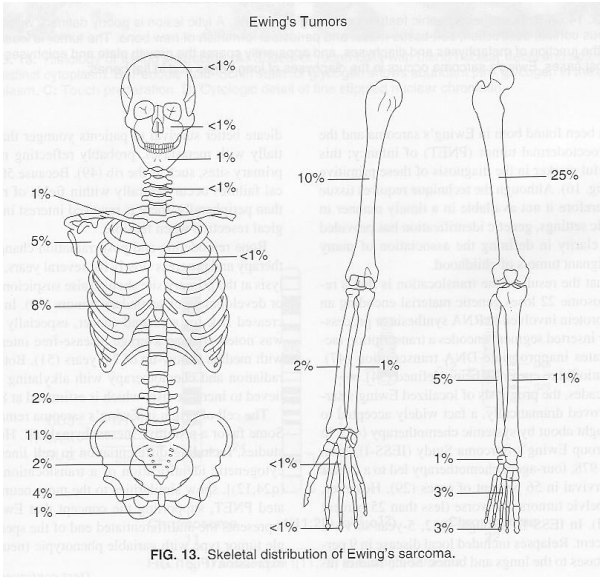
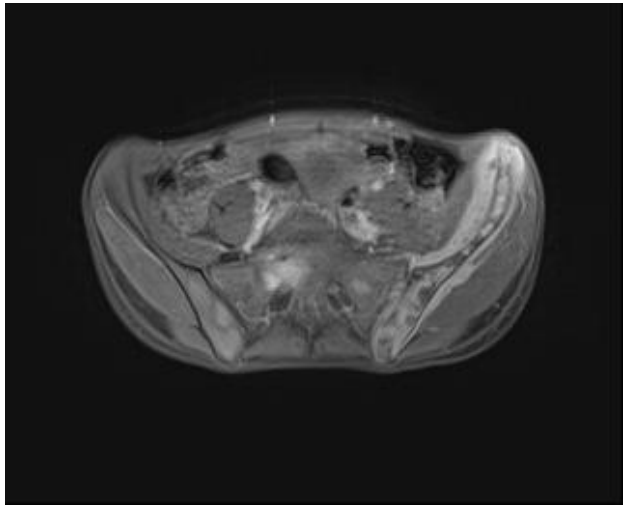
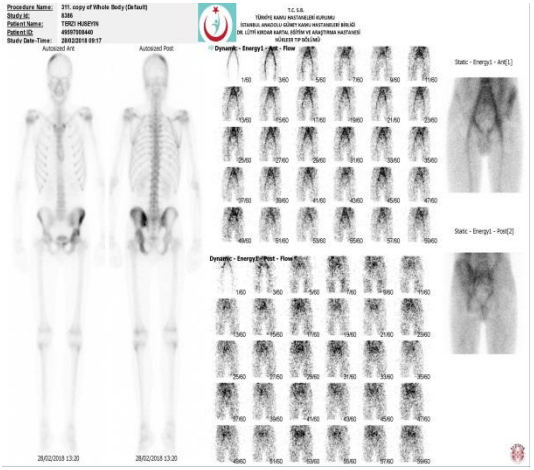
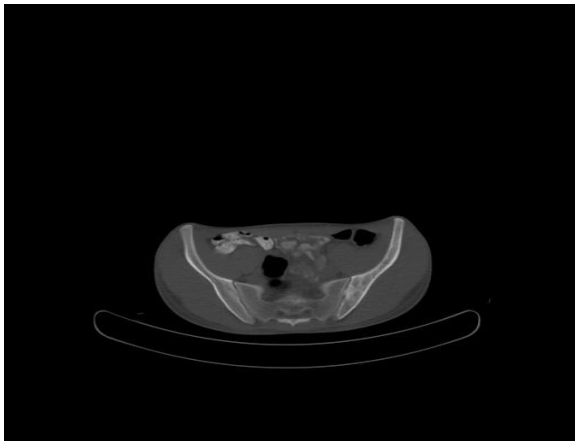


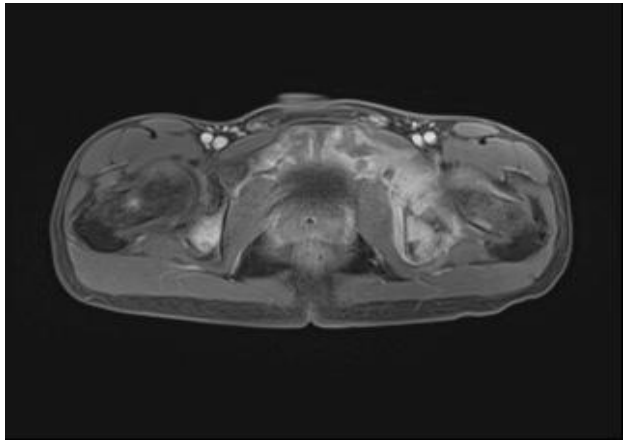
FIG. 13. Skeletal distribution of Ewing's sarcoma.

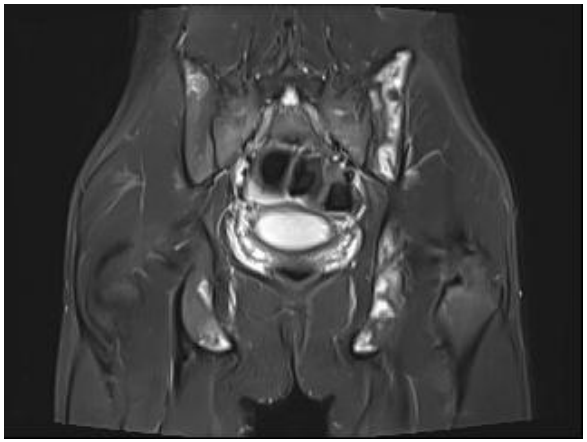
Metastatik Ewing Sarkom











Ewing Sarkom-Radyoloji

- Kötü sınırlı, **permeatif** lezyon
- ‘’**Onion skin**’’ periost reaksiyonu
- Büyük **yumuşak doku** kitlesi
- Ayırıcı tanı: Büyük taklitçi (**Osteomiyelit**, osteosarkom, eozinofilik granülom, osteoid osteoma, lenfoma)

Ewing Sarkom



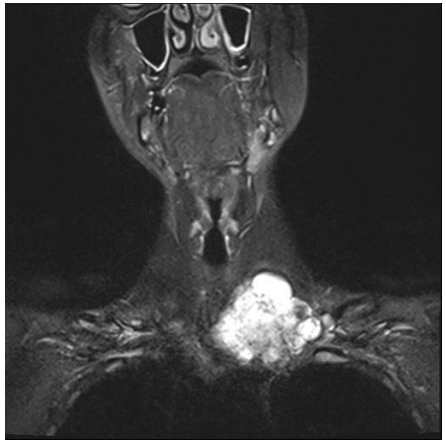

Ewing Sarkom-Tedavi

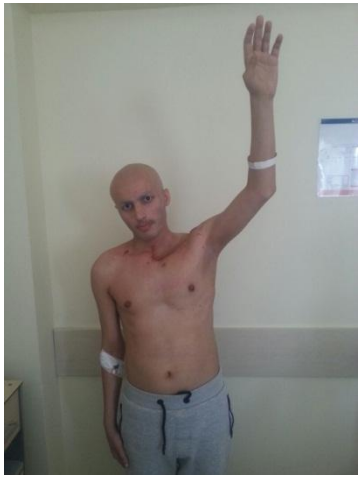
- **Neoadjuvan** (VAC-IE)
  - Vincristine, Adriamycin (Doxorubicin), Siklofosfamid, İfosfamide, Etoposide
- **Geniş rezeksiyon**
- **Adjuvan KT**
- **RT** (definitif-inoperable, adjuvan-pelvis)

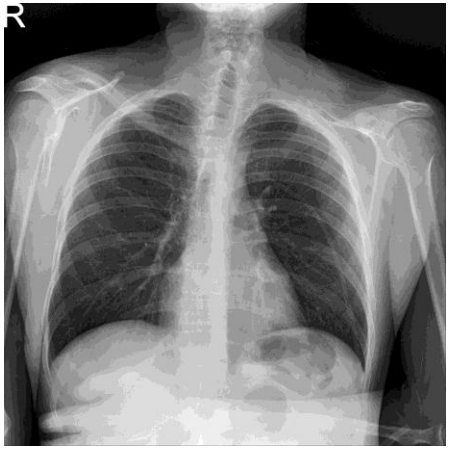
Ewing Sarkom
Kötü Prognostik faktörler

- **Metastaz** varlığı (Ac, Kemik, Kemik iliği)
- **KT'ye yanıt** (<%90)
- **Pelvis/Omurga**

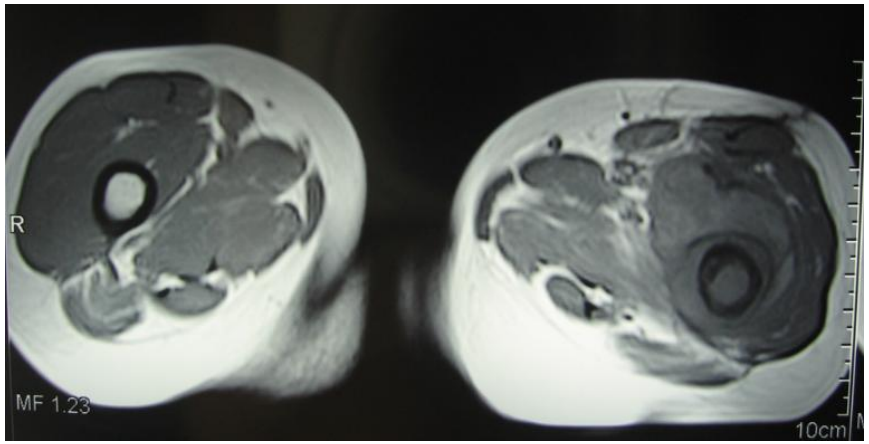
Sol klavikula ewing sarkom



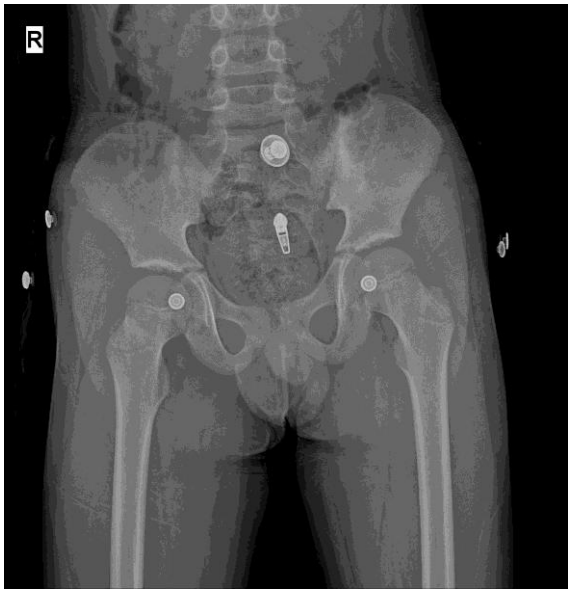


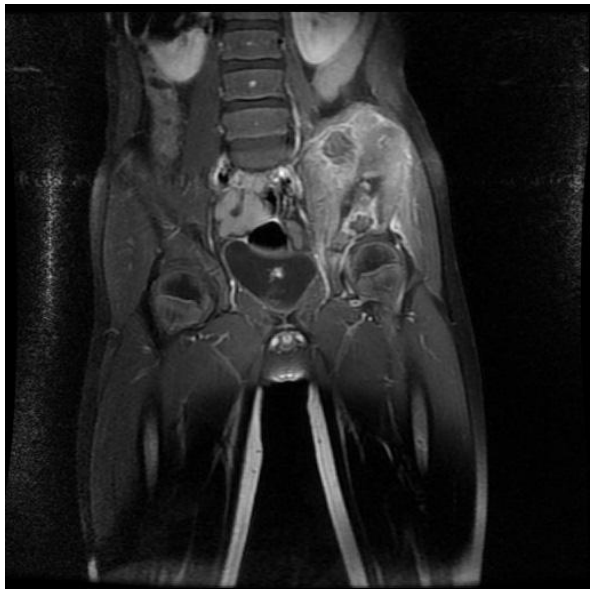


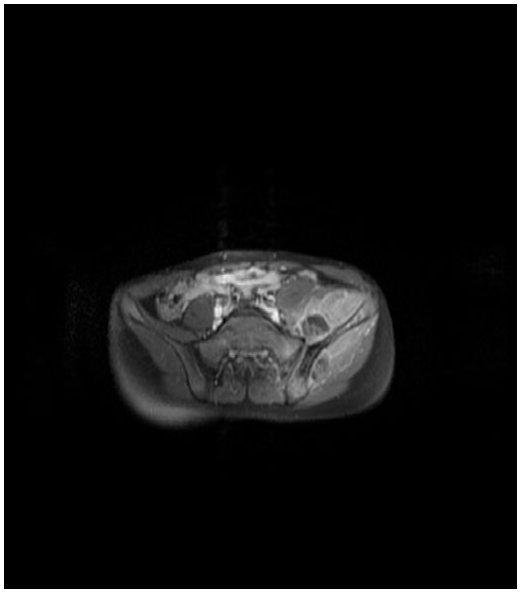
Proksimal femur patolojik fraktür



Sol İliak Kanat Ewing

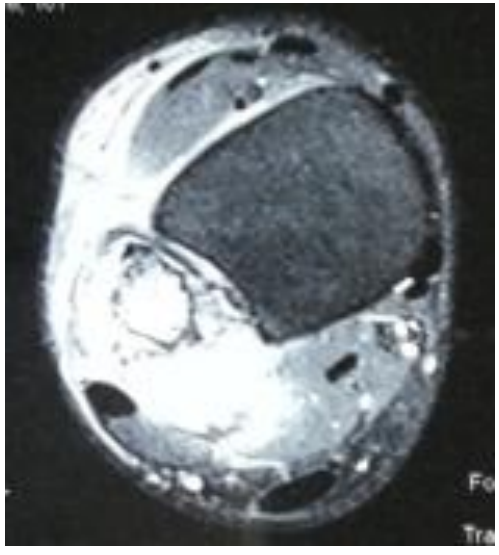






Ayak bileği ağrısı

Distal fibula ewing



MMR PROTOKOLÜ

GÜNLER	1	2	3	8	15
SİSPLATİN 90 mg/m²	X				
ADRİAMİSİN 30 mg/m²	↑	↑			
VİNKRİSTİN 1.5mg/m²	X			X	X
SİKLOFOSFAMİD 10mg/kg	X	X	X		

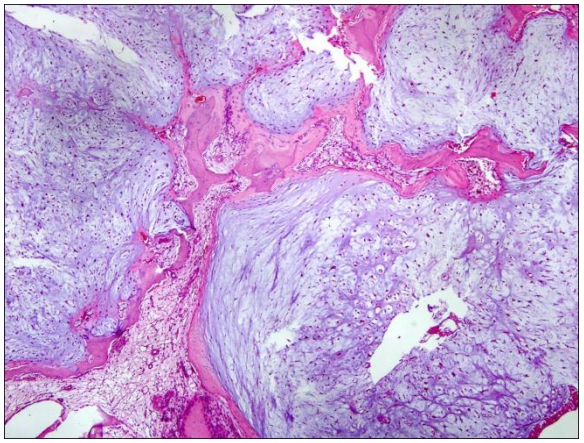
Prof.Dr. Ali VARAN

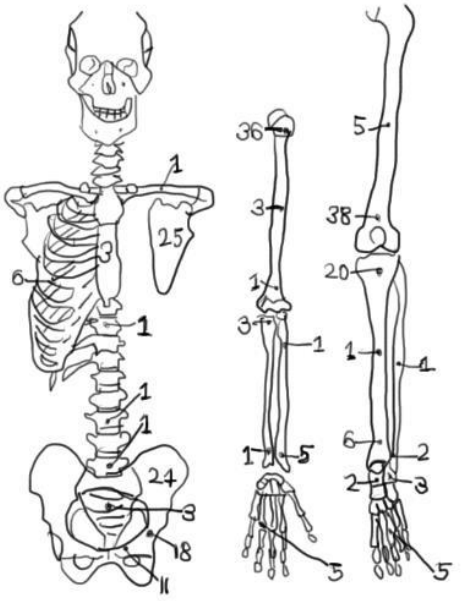
Kemoterapi başlanmadan günlük sıvı alımı en az 3000cc/m² olmalı, idrar dansitesi 1010'un altında olduğunda tedaviye başlanmalıdır. Verilecek sıvı 1/3 SF olmalıdır. Hastanın günlük aldığı-çıkardığı sıvı yakın takip edilmelidir.

- ....135.....mg **SİSPLATİN** .....500.....cc SF içinde IV infüzyonla ışıktan korunarak 4 saatte verilir. Sıvı içine .....5.....cc $MgSO_4$ konulur.
- ...45... mg **ADRİAMİSİN**.....500..... cc %5 dekstroz içinde 4 saatlik infüzyonla ışıktan korunarak verilir.
- .....2.....mg **VİNKRİSTİN** SF ile 10 cc'ye tamamlanarak damara puşe edilir.
- .....600.....mg **SİKLOFOSFAMİD** .....250.....cc SF içerisinde ışıktan korunarak 1 saatte infüzyonla verilir. +Mesna 600mg
- Antiemetik olarak **ONDANSETRON** 5 mg/m2 dozunda kemoterapi başlangıcından 30 dk önce intravenöz yavaş puşe edilir.

Kondrosarkom-Klinik

- 40-75 yaş arası
- Pelvis, proksimal femur, scapula
- Yavaş büyüyen şişlik ve uzun süreli ağrı





Sekonder Kondrosarkom

- **Soliter** Osteokondrom (<%1)
- **Multıpl Herediter** Osteokondromatozis (%5-10)
- **Ollier** Disease (%25-30)
- **Maffucci** Disease (%23-100)

Sekonder Kondrosarkom-Radyoloji

- Osteokondrom şapkasında düzensizlik ve hızlı kalınlaşma (>2cm)
- Osteokondrom sapında destrüksiyon
- Karnıbahar görünümü
- Matriks kalsifikasyonu
- Komşu yumuşak dokudan tam sınırlanamamak

Kondrosarkom -Histolojik Grade

- Grade 1:%60
- Grade 2: %25
- Grade 3: %5
- Dedifferentiated: %10

- **İğne biyopsisi grade tespiti için uygun değil**

Kondrosarkom-Radyoloji

- **Grade I:**

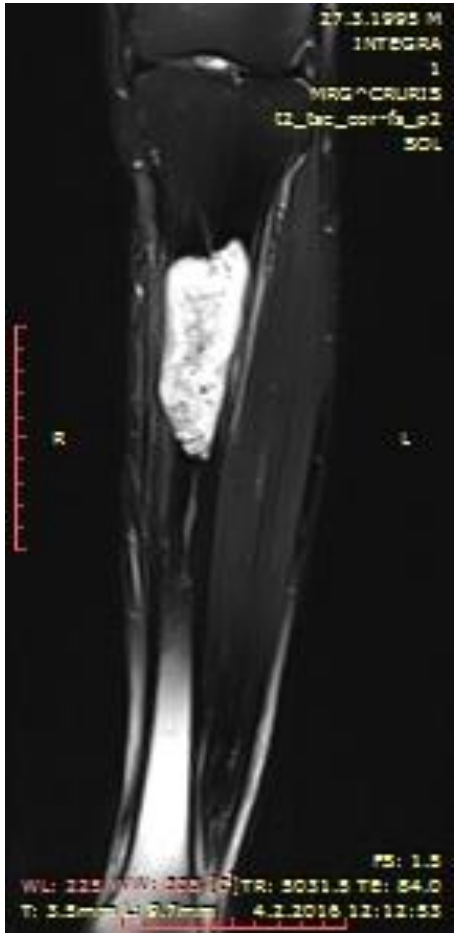
➢ Kortikal kalınlaşma/ekspansiyon

➢ Endosteal erozyon

➢ >8-10 cm

➢ Yüzük/Ark şekilli kalsifikasyon

Grade I Kondrosarkom



Kondrosarkom-Radyoloji

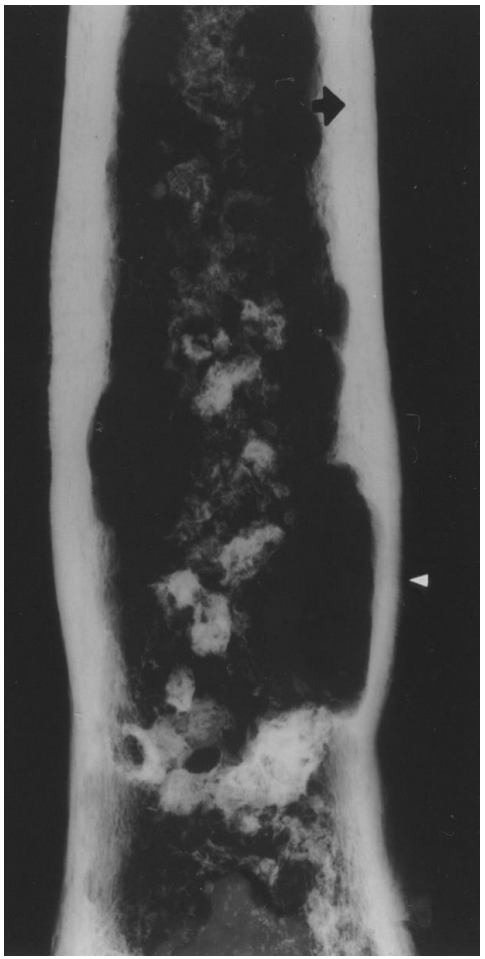
- **Grade II-III:**

➢ Kortikal destrüksiyon

➢ Yumuşak doku uzanımı

➢ Artmış sintigrafik tutulum

Grade II Kondrosarkom



Kondrosarkom Tedavi

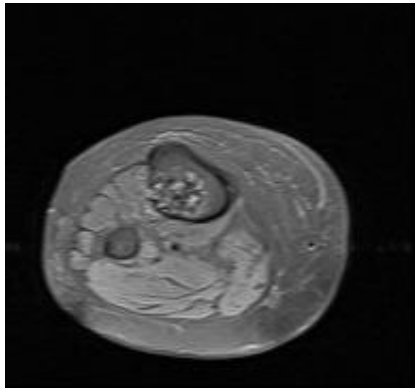
- Grade **I**: İntralezyonel küretaj/Geniş rezeksiyon

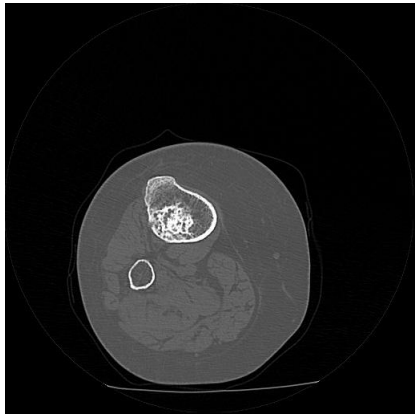
➢ Pelvik Grade I kondrosarkom: Geniş rezeksiyon

- Grade **II-III**: Geniş rezeksiyon

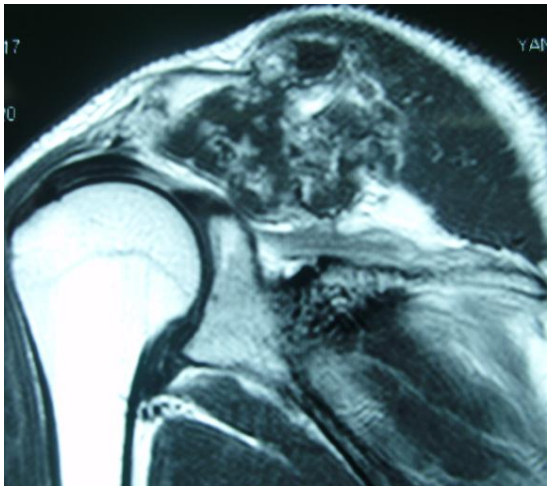
- **Dediffrentiated/Mezenkimal**: KT+Geniş rezeksiyon

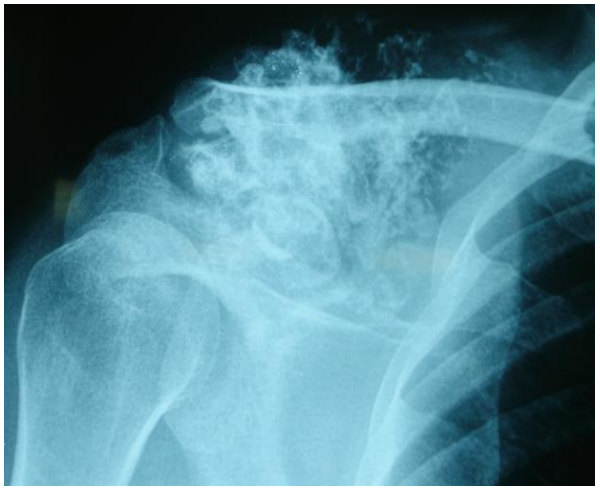
Grade I Kondrosarkom

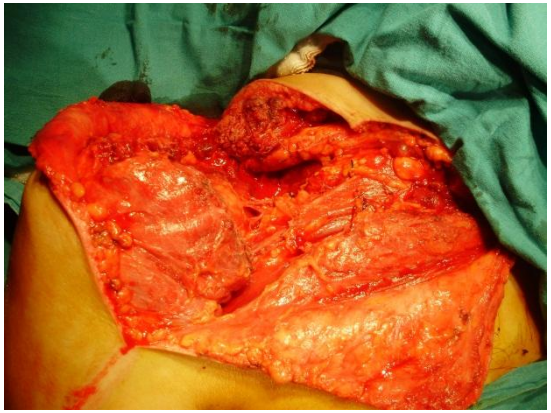


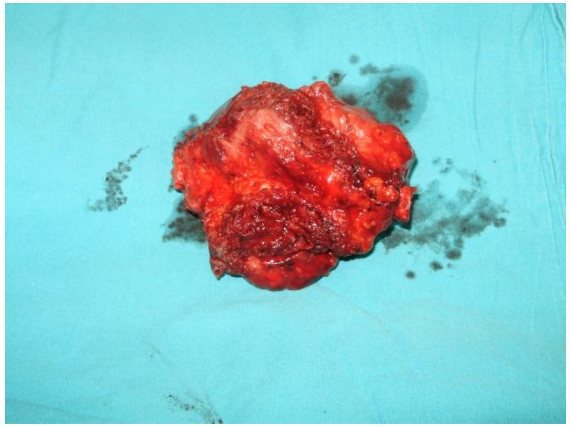


Distal Klavikula Grade II Kondrosarkom

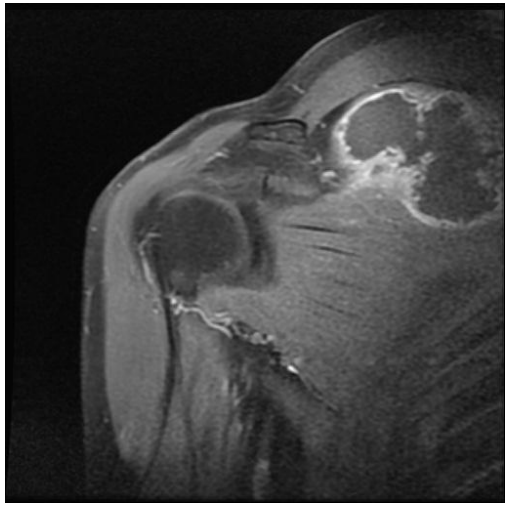


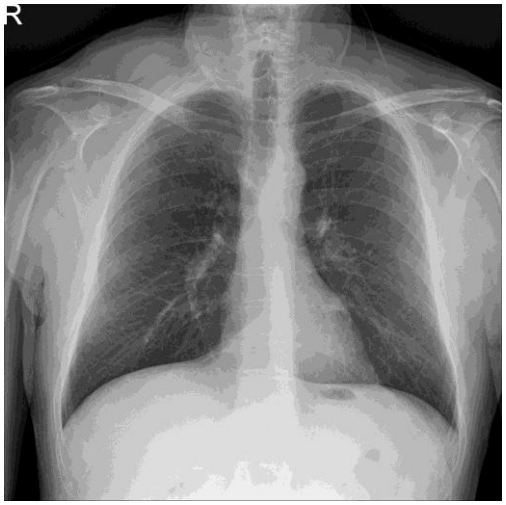


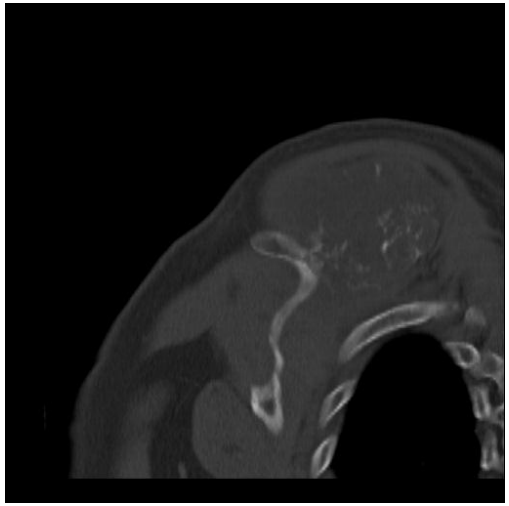


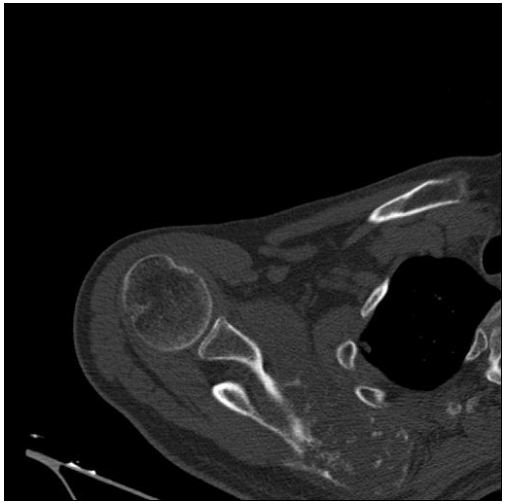


Scapula Grade II Kondrosarkom

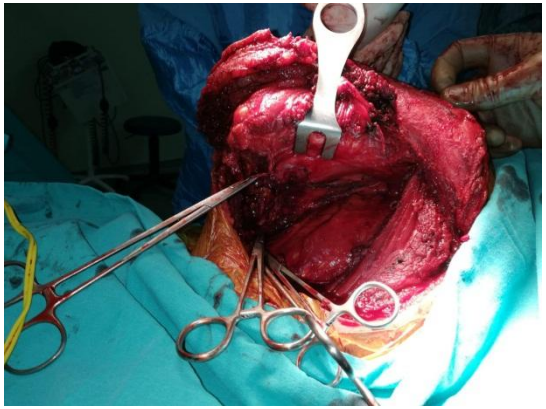


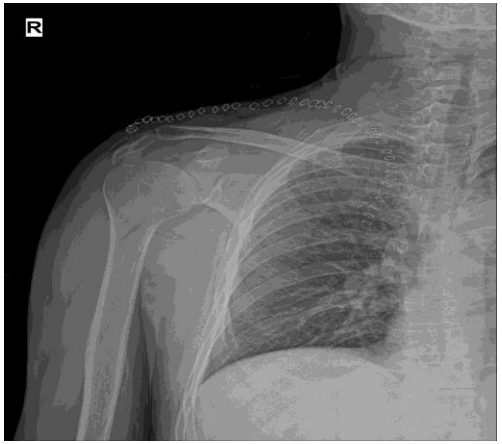


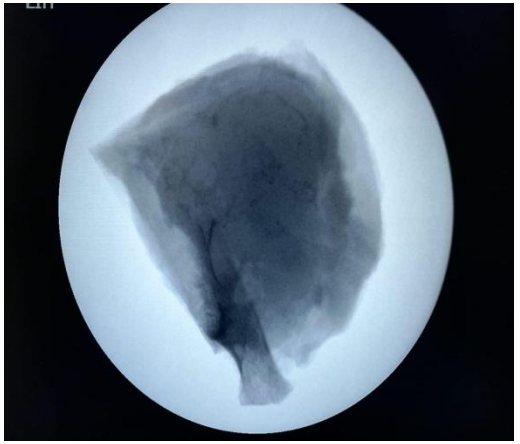




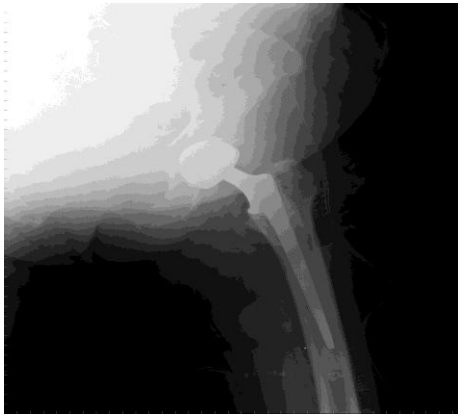
Skapulektomi

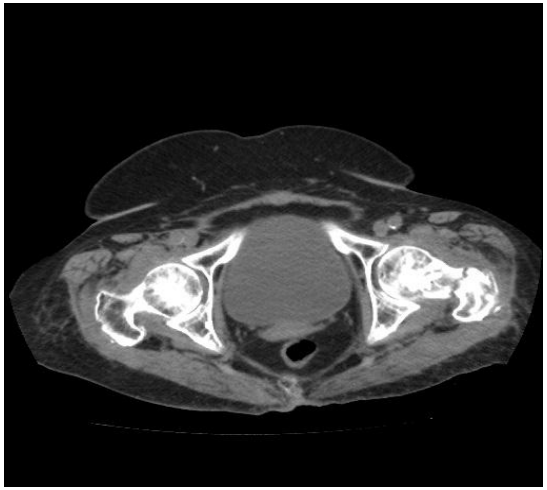




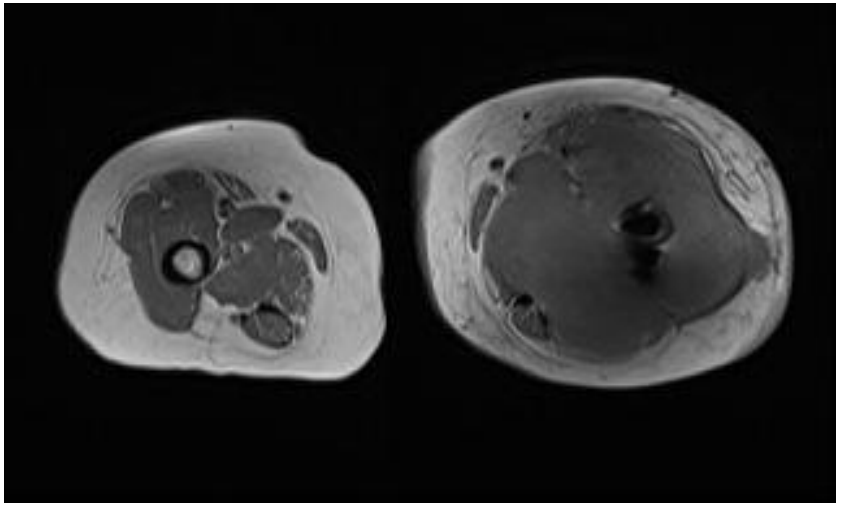


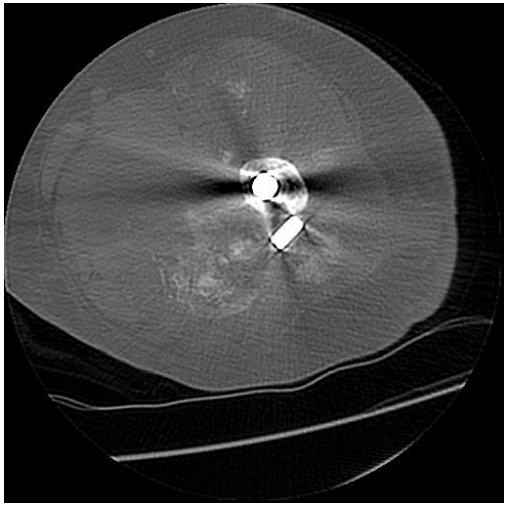
Sol Femur Boyun Fraktürü?

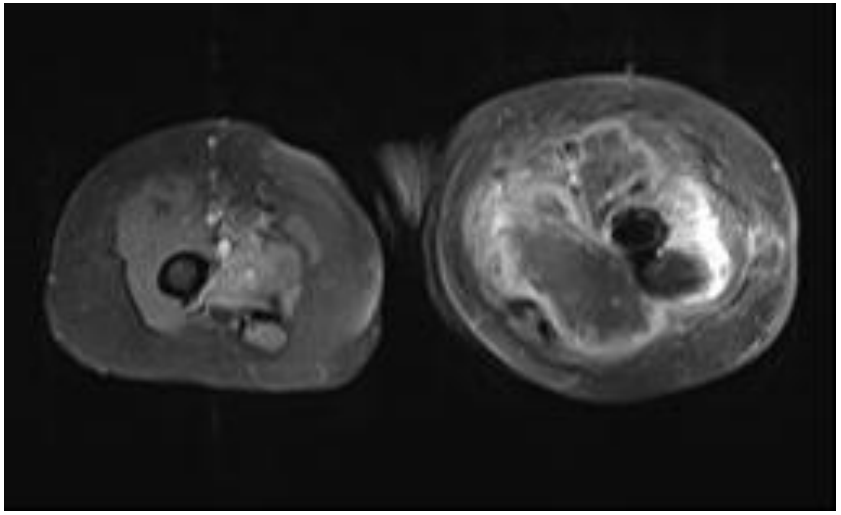


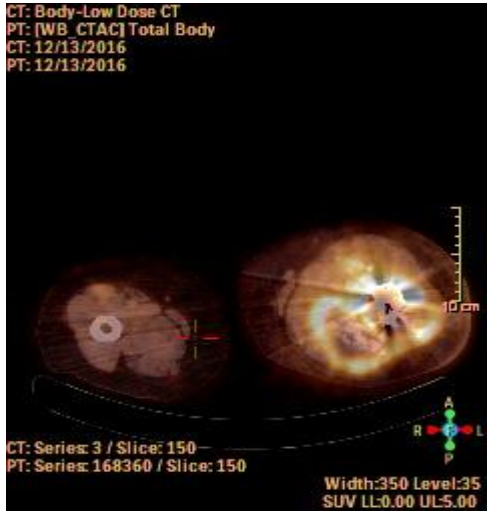


Dediferansiye Kondrosarkom

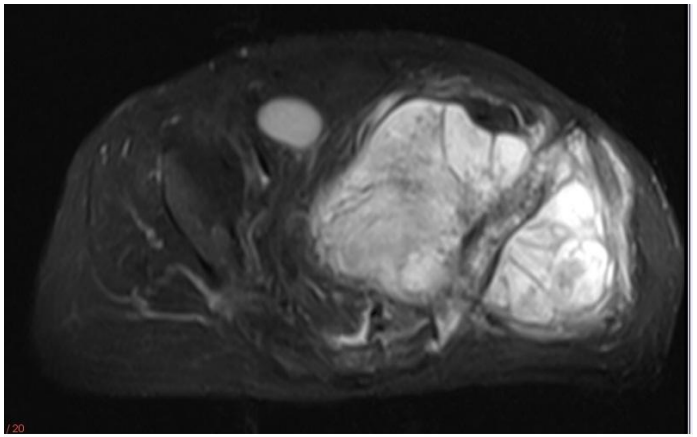


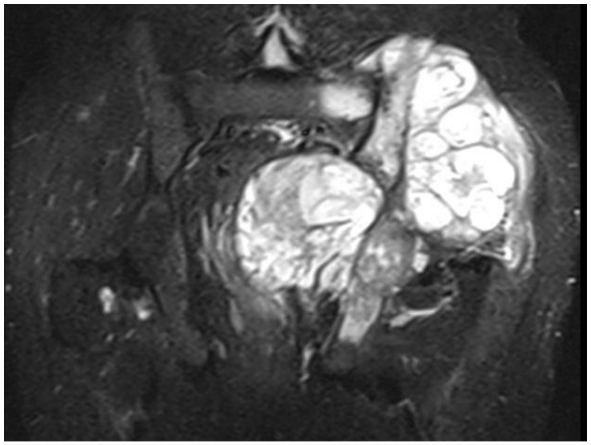


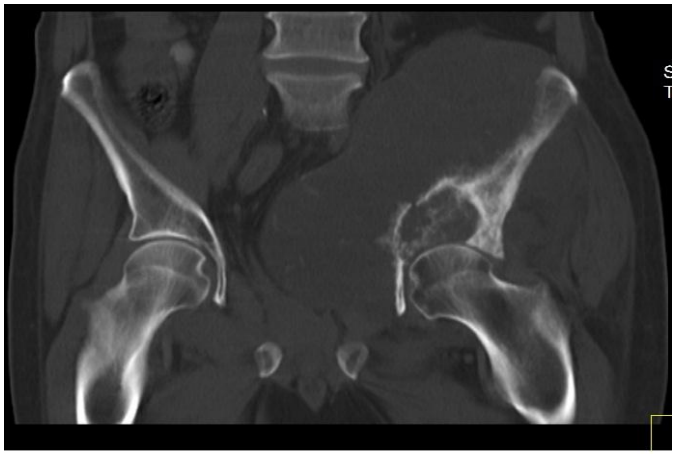




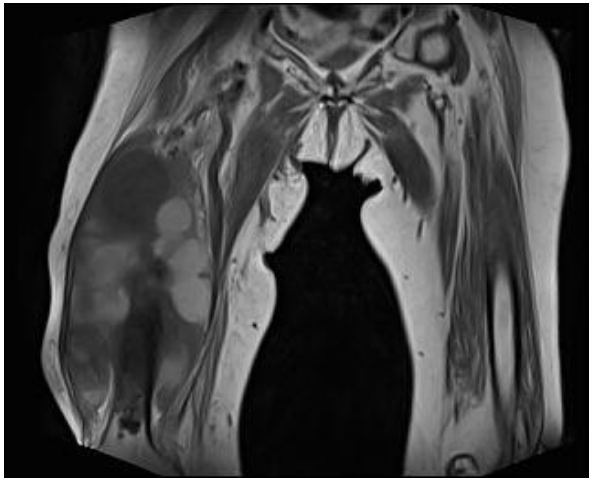
Grade III Kondrosarkom

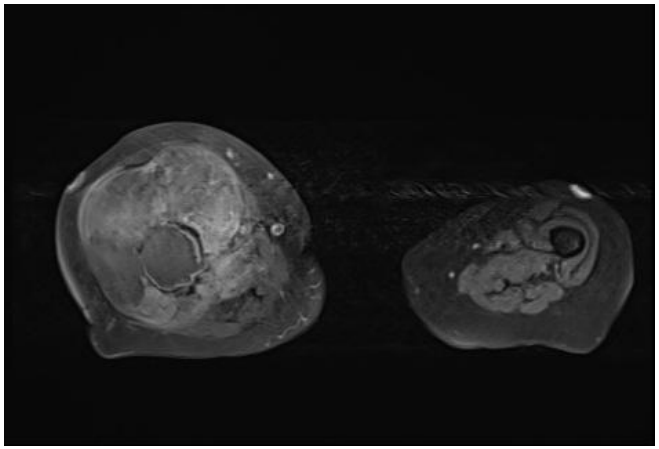


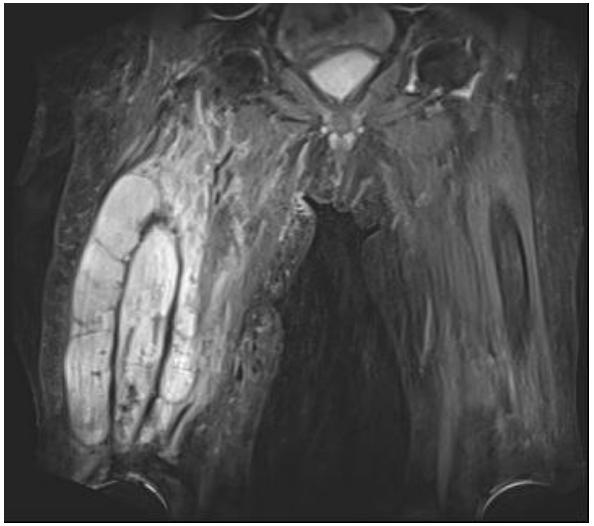




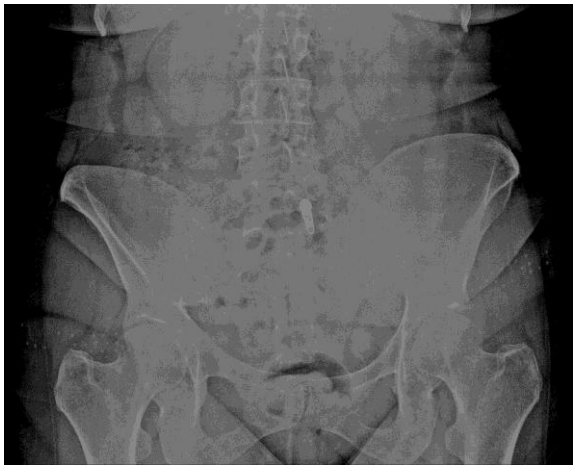
Grade III Kondrosarkom

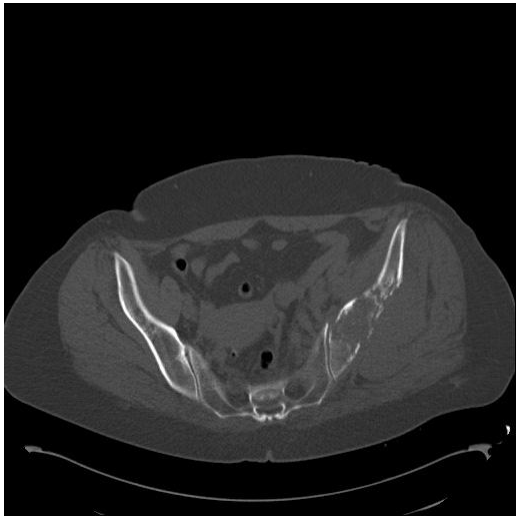


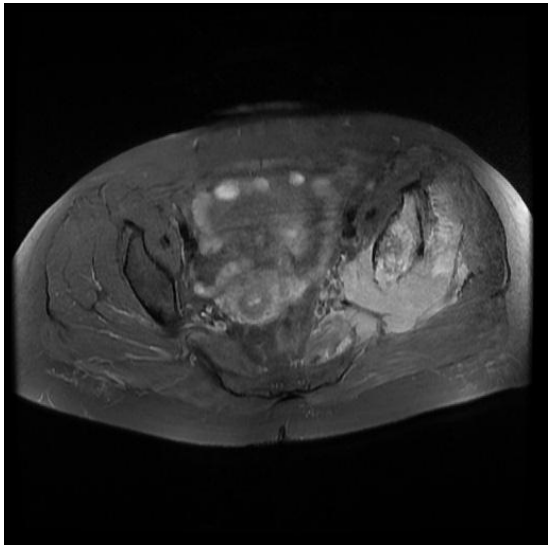




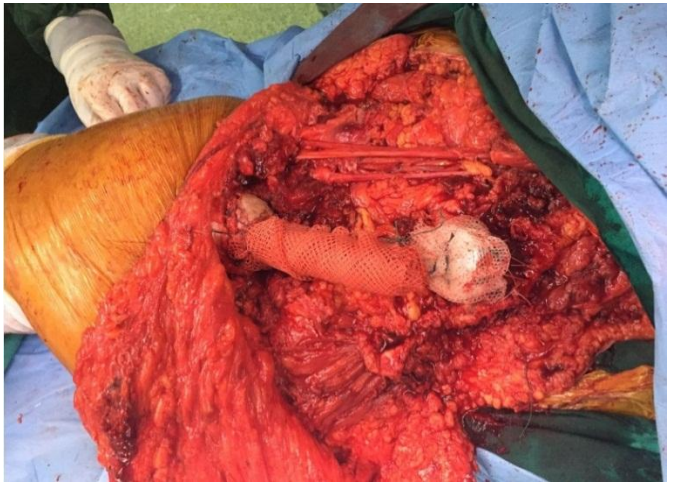
Sol Hemipelvis
Dediferansiye Kondrosarkom

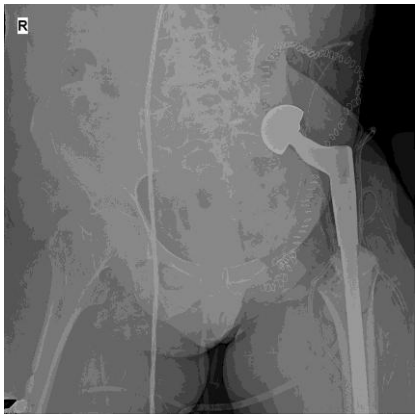






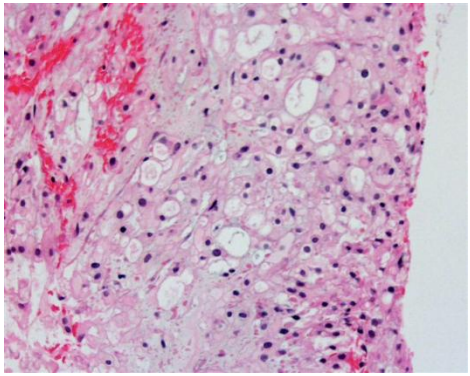
Hip Transpozition





Kordoma

- Notokordal kalıntıdan kaynaklı
- **>40 yaş**
- Omurga (%50 **sakrum**, %35 kafa tabanı, %15 mobil vertebra)
- Yavaş büyüyen (**Geç tanı**)
- Bası semptomları (kabızlık, inkontinans)



Kordoma-Görüntüleme

- Pelvis grafisinde **tanı zor** (barsak gazları), koksiks lateral grafi alınmalı

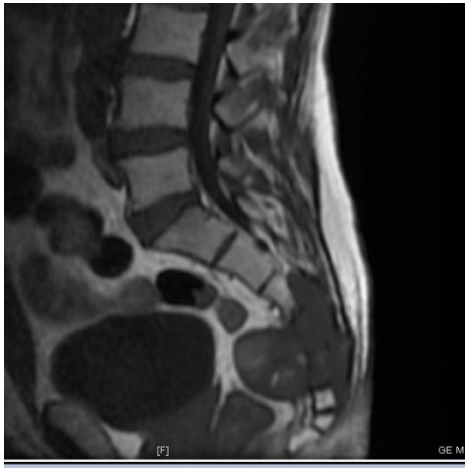
- BT: Kalsifikasyon, destrüksiyon

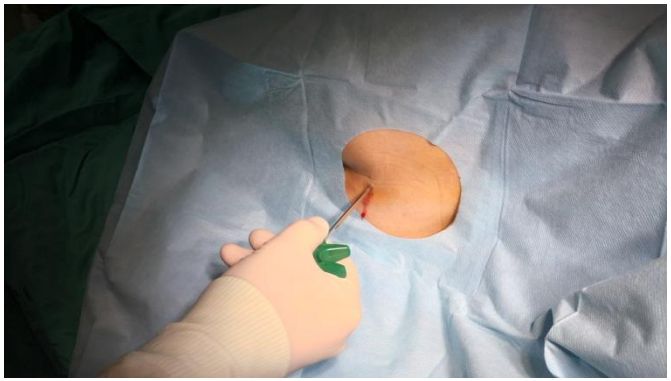
- **MR**: Çoklu seviye tutulumu, büyük yumuşak doku

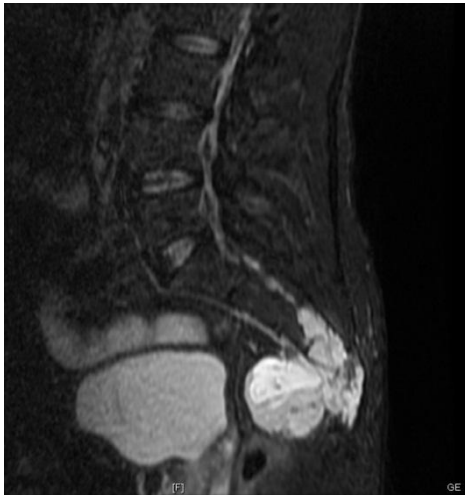
Kordoma-Tedavi

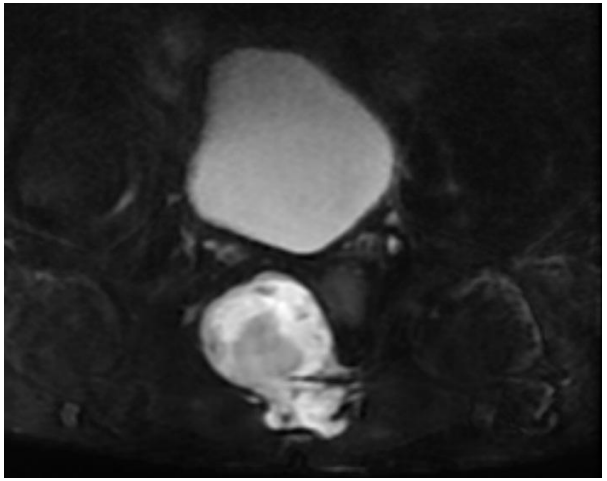
- Geniş rezeksiyon
- Nüks sık (%50)
- KT rutin uygulanmaz
- RT: Adjuvan, Primer (İnop)
- Geç metastaz (Ac, Kemik) nedeniyle uzun süreli (20 yıl) takip

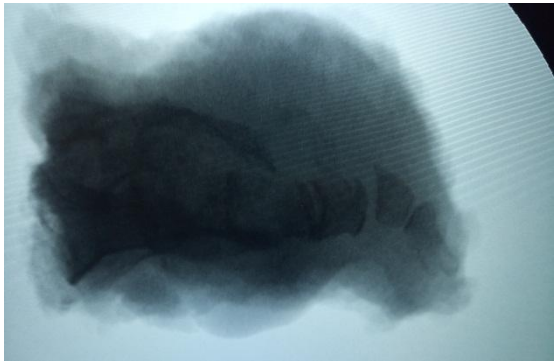
Kordoma

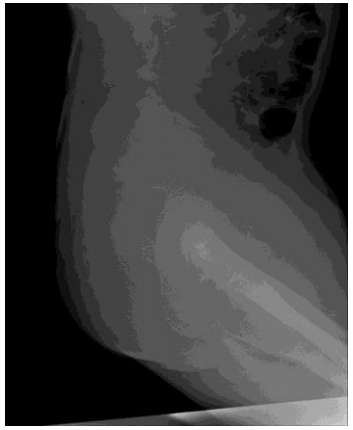




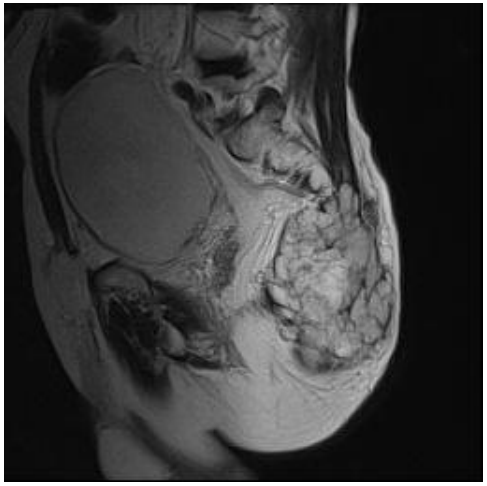


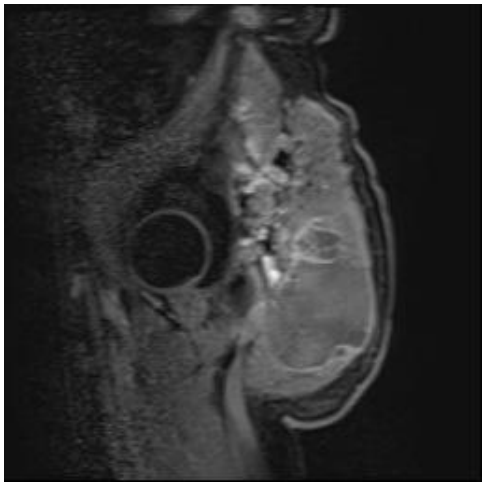


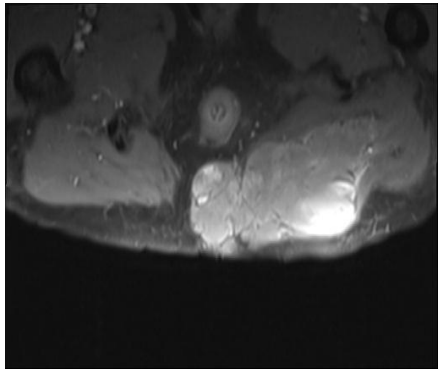


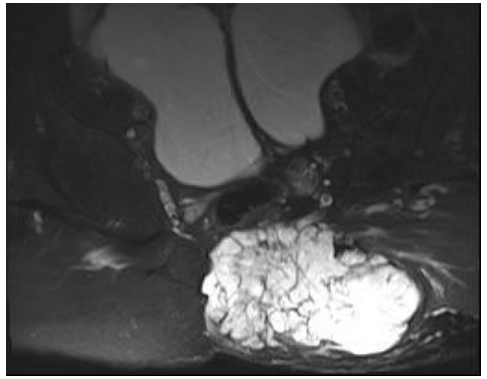


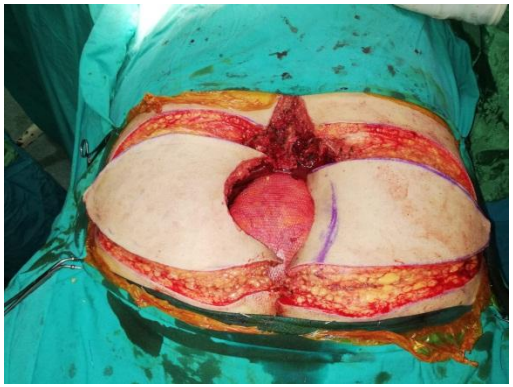
Kordoma

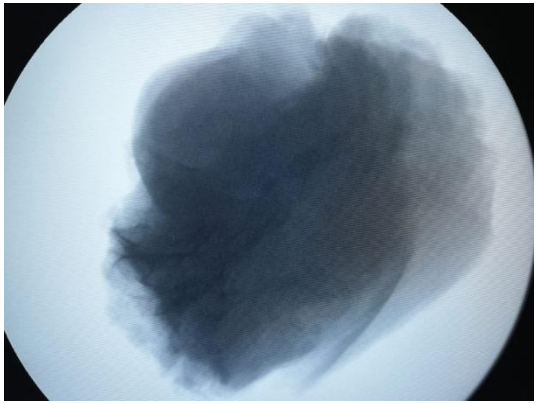


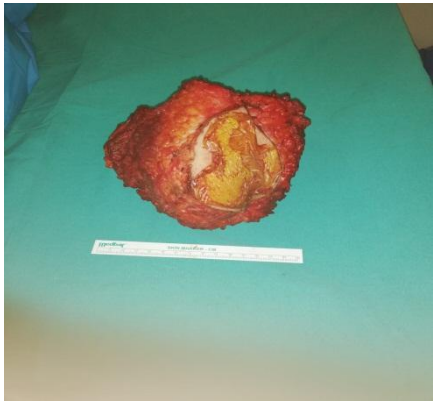






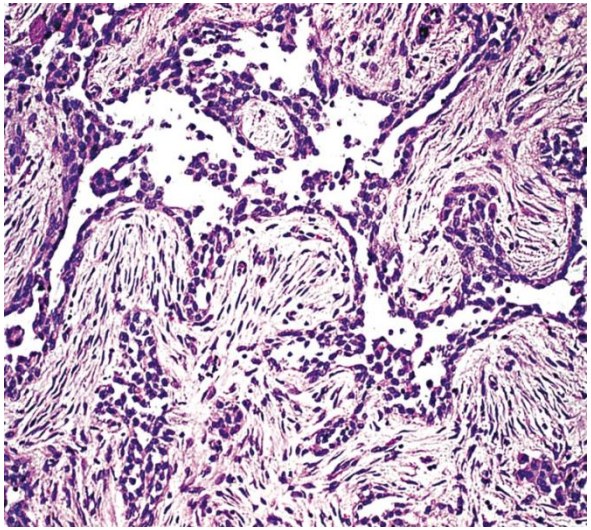






Adamantinoma-Klinik

- **20-40 yaş**
- **Tibia diafiz ön yüz**
- Lokal hassasiyet ve **deformite**
- Yavaş ilerler (Uzun süreli hikaye)



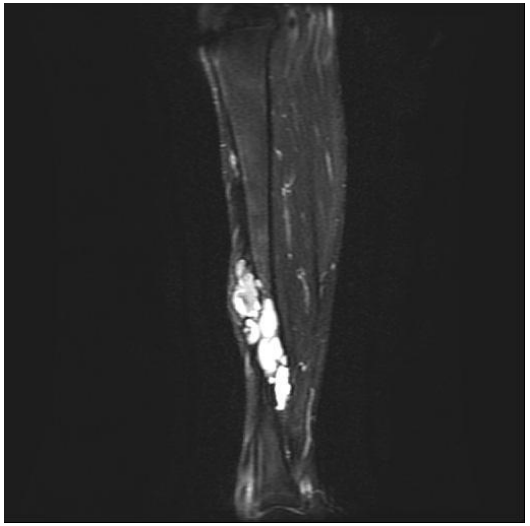
Adamantinoma-Radyoloji

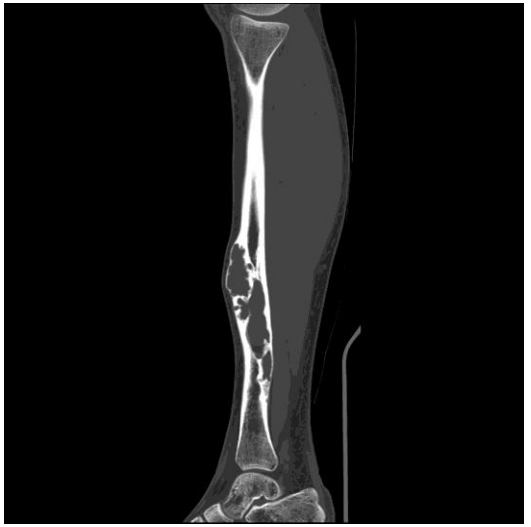
- Çoklu iyi sınırlı **litik** lezyonlar.(Bir tanesi major)
- **Sabun köpüğü** görünümü
- Periost reaksiyonu görülmez
- Çok nadiren (%10) yumuşak doku kitlesi

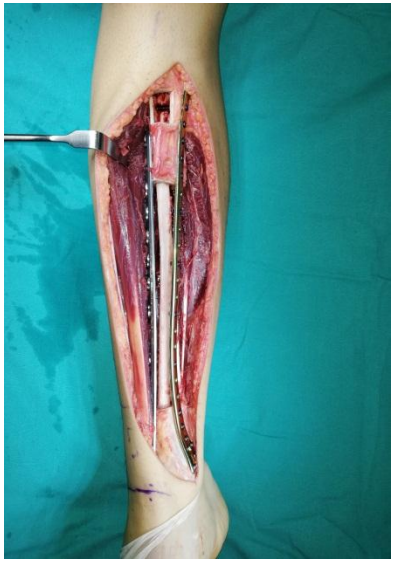
Adamantinoma-Tedavi

- Geniş rezeksiyon
- KT ve RT uygulanmaz
- Geç metastaz (%15-20 Ac, Lenf nodu) nedeniyle uzun süreli takip

Adamantinoma

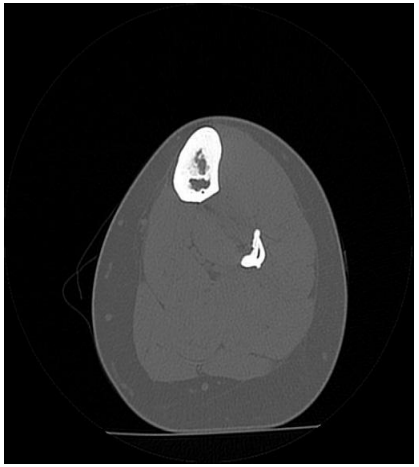


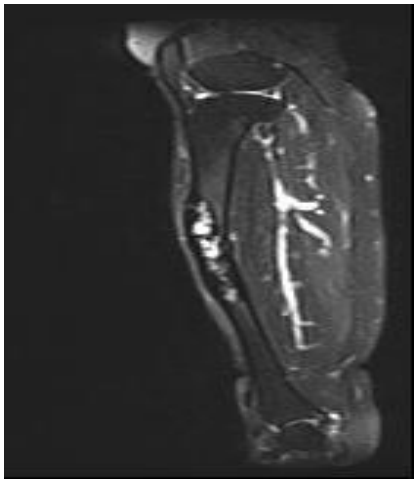


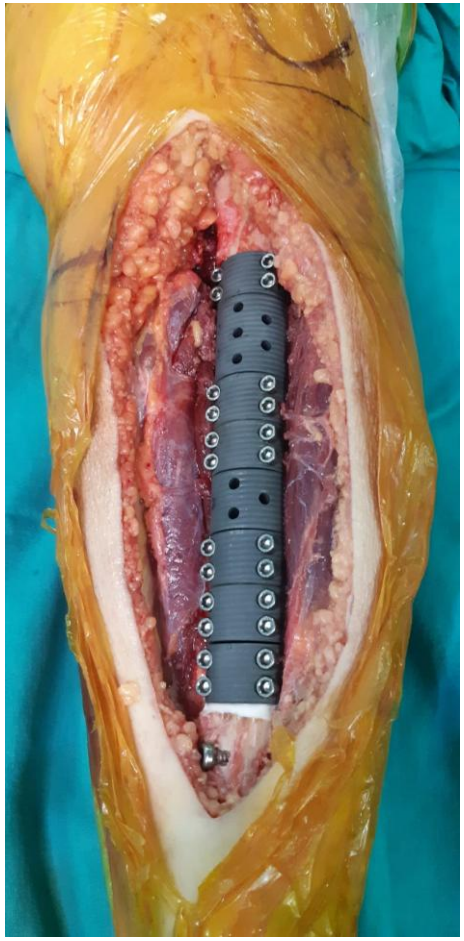



R

Adamantinoma







Postradyasyon Sarkomu

- Her hangi bir tümör zemininde gelişebilir
- Çocukluk çağında maruz kalanlarda daha sık (20 yıllık süreçte %5-10 risk)
- Eş zamanlı KT (özellikle alkali ajanlar) uygulanmış olması riski artırır

Postradyasyon Sarkomu

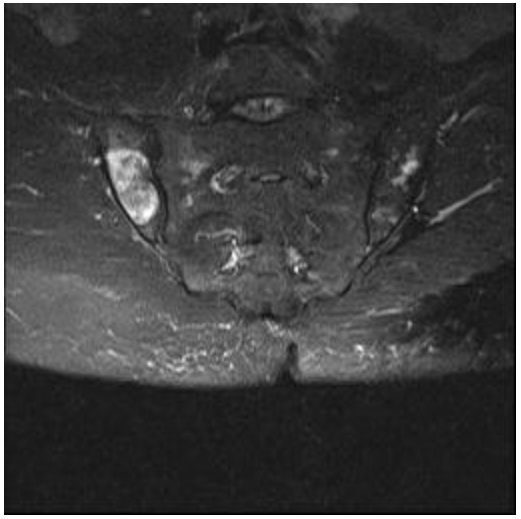
- Litik, destrüktif lezyon

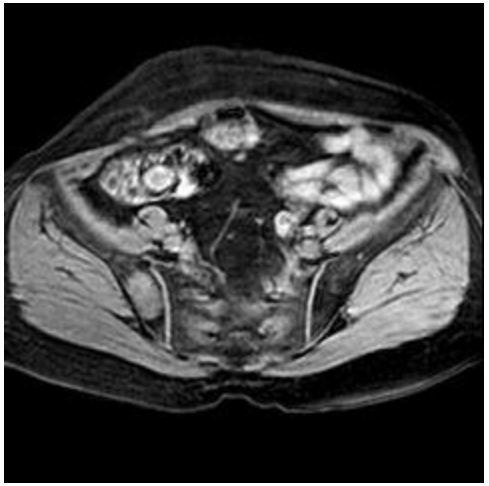
- Histoloji: **Yüksek grade** sarkoma (osteosarkom, undiferansiye pleomorfik sarkom, fibrosarkom, kondrosarkom)

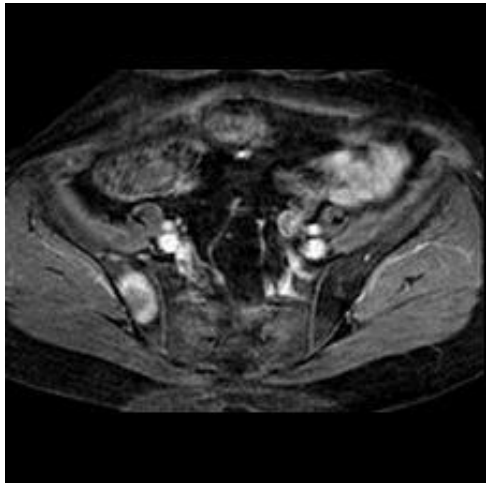
Postradyasyon Sarkomu

- Tedavi: KT+Geniş rezeksiyon
- RT uygulanabilir
- Kötü prognoz (5 yıllık %25-50)

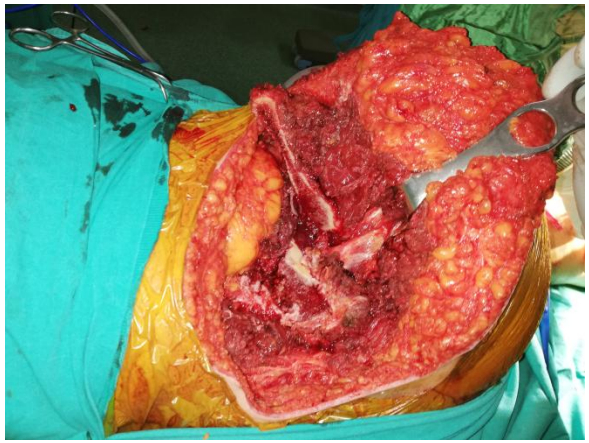
Postradyasyon Sarkomu (IPM)

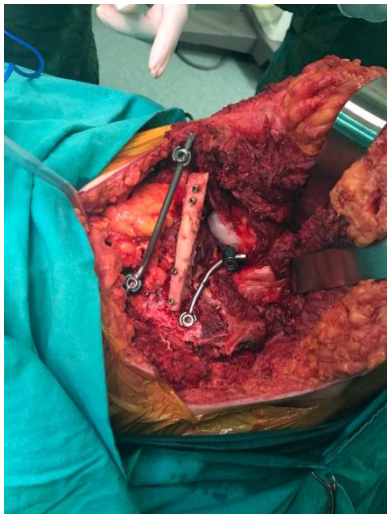


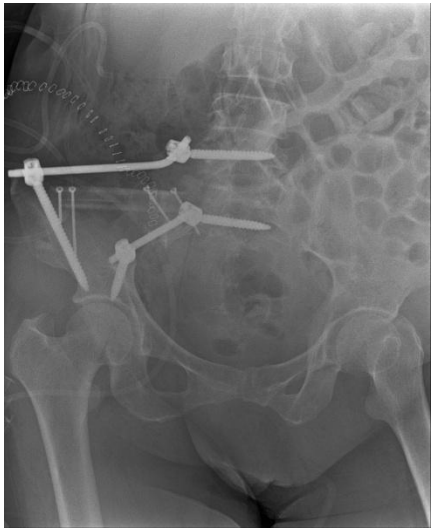


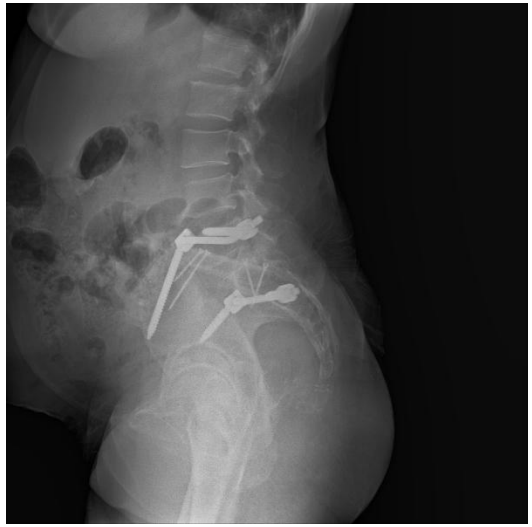


Postradyasyon Sarkomu (IPM)









Sonuç

- Şüphe

✓ Israrcı/İstirahat-Gece ağrısı?

✓ Patolojik kırık? (öncesinde ağrı, minimal travma, litik lezyon)

✓ Periost reaksiyonu?

- Ek tetkik

✓ Tomografi, Kontrastlı MRI, Sintigrafi

- Biyopsi?

Kötü Huylu Kemik Tümörleri

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Dr. Lütfi Kırdar Kartal E.A.H

Teşekkür Ederim