

Pediatric Over Tumörleri Olgu sunumu

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Başkent Üniversitesi Tıp Fakültesi

Patoloji AD



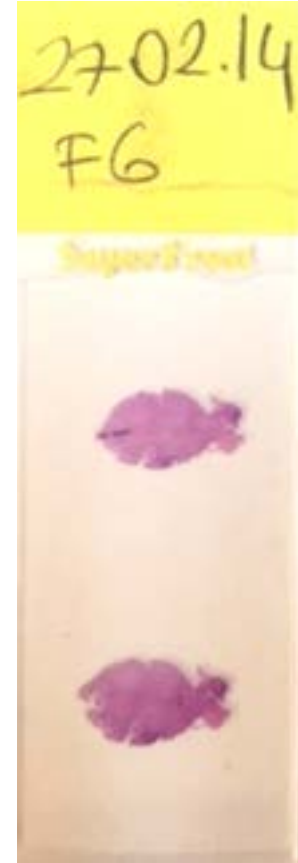
25 Ulusal Patoloji Kongresi
6 Sitopatoloji Kongresi

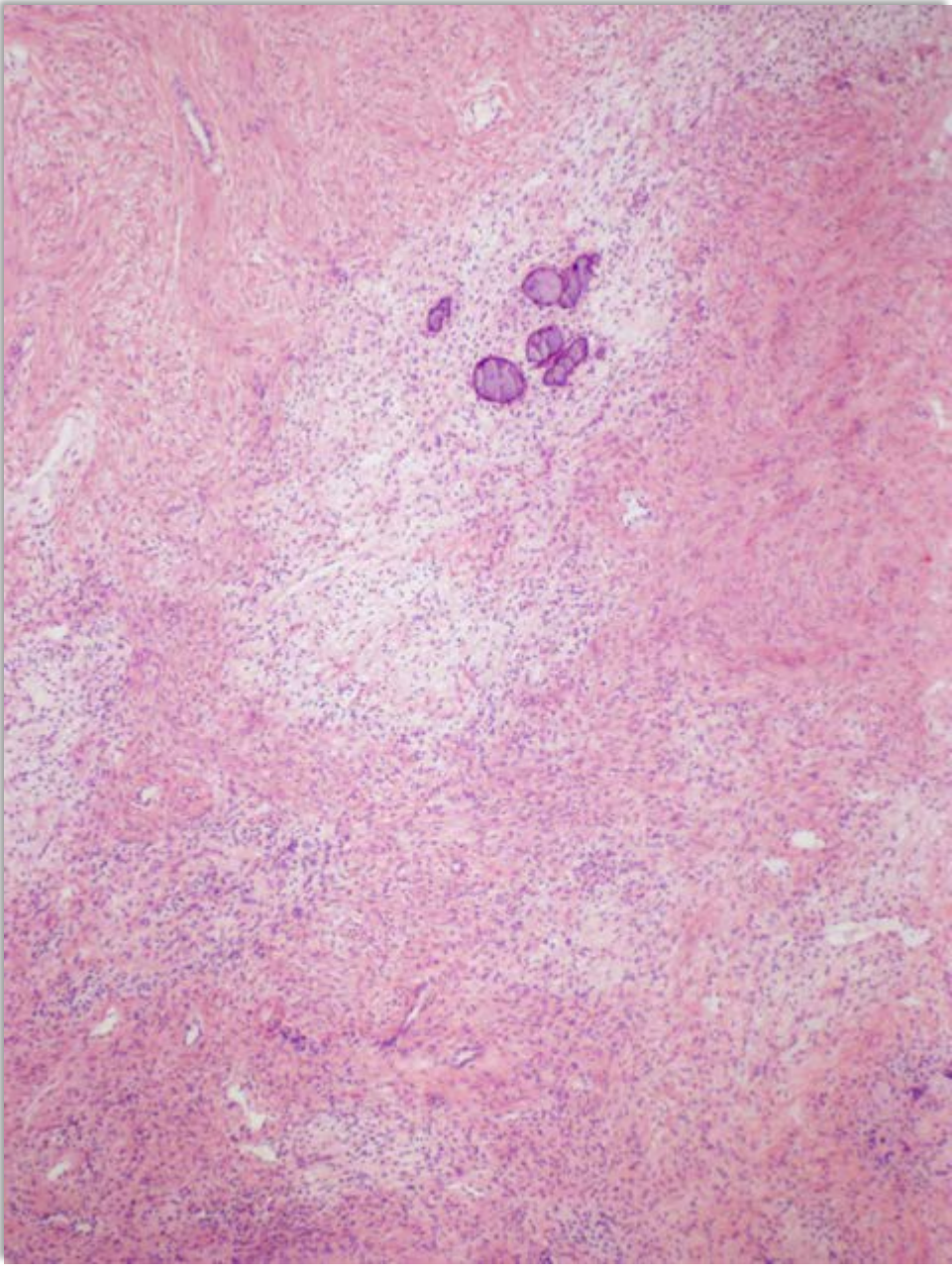
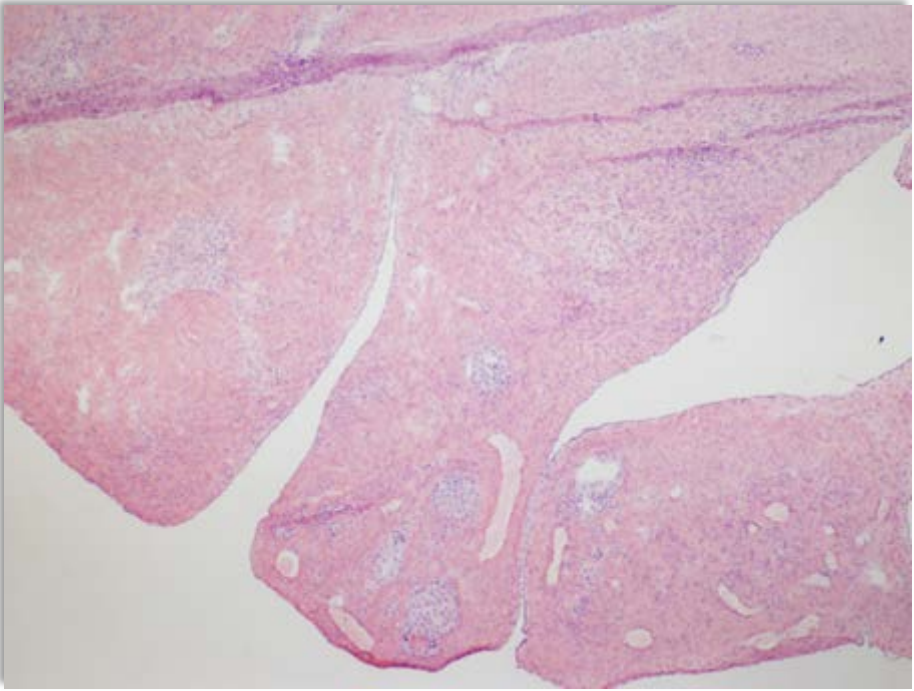
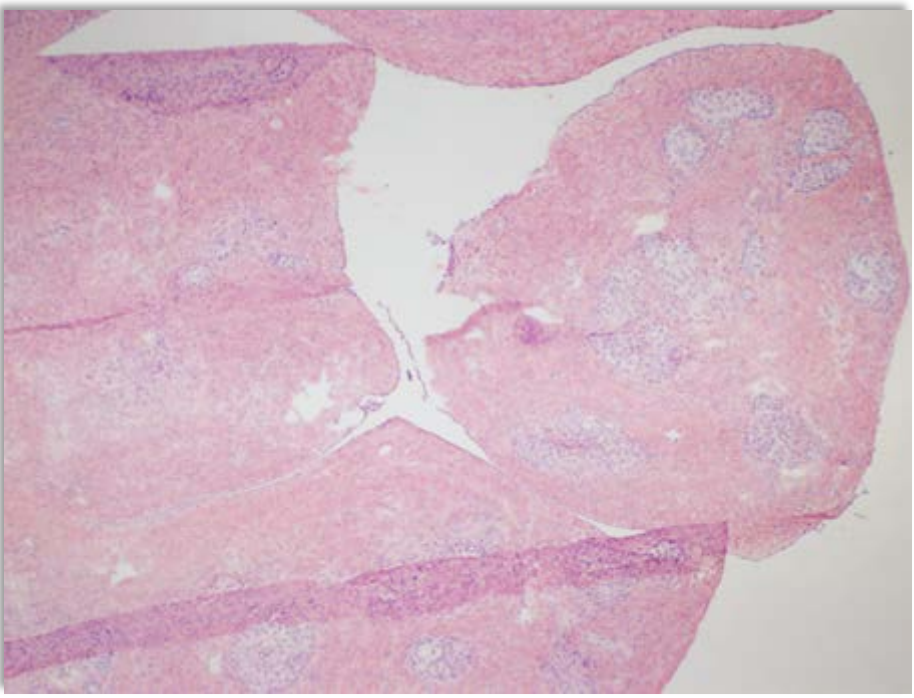
14 - 17 Ekim 2015 / Merinos AKKM - BURSA

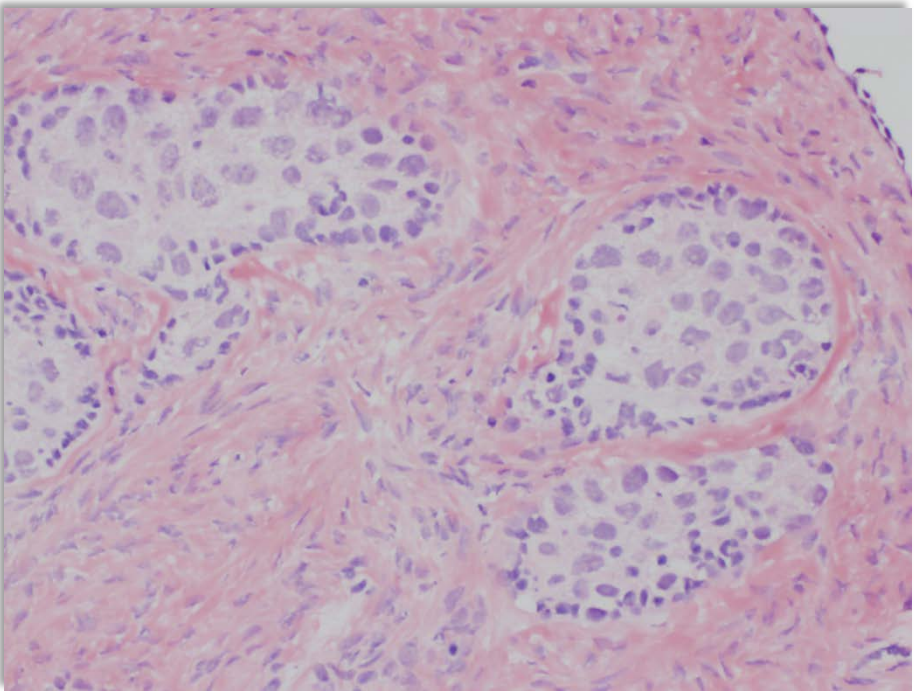
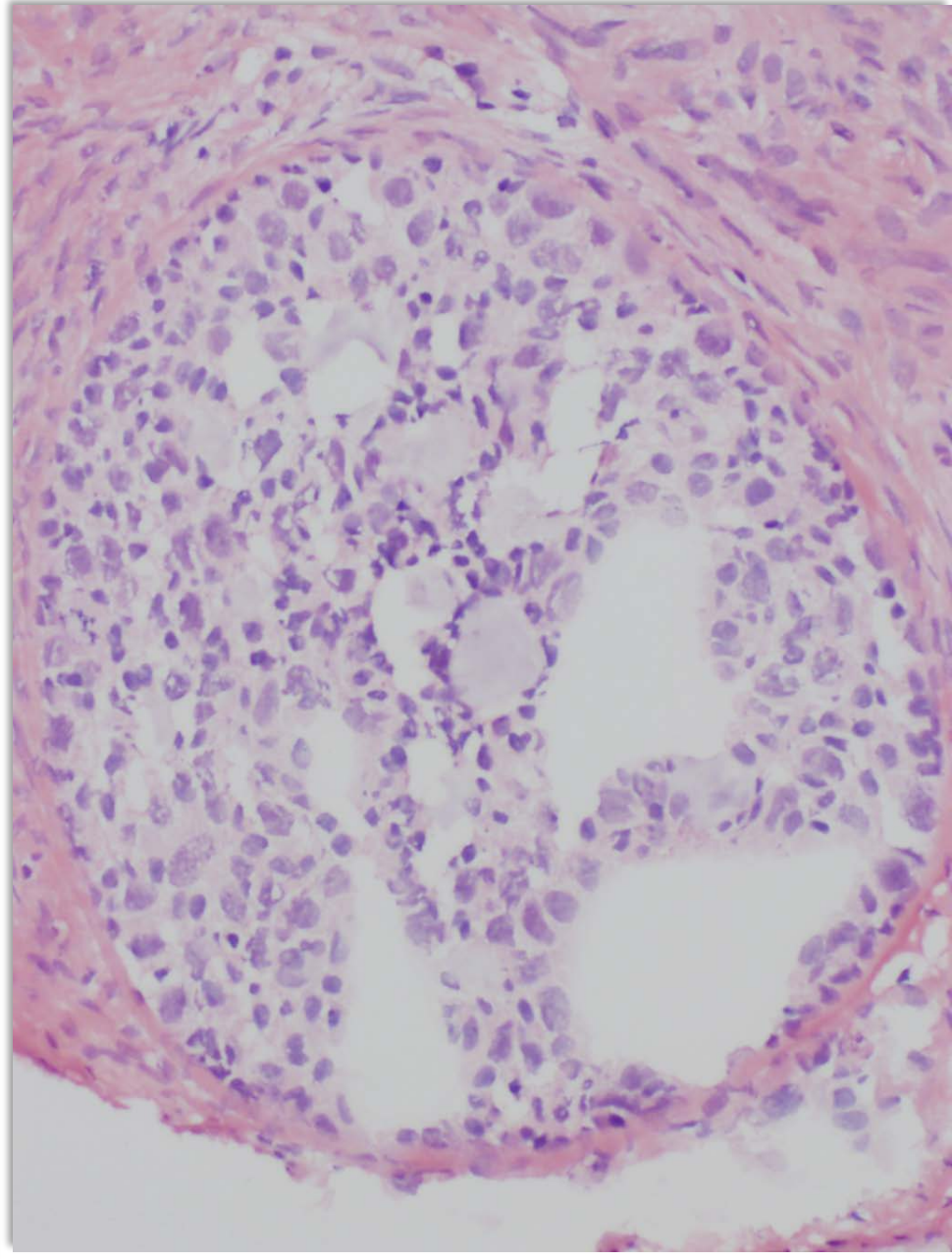
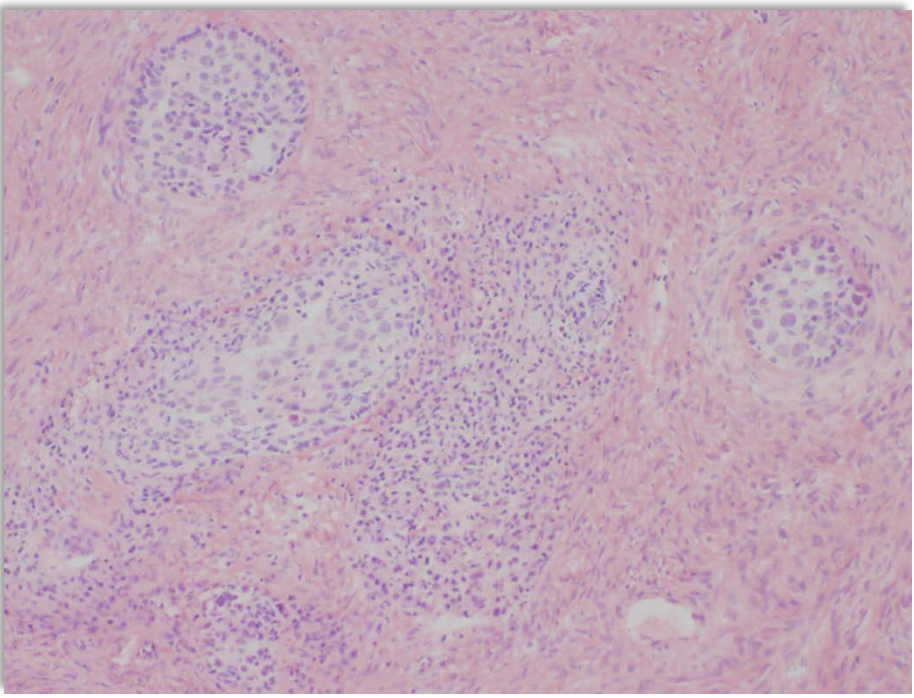


Olgu Sunumu

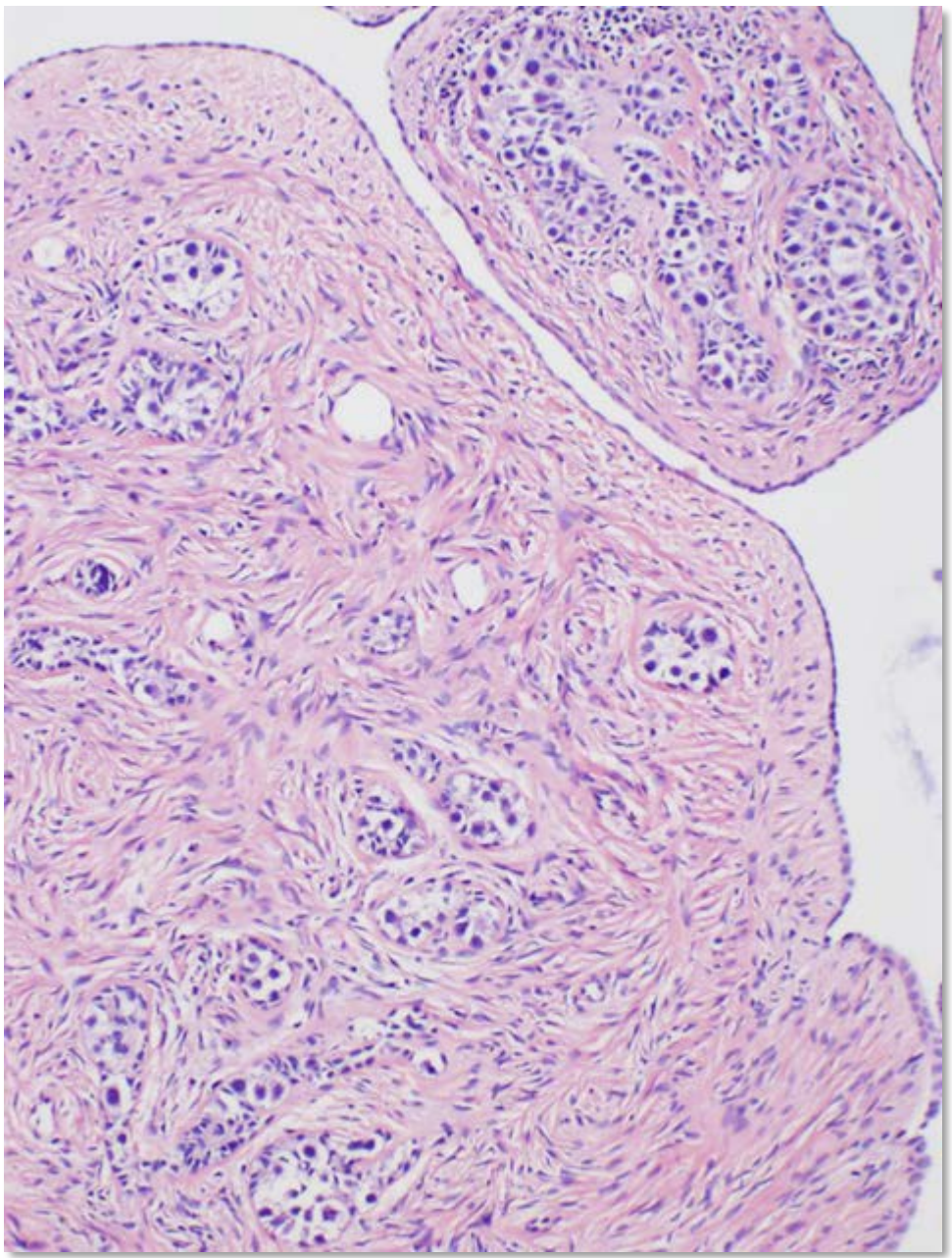
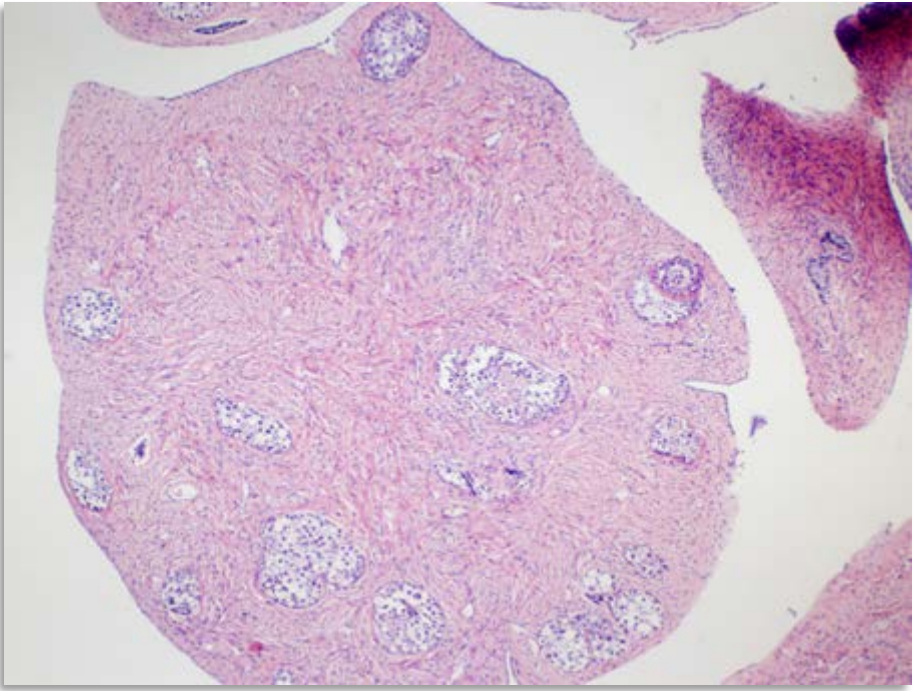
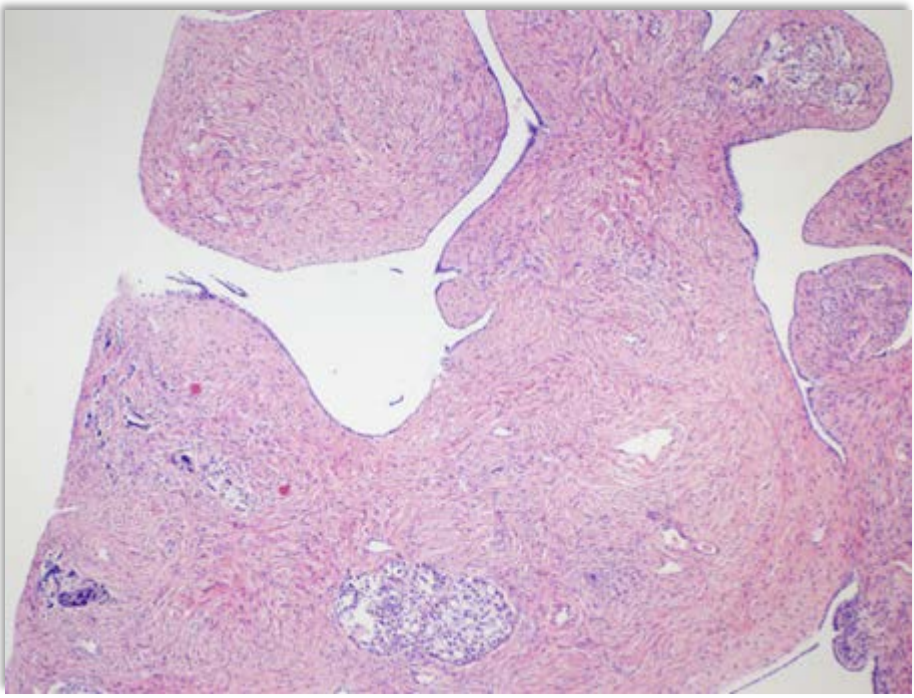
- İntraoperatif inceleme
- 16 yaş/F
- Gecikmeli adet
- Sağ over üzerinden biyopsi

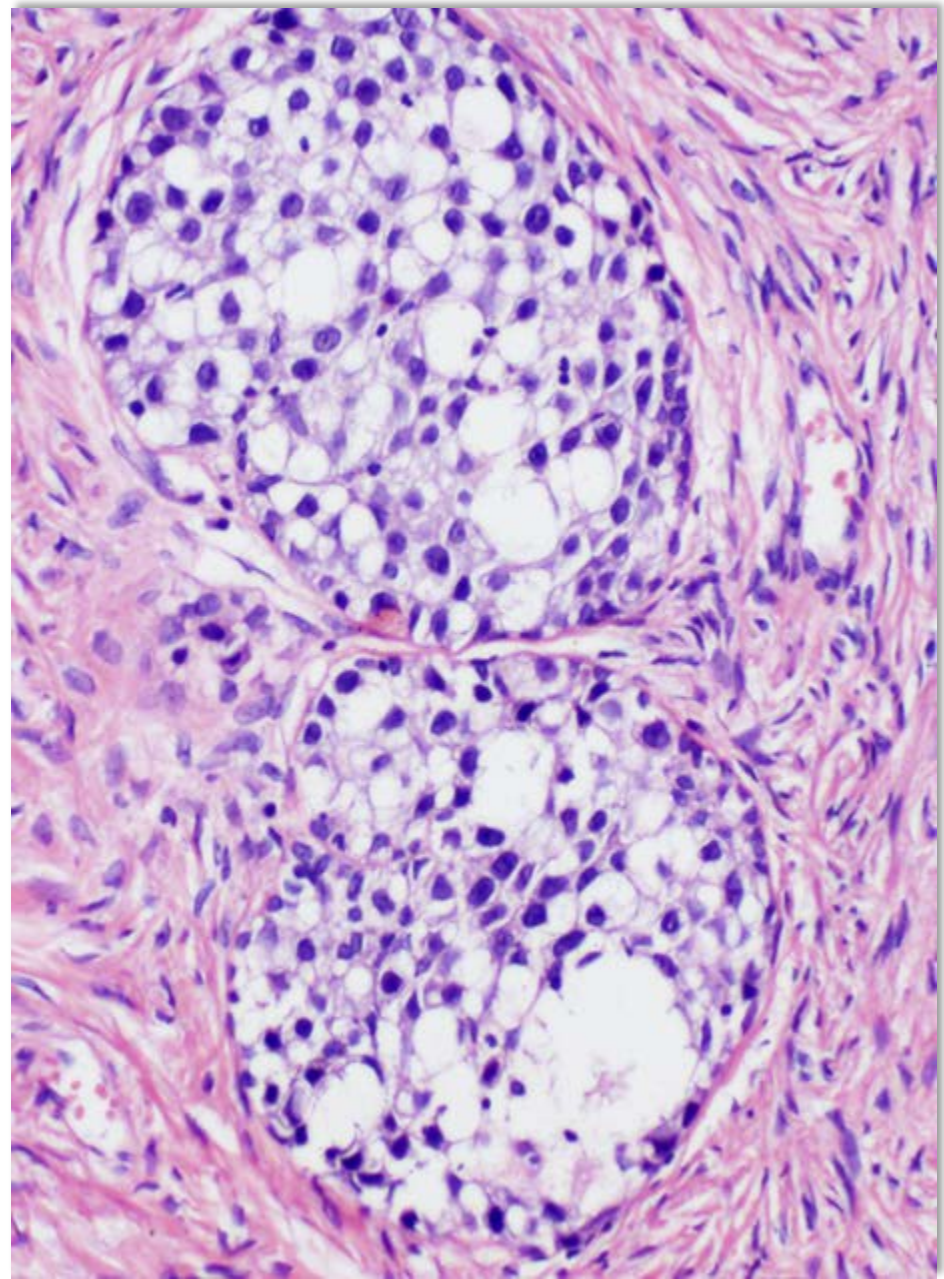
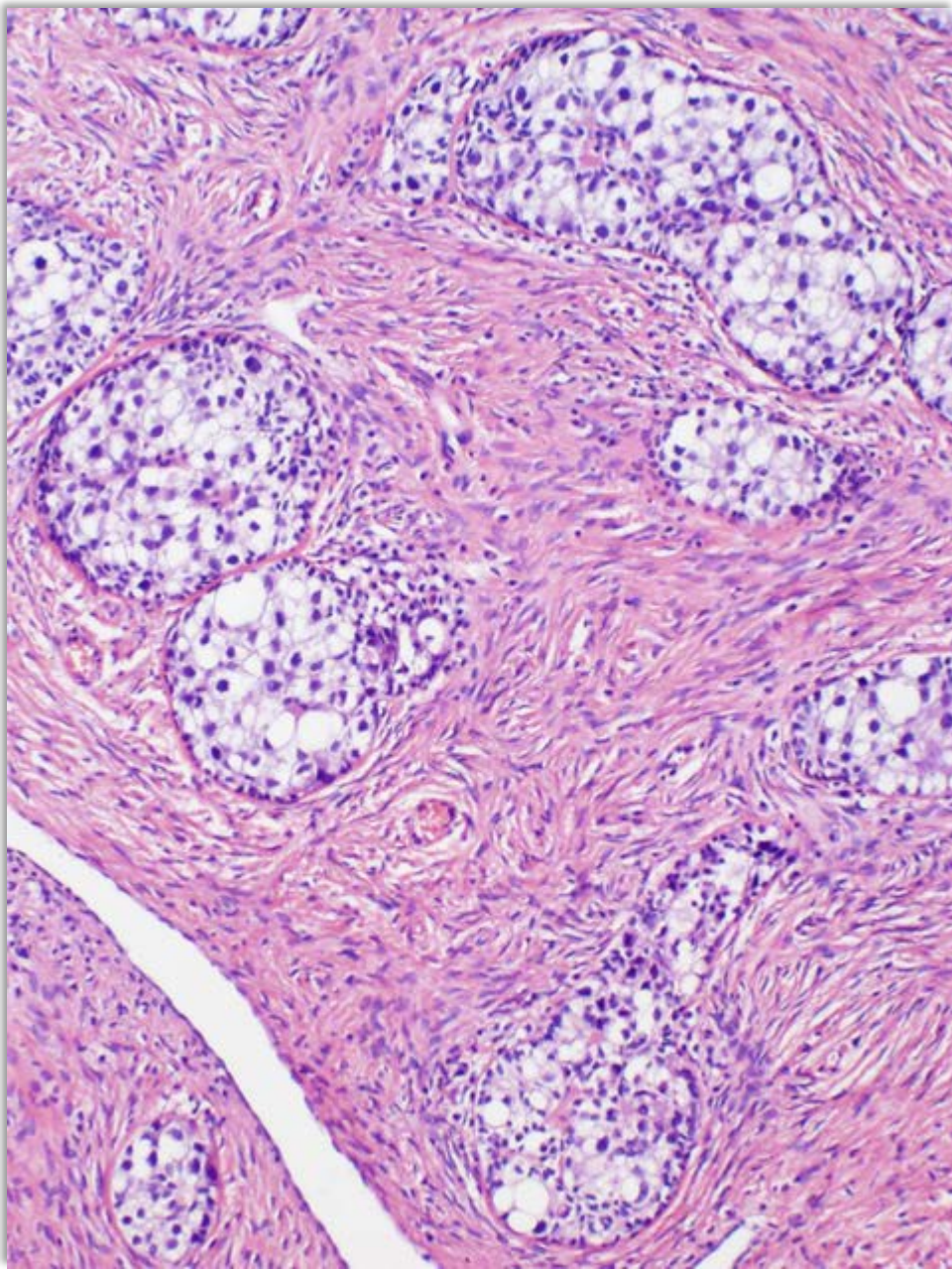


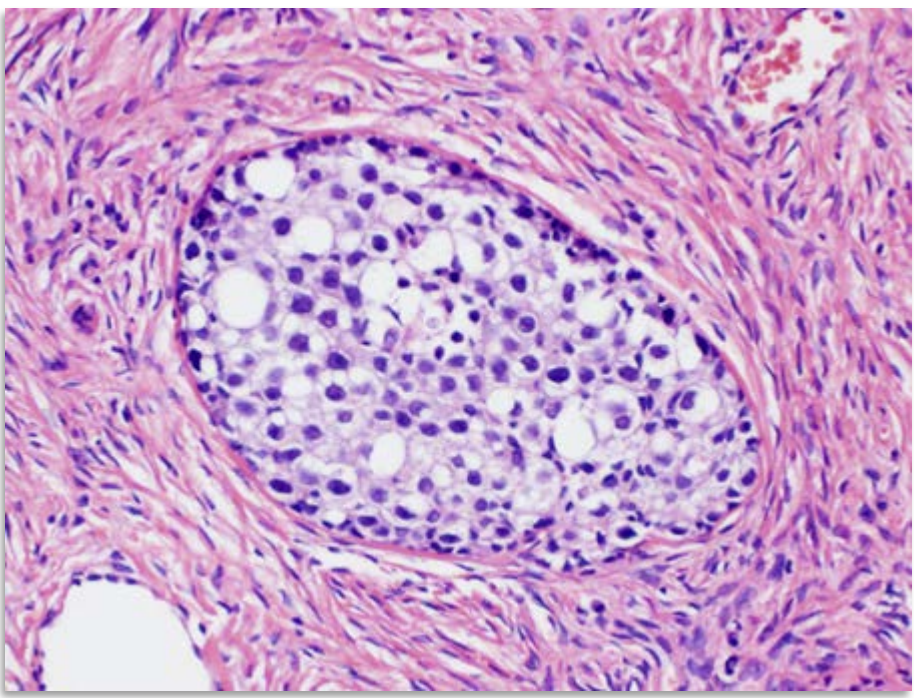
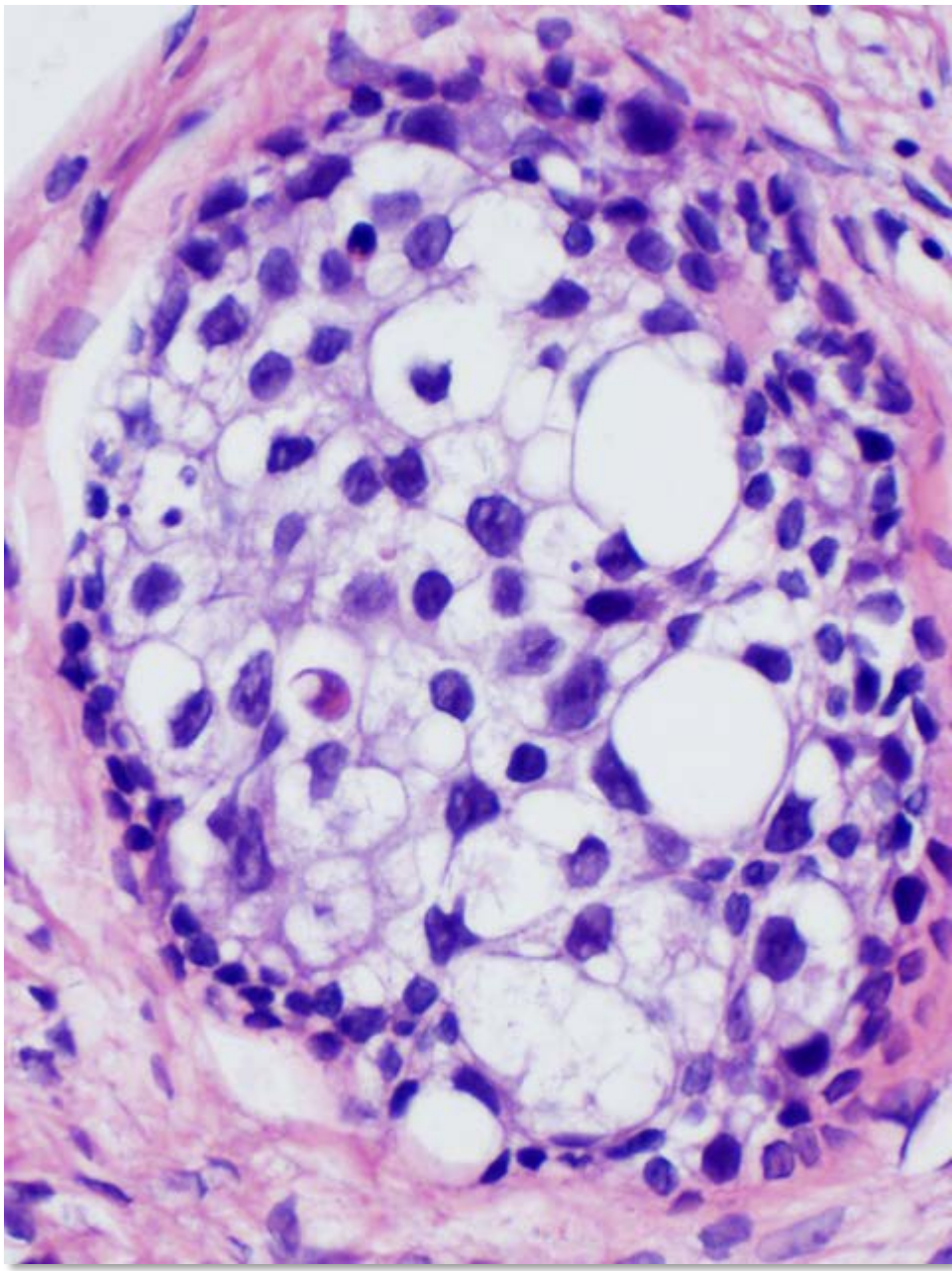
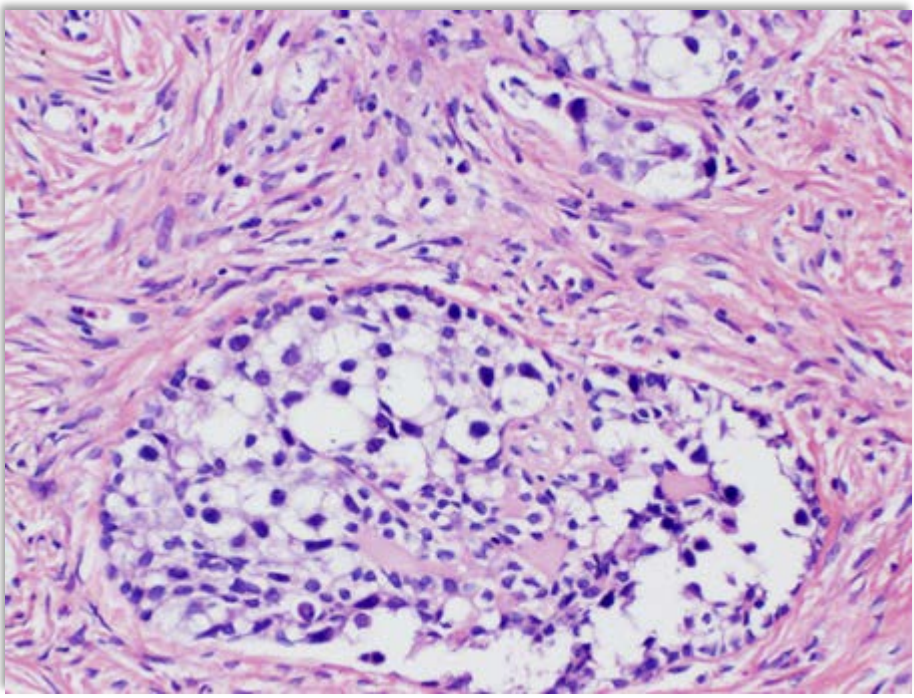


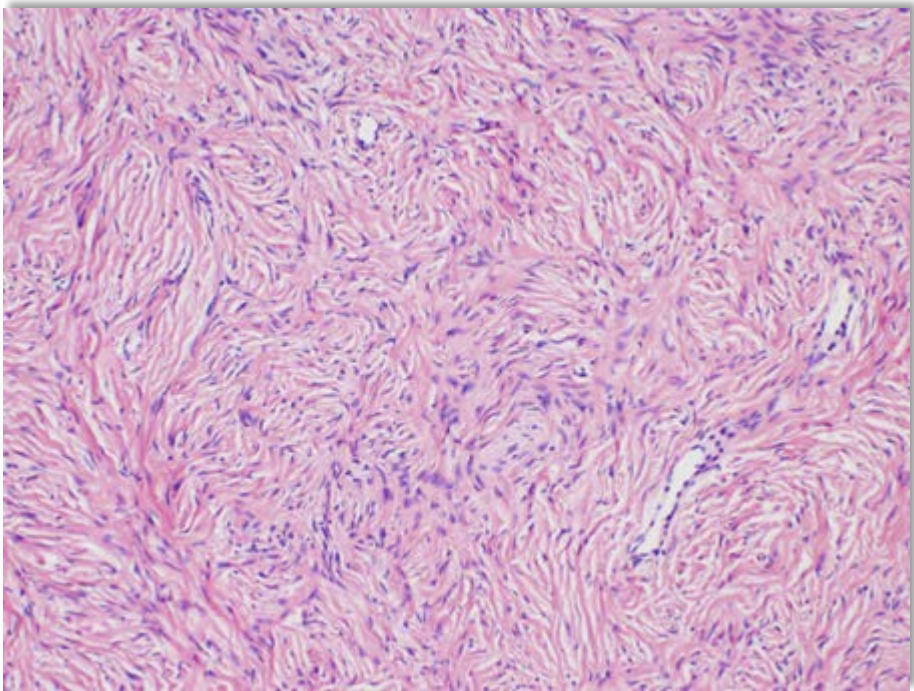
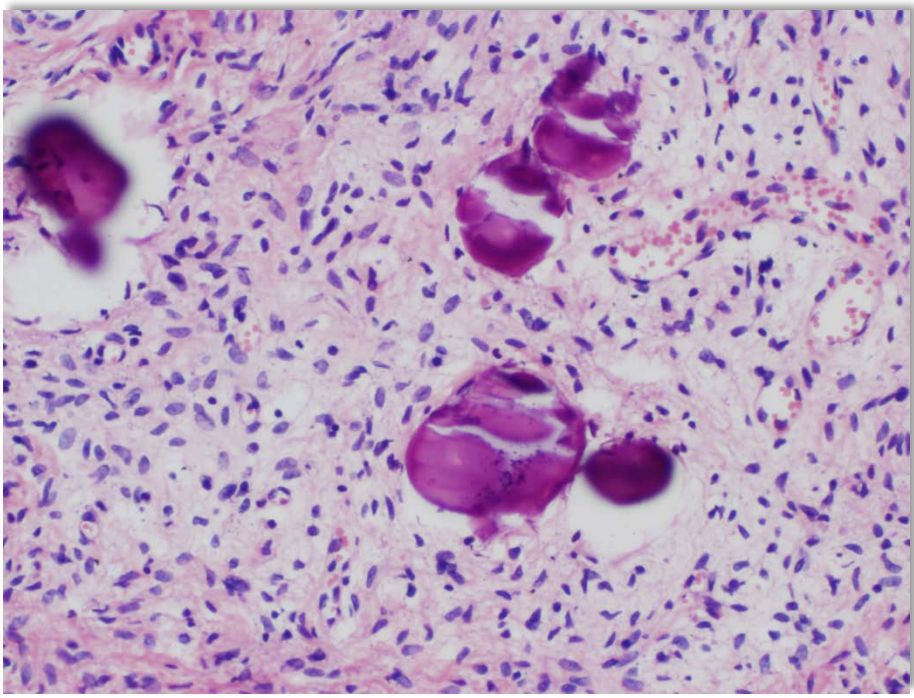
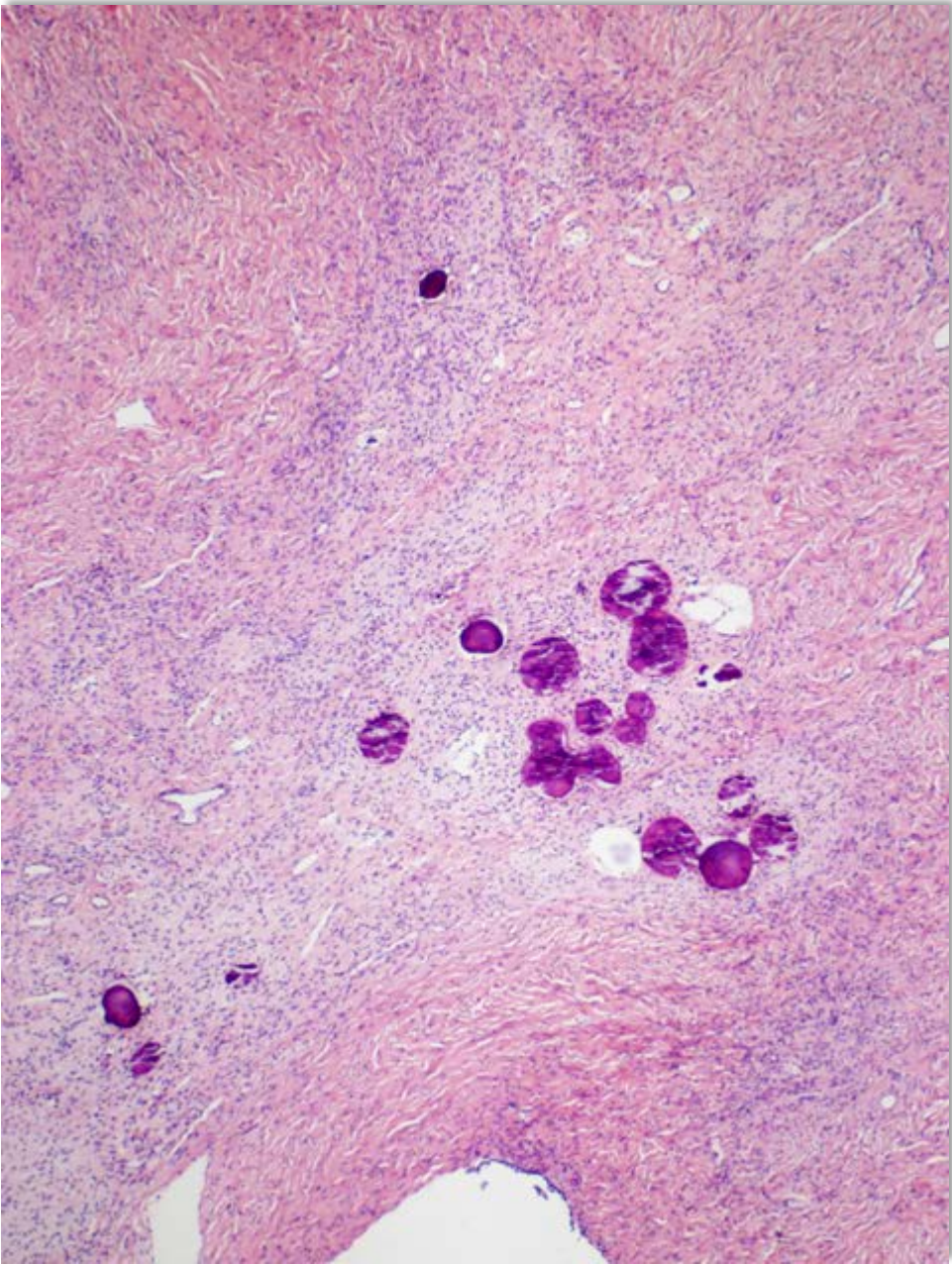


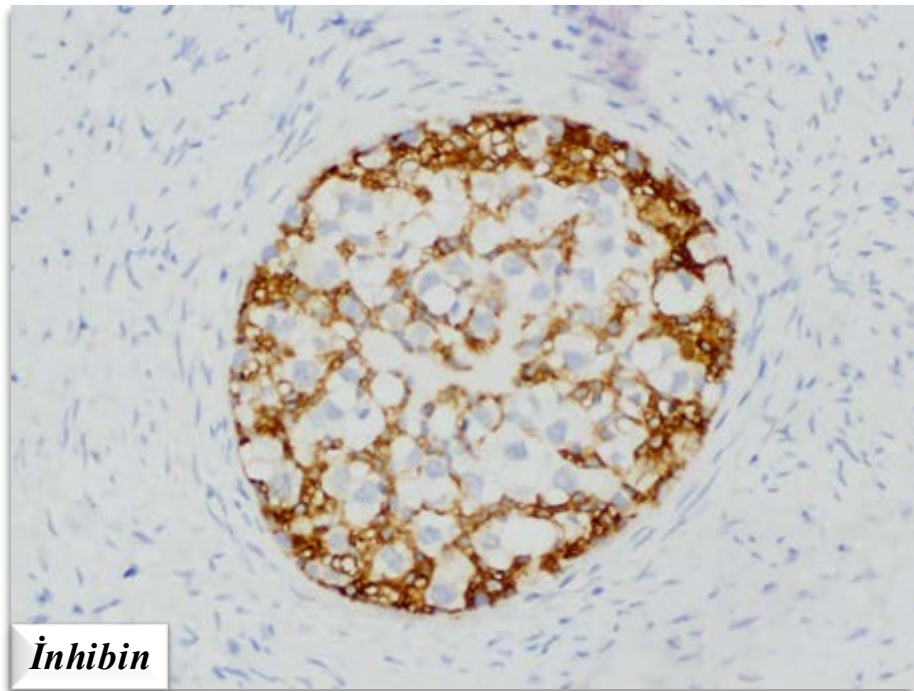
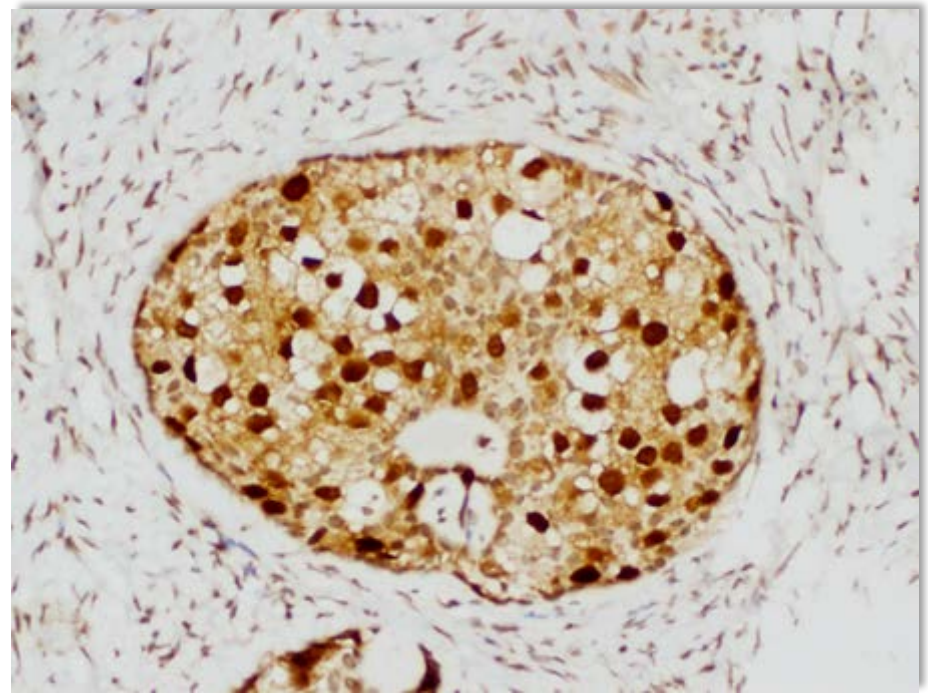
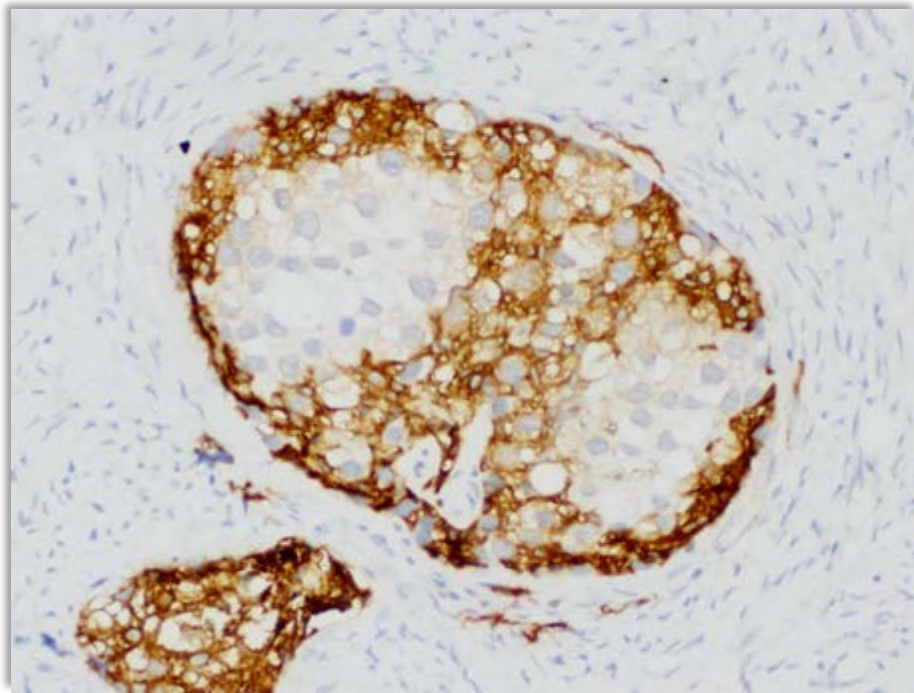
- Gecikmeli adet
- HRT (Cyclo-Progynova)
- Biyokimya normal
- Alt abdomen MR
 - Hipoplazik over
 - Sağ overde tümöral lezyon açısından şüpheli görünüm
- Laparoskopi önerisi



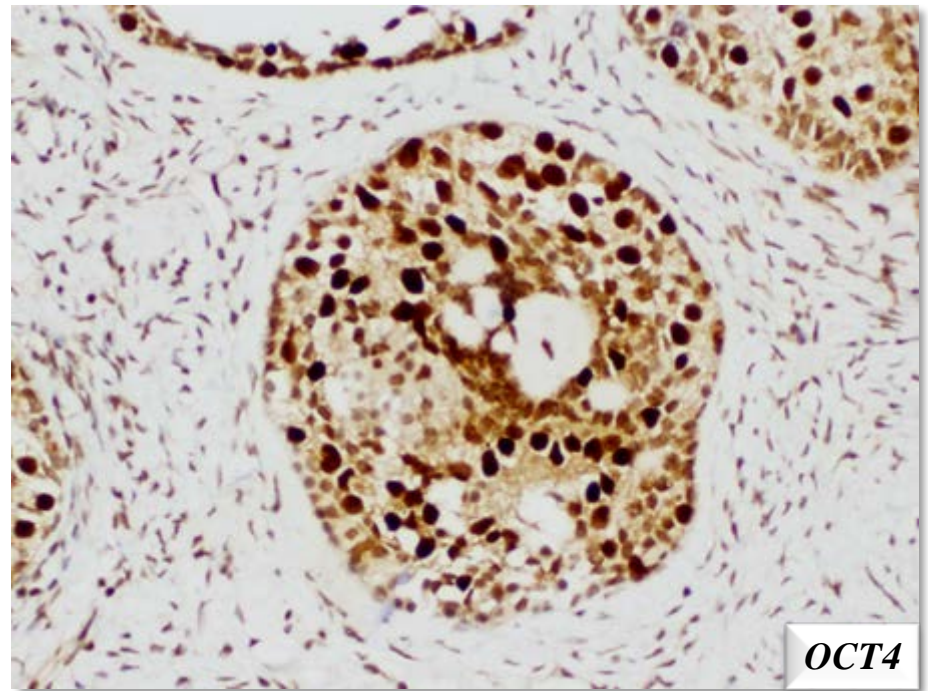








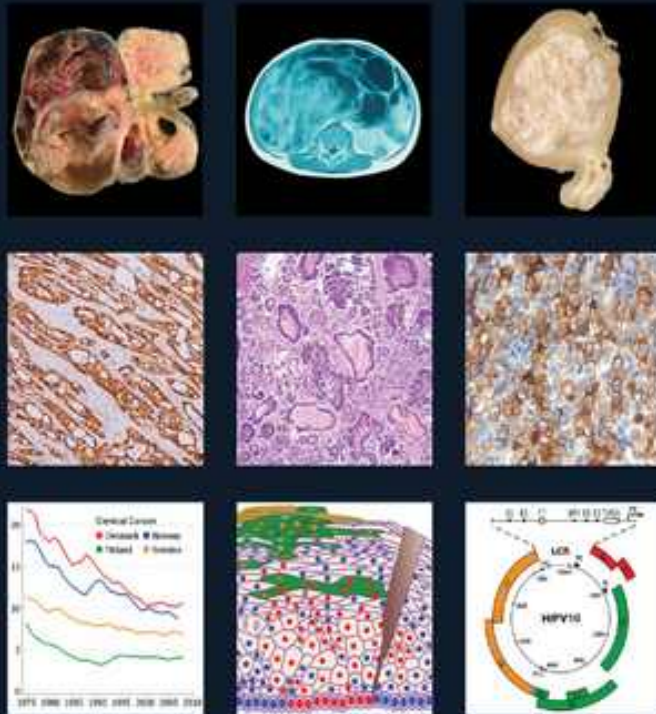
Inhibin



OCT4

WHO Classification of Tumours of Female Reproductive Organs

Edited by Robert J. Kurman, Maria-Luisa Carcangiu, C. Simon Herrington, Robert H. Young



WHO 2014

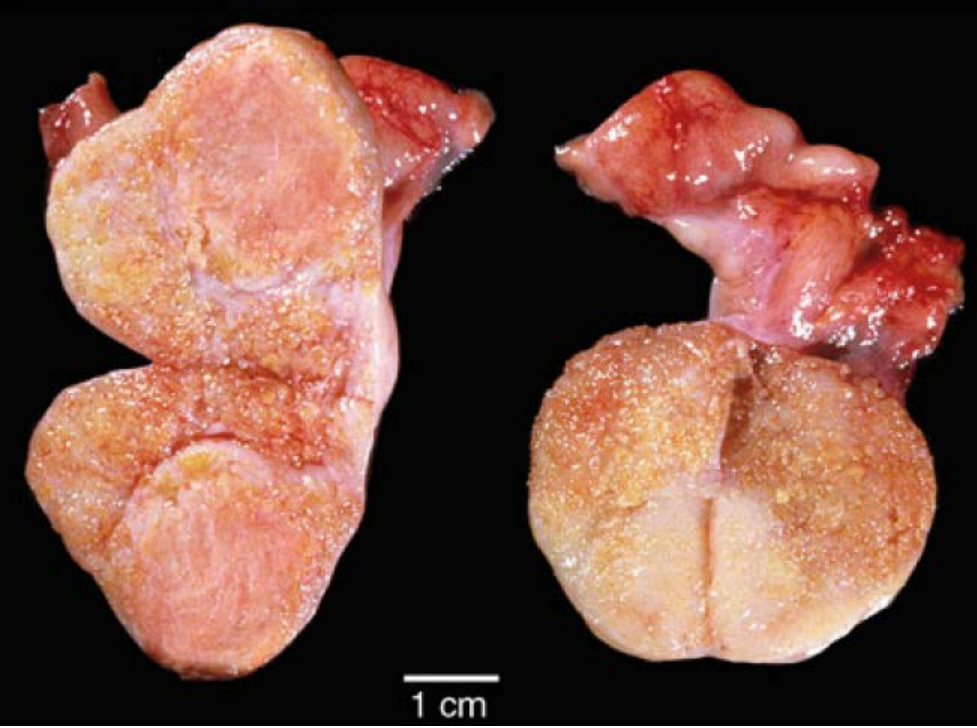
- **Germ cell-sex cord-stromal tumours**
 1. Gonadoblastoma, including gonadoblastoma with malignant germ cell tumour
 2. Mixed germ cell-sex cord-stromal tumour, unclassified

Gonadoblastoma

- İmmatür seks kord hücreleri (Sertoli/granuloza hücreleri) ve germ hücrelerinden oluşur - **Scully (1953)**
- Gonadoblastoma şeklinde metastaz ~~Ø~~
- %50 olguda **disgerminom** birlikteliği,
- %10 olguda ise diğer malign germ hücre neoplazisi tanımlanmıştır
- Malign germ hücreli tümörün “in situ” formu

Klinik Özellikler

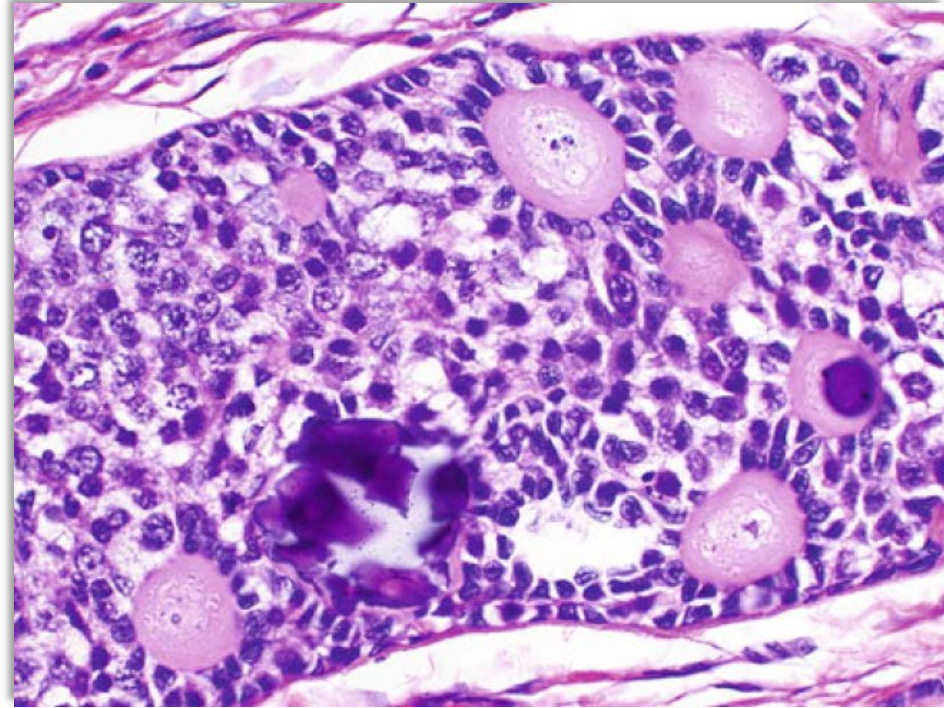
- İkinci dekad (Doğum-4. dekad)
- Female fenotipte (%80) gonadal disgenezis ve anormal karyotipli hastalar
- Nadiren 46 XX fenotip
- Primer veya sekonder amenore/ genital organlarda anomali
- Virilizasyon (%60)

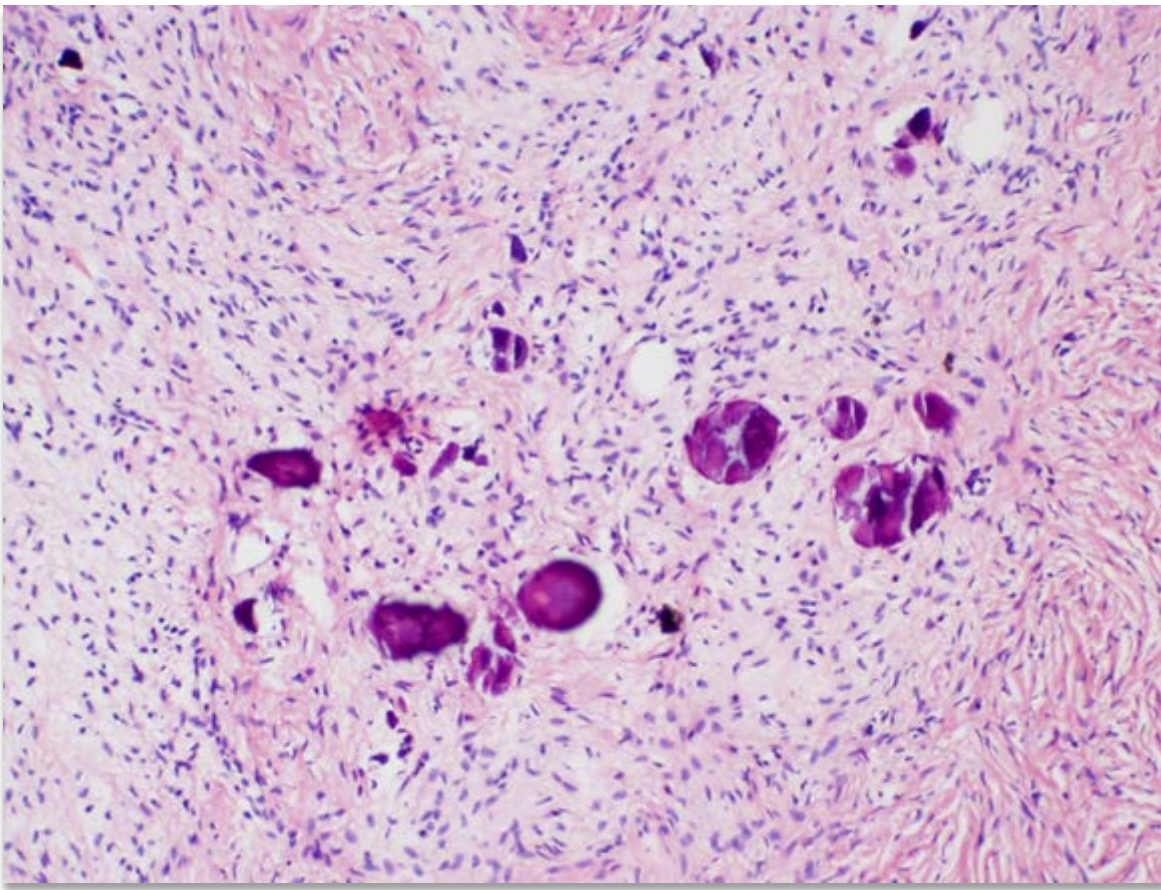


- 2-3 cm (8 cm >) **RO***
- %25 mikroskopik
- Beyaz-gri-sarı-kahverengi
- Kalsifikasyon nedeniyle sert-granüler
- %30-40 bilateral

Sertoli ve granuloza hücreleri

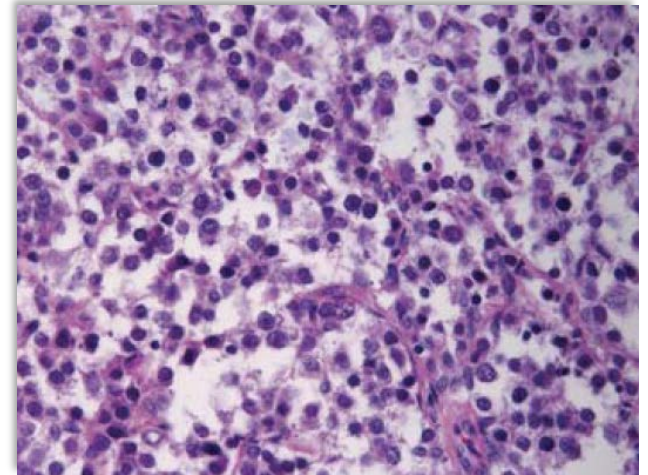
- Kümelerin periferinde koronal paternde izlenebilir
- Primer follikülde ovumun etrafını saran follikül epiteli gibi **germ hücreleri**nin etrafını sarar
- Call-Exner cisimciklerine benzeyen yapılar oluşturur





Leydig veya luteinize stromal hücrelere benzer hücre toplulukları (%66)

1. Hyalinizasyon
2. Kalsifikasyon
3. Disgerminom gelişimi



İmmunohistokimyasal Özellikler

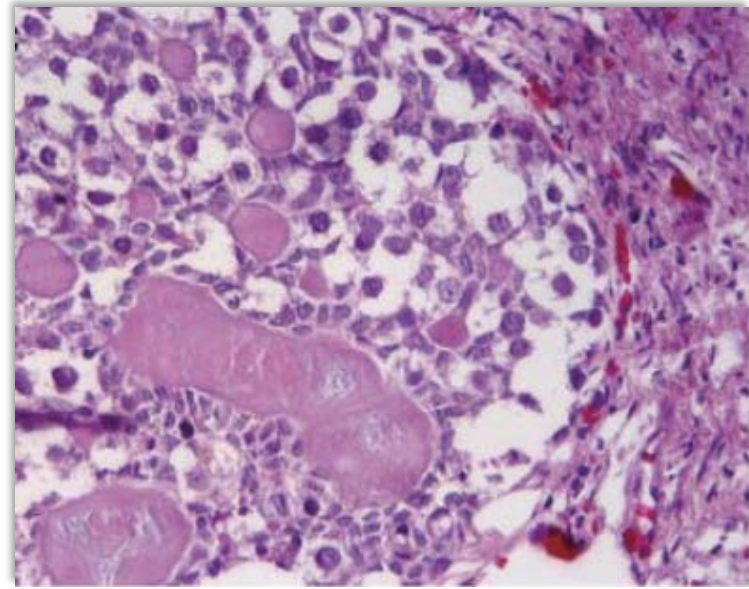
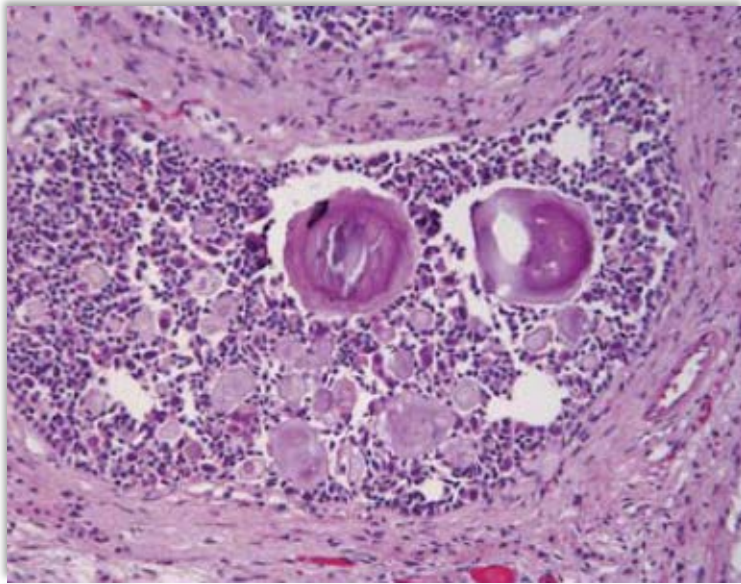
Germ hücreleri

- PLAP
- CD117
- OCT-4
- SALL4

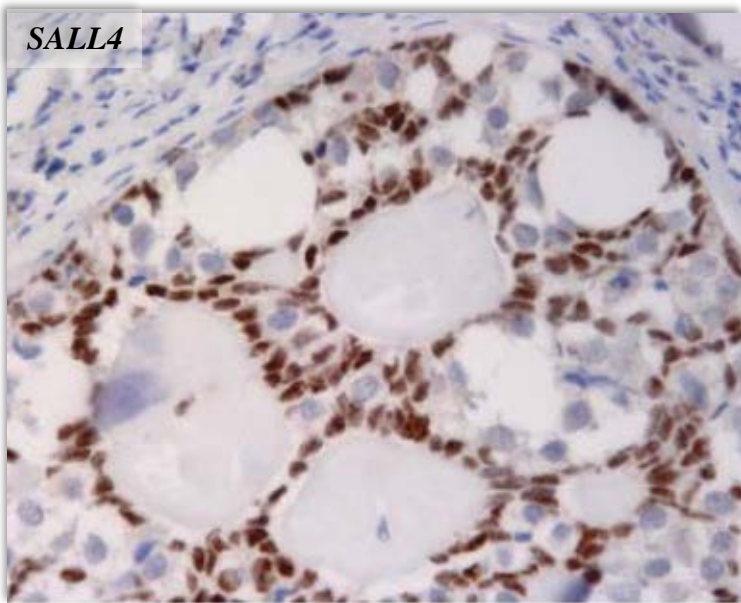
Seks kord hücreleri

- İnhibin
- Kalretinin
- WT1
- FOXL2

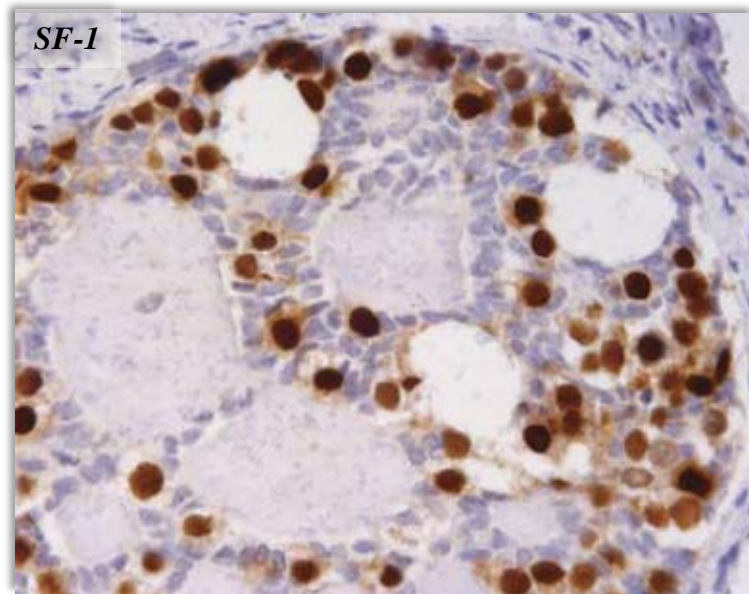
SALL4 AND SF-1 EXPRESSION PATTERNS IN GONADOBLASTOMA



SALL4



SF-1



Genetik Özellikler

- Pür veya mikst gonadal disgenezis/ Male psödohermafrodit
- %96 olguda Y kromozom (TSPY1 gen)
- En sık saptanan karyotip: 46XY (%50)
- 45X/46XY mozaizizm (%25)
- 46XX (8 olgu), 4 olgu fertil
- Gonadal disgenezis ve gonadoblastoma
Y kromozomal DNA'sı olmayan hastalarda görülebilir

Patogenez ve Prognoz

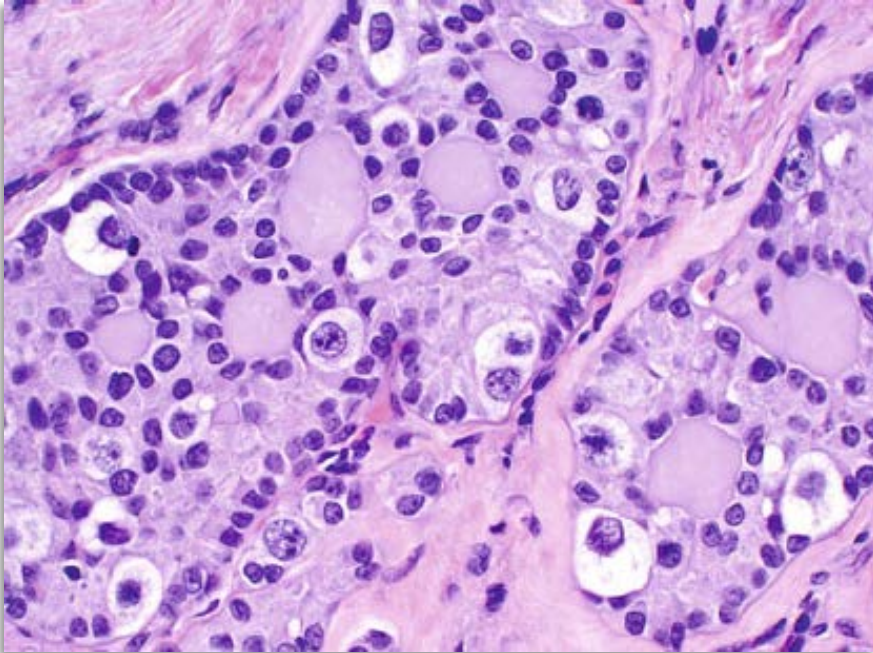
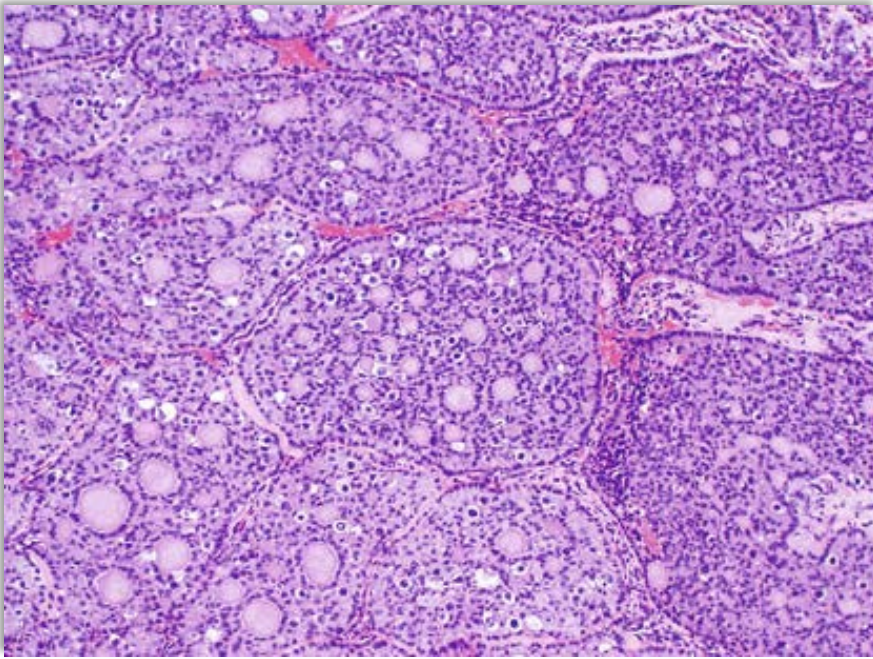
- Disgenetik gonadlardaki andiferansiye gonadal dokunun OCT3/4-pozitif germ hücrelerinden geliştiği öne sürülmektedir
- Tek başına benign
- Malign transformasyon olursa oluşan tümörün tipine, boyutuna ve evresine göre prognoz değişir

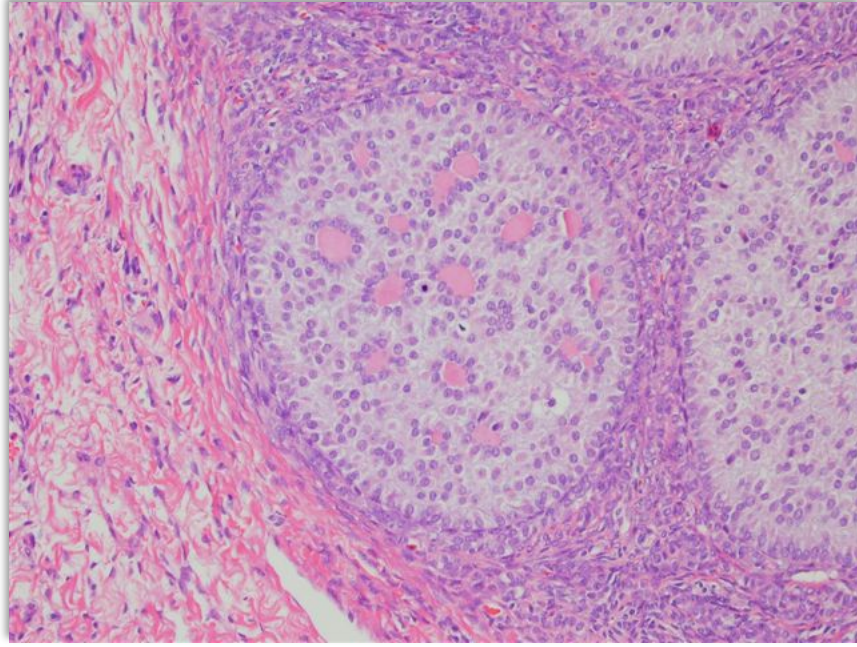
Ayırıcı Tanı

- Mikst germ hücreli-seks kord-stromal tümör
- Annular tubüller içeren seks kord tümör
- Normal fetüs ve yenidoğanda %15 oranında mikroskopik gonadoblastoma odağı görülebilir

Mikst germ hücreli-seks kord-stromal tümör

- Unilateral /Boyut 7,5-18 cm
- Diğer over normal
- 46XX karyotip
- Gonadal disgenezis ve somatoseksüel anomali yok
- Virilizasyon **yok**
- Germ hücre ve seks kord komponentte **proliferatif aktivite ↑**
- Kalsifikasyon, hıyalinizasyon **yok**
- Leydig ve lutein benzeri hücreler **yok**
- Çoğu **benign** (%10 oranında disgerminom ve diğer MGHT gelişimi)





Annular tubüller içeren seks kord tümör

- Sporadik veya Peutz-Jeghers sendromuna eşlik eder
- 3. ve 4. dekadlar
- Çoğunlukla bilateral (PJS)
- Sertoli ve granulosa benzeri hücrelerin oluşturduğu tubüller
- Call-Exner benzeri yapılar
- Kalsifikasyon
- Germ hücreleri **yok**
- Sporadik: düşük dereceli malignite



Inhibin

46 XX

Alt abdominal MR
(15/06/2015)

- Sağ over boyutlarında azalma
- Hipoplazik yapıda sol over dokusu

Önerilen

Bilateral oofektomi (diğer over normal görünümde olsa bile)

Disgenetik gonadlar malign germ hücreli tümörlerin kökeni olabilir

