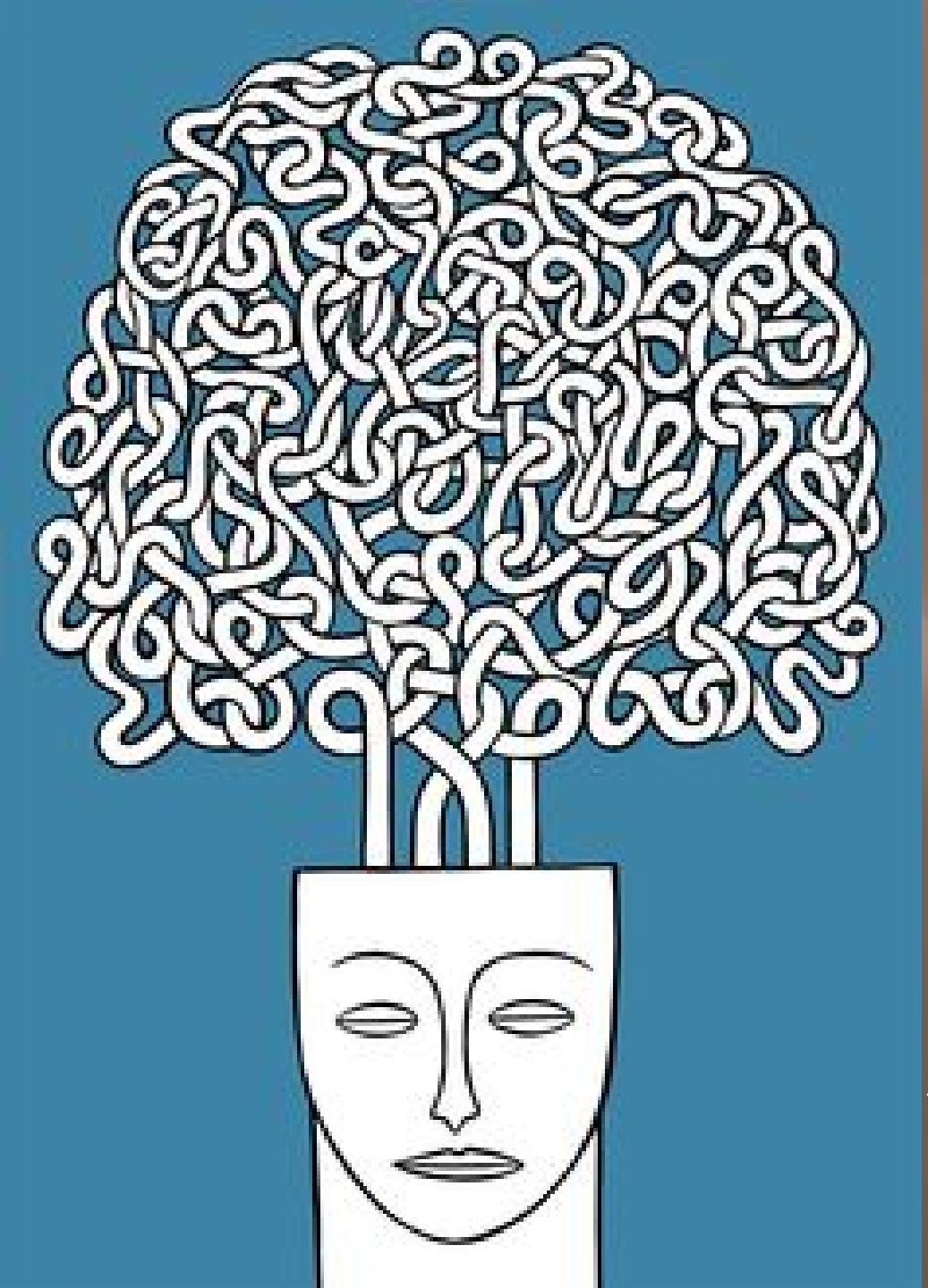


NÖROPATOLOJİ SLAYT SEMİNERİ

SELLAR BÖLGE KİTLE LEZYONLARI

OLGU-4

Doç.Dr.Güzide Ayşe OCAK

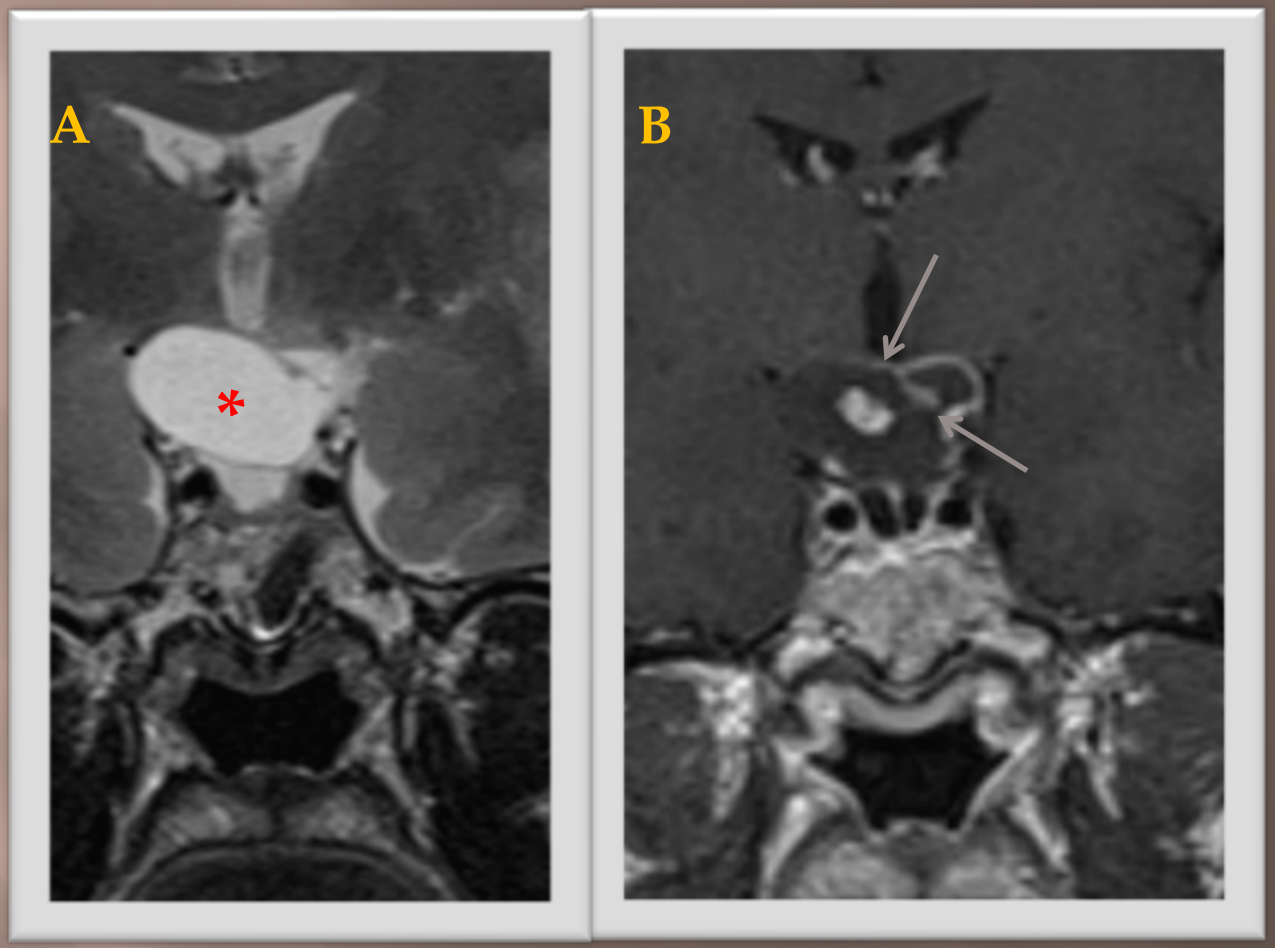
1.Olgu

- 26 yaşında erkek hasta
- Şikayet: Görme bozukluğu ve baş ağrısı
- MRI: Suprasellar yerleşimli, 4 cm çapında solid/kistik kitle. Solid alanlarda heterojenöz kontrast tutulumu var.
- Operasyon: Transsfenoidal yolla kitle eksizyonu

2.Olgu

- 45 yaşında kadın hasta
- Şikayet: Görme bozukluğu ve baş ağrısı

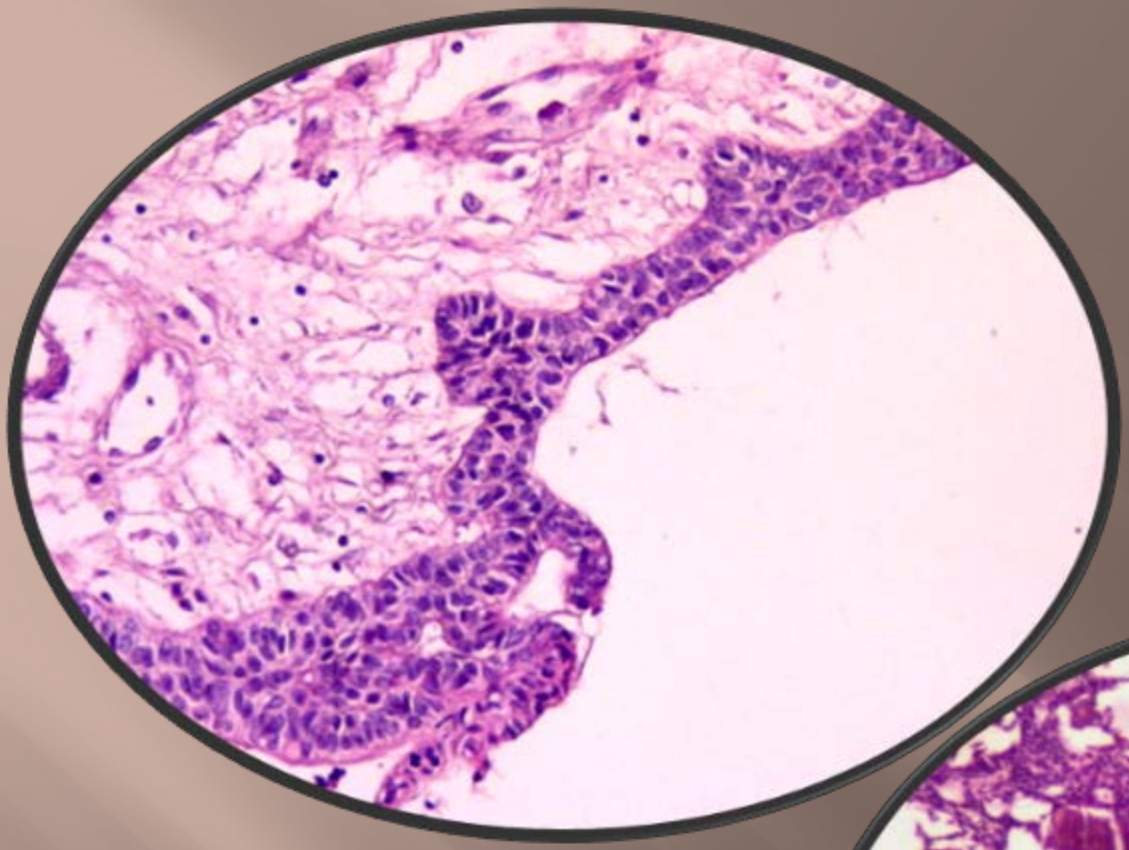
MRI

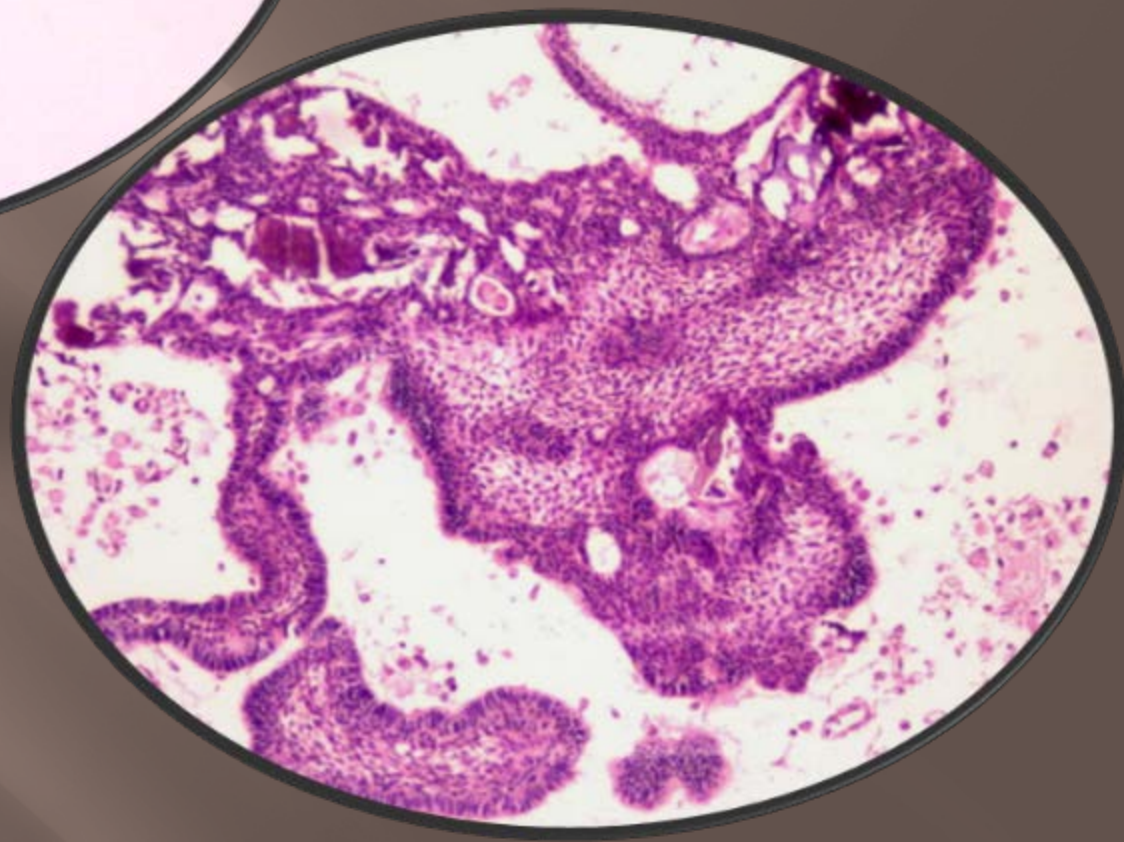


T2-ağırlıklı koronal kesitte (A) suprasellar kistik kitle (*) izleniyor
Kontrast sonrası T1-ağırlıklı koronal kesitte çevresel kontrastlanma (oklar) dikkat çekiyor.

- Özgeçmiş: Aynı bölgede kitle eksizyonu

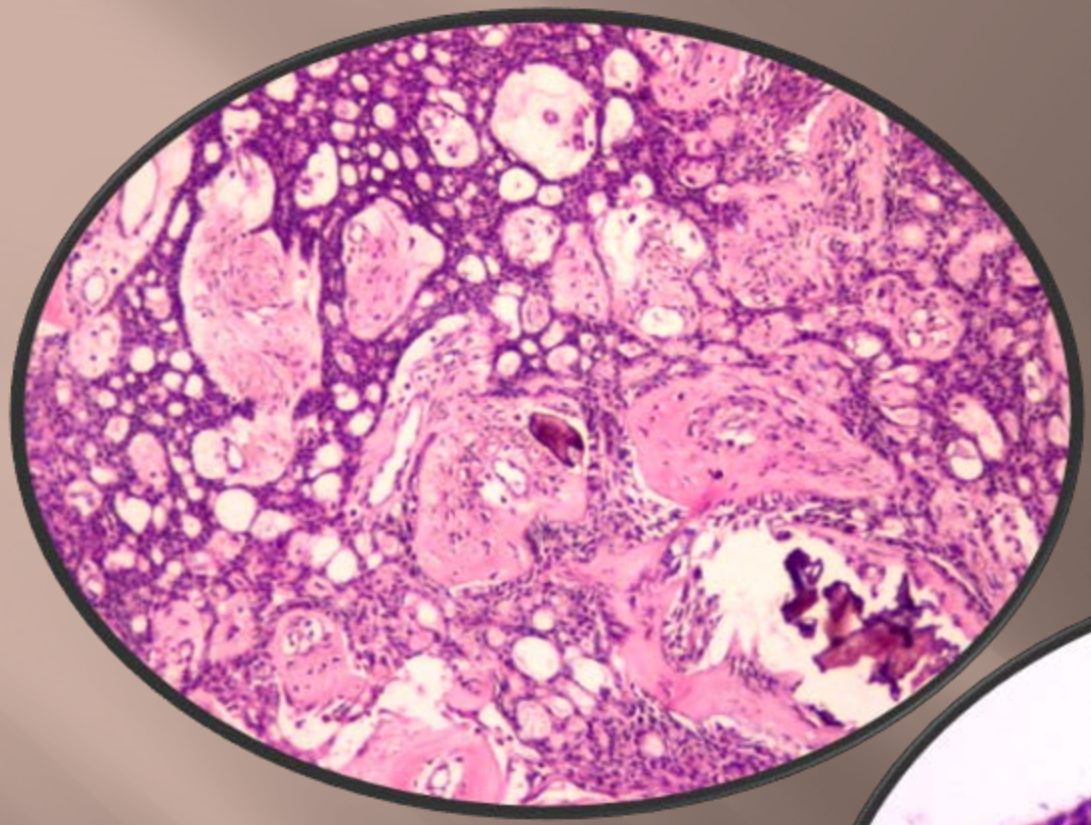
- Operasyon: Nöronavigasyonla kitle eksizyonu

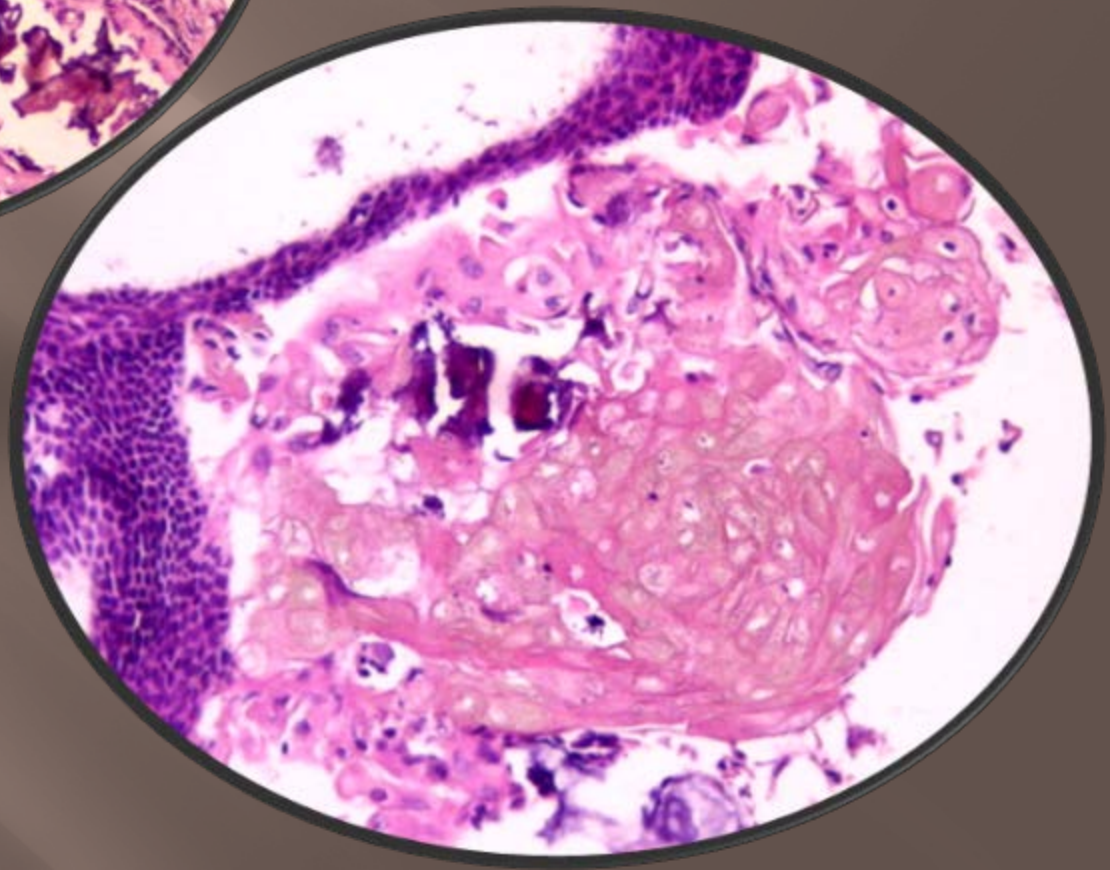


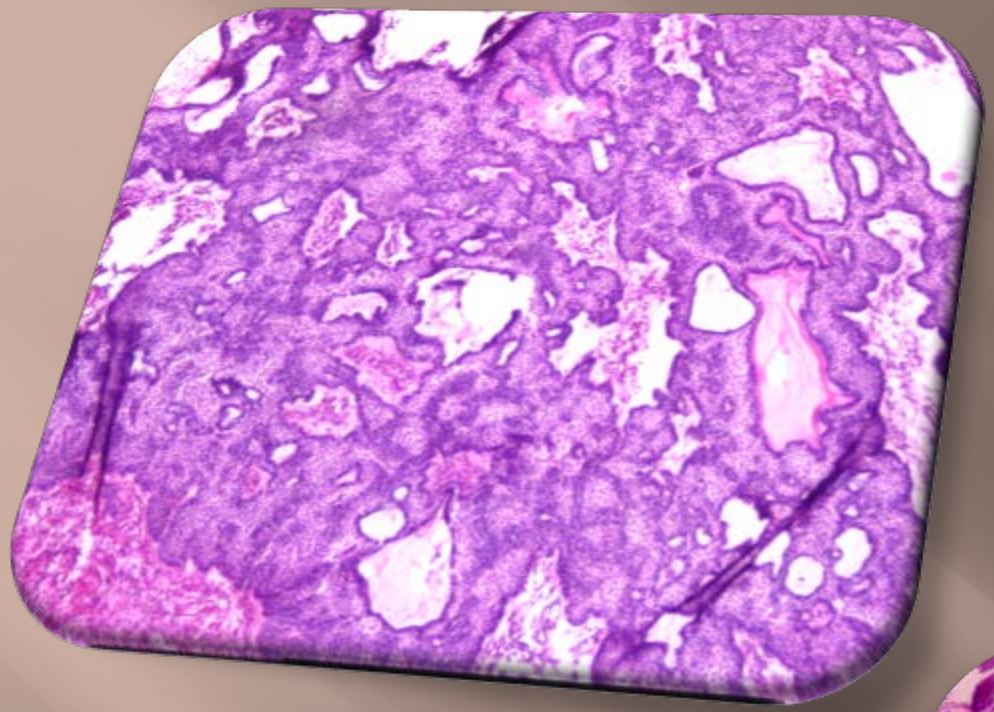


1.Olgu

1.Olgu







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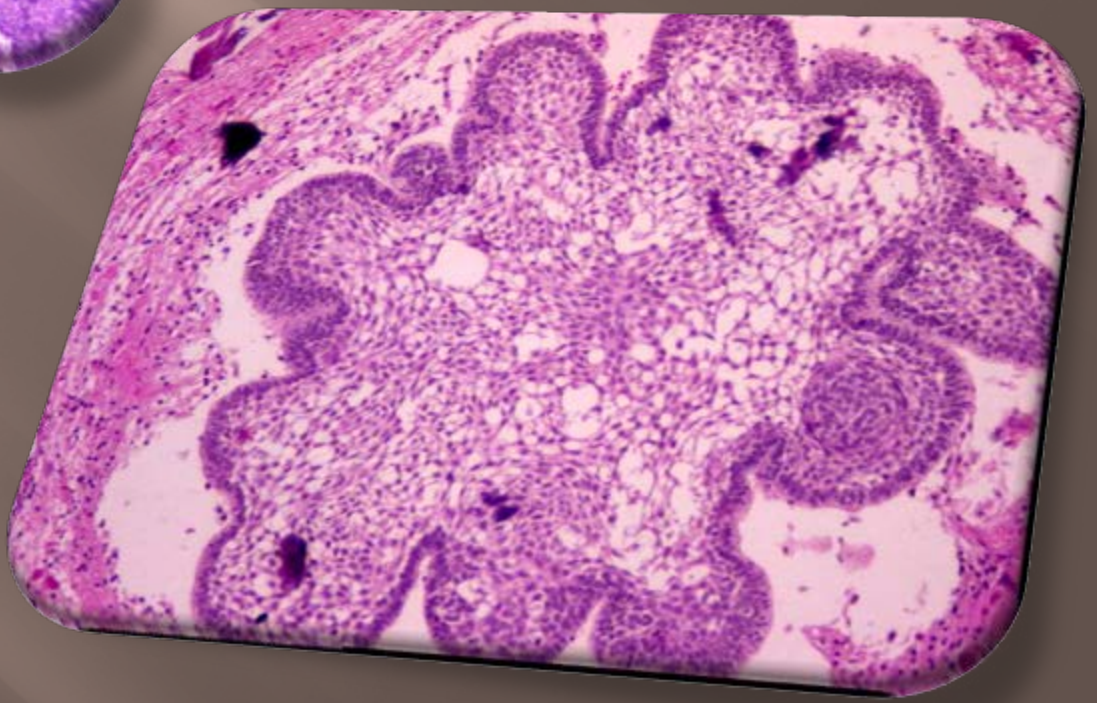

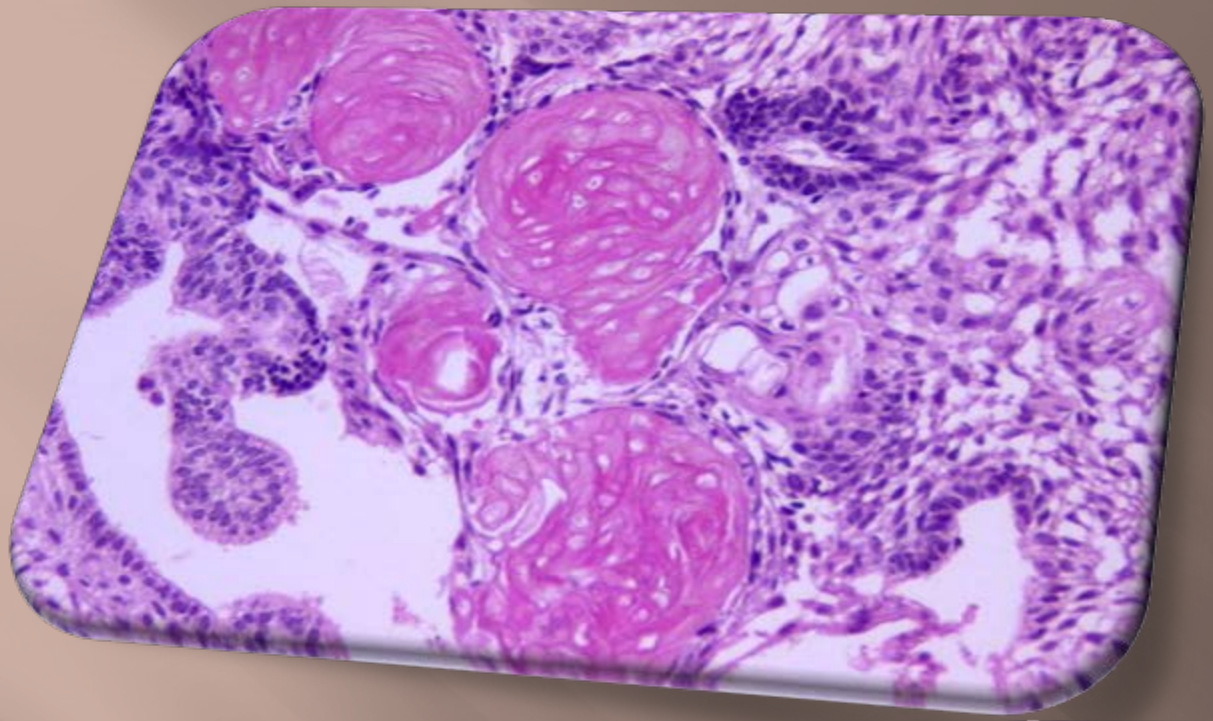

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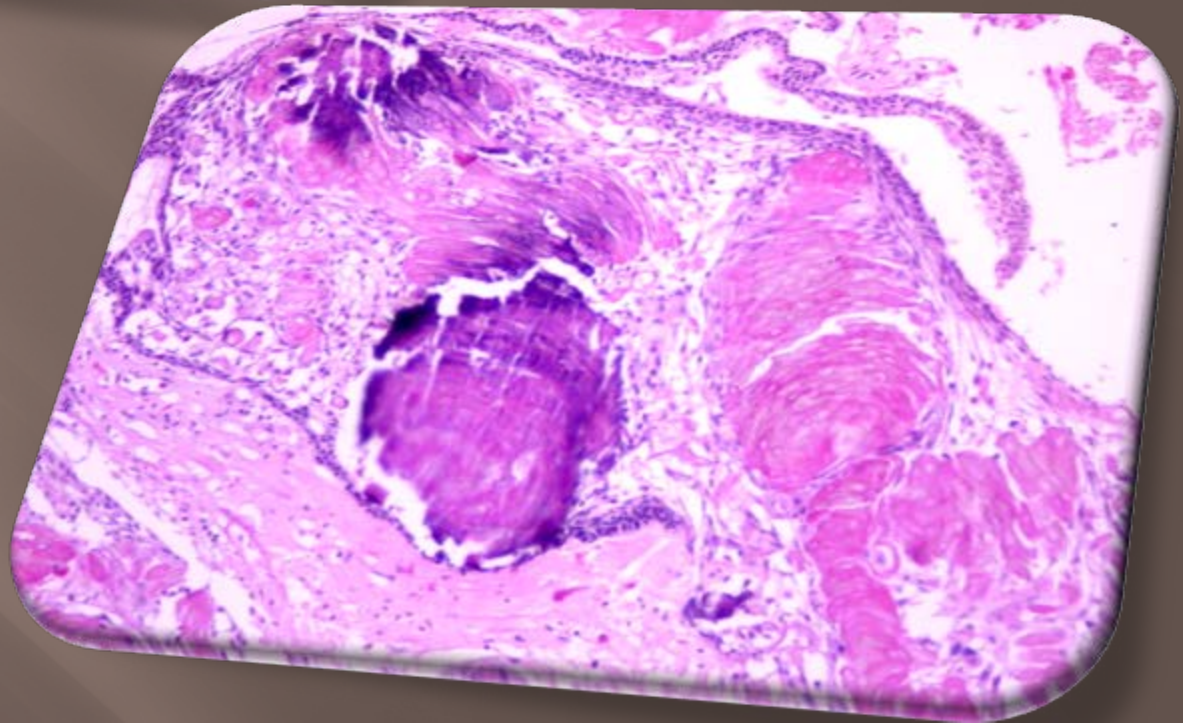
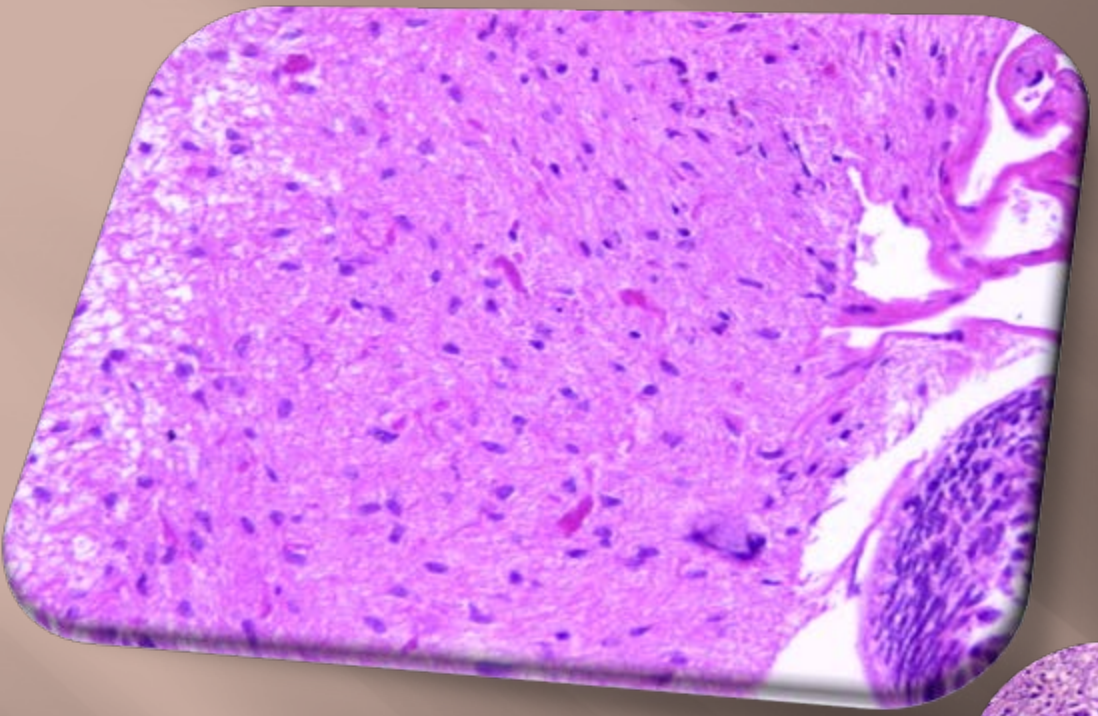
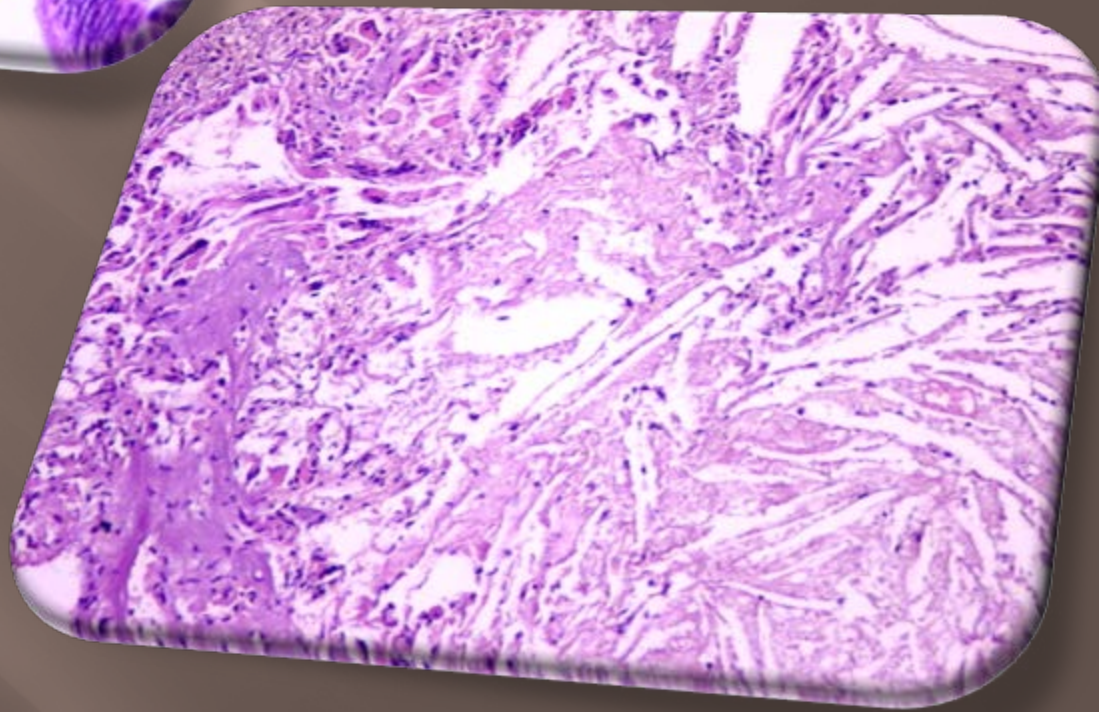
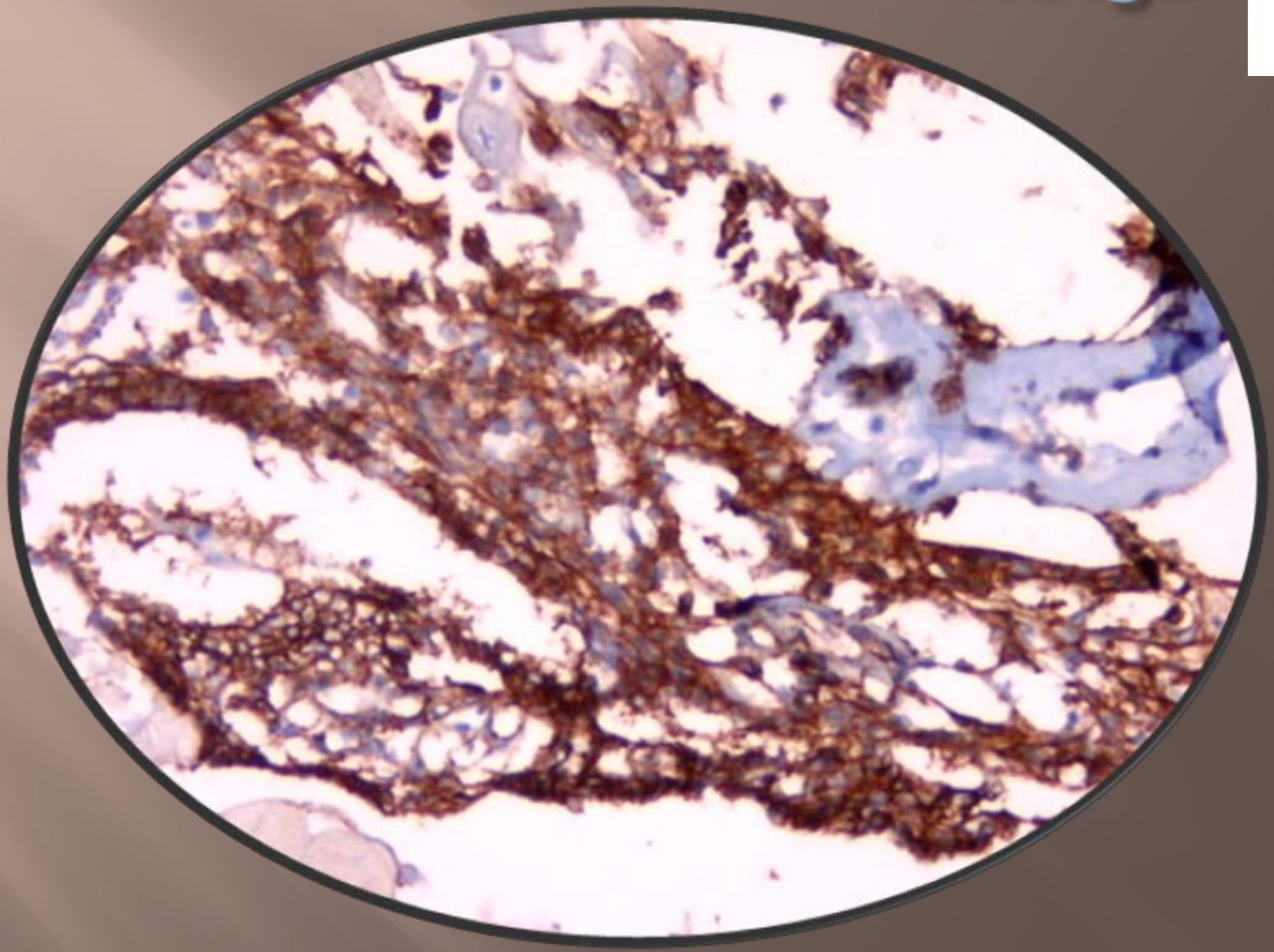






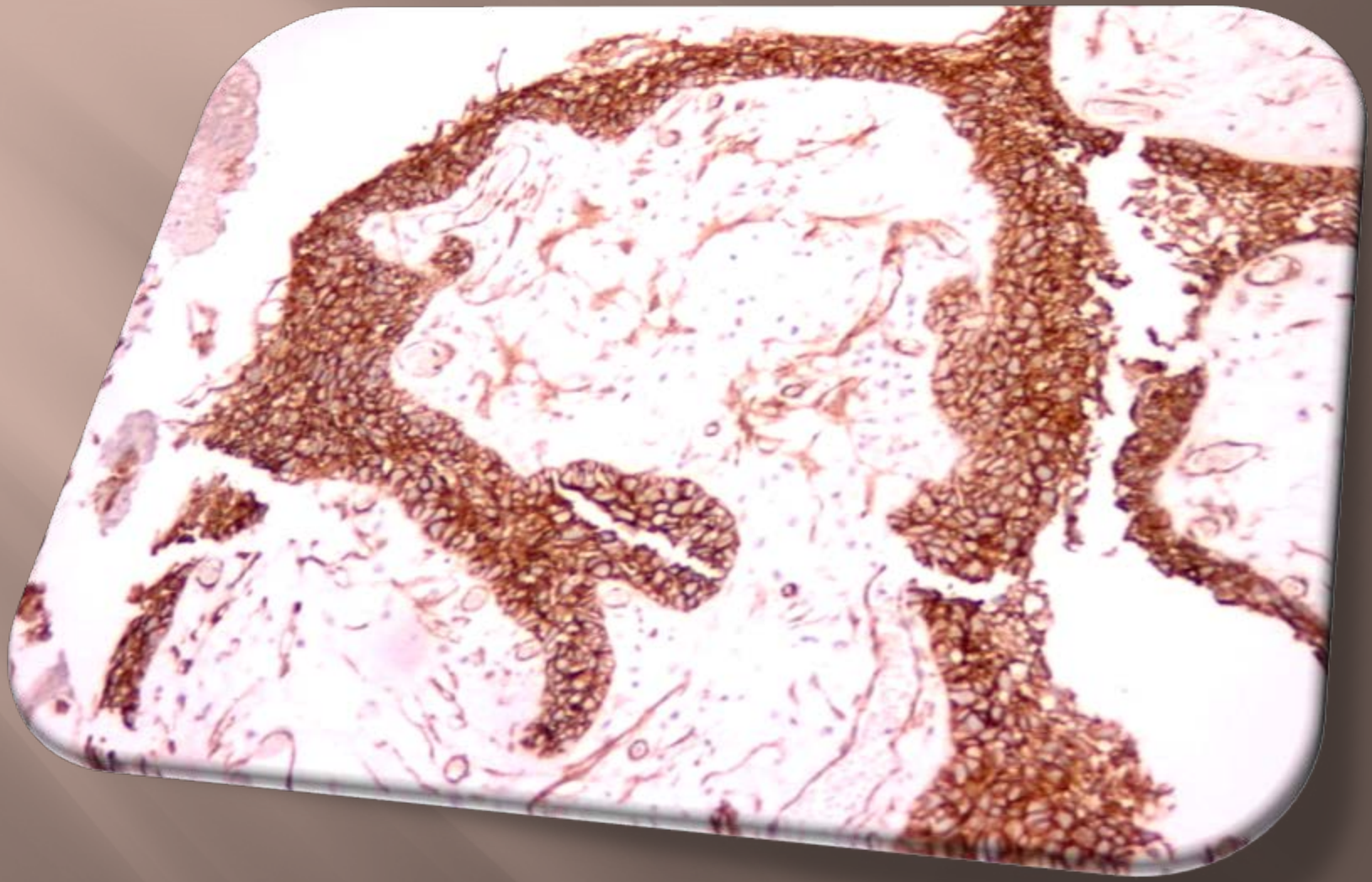
2.Olgu

1.Olgu



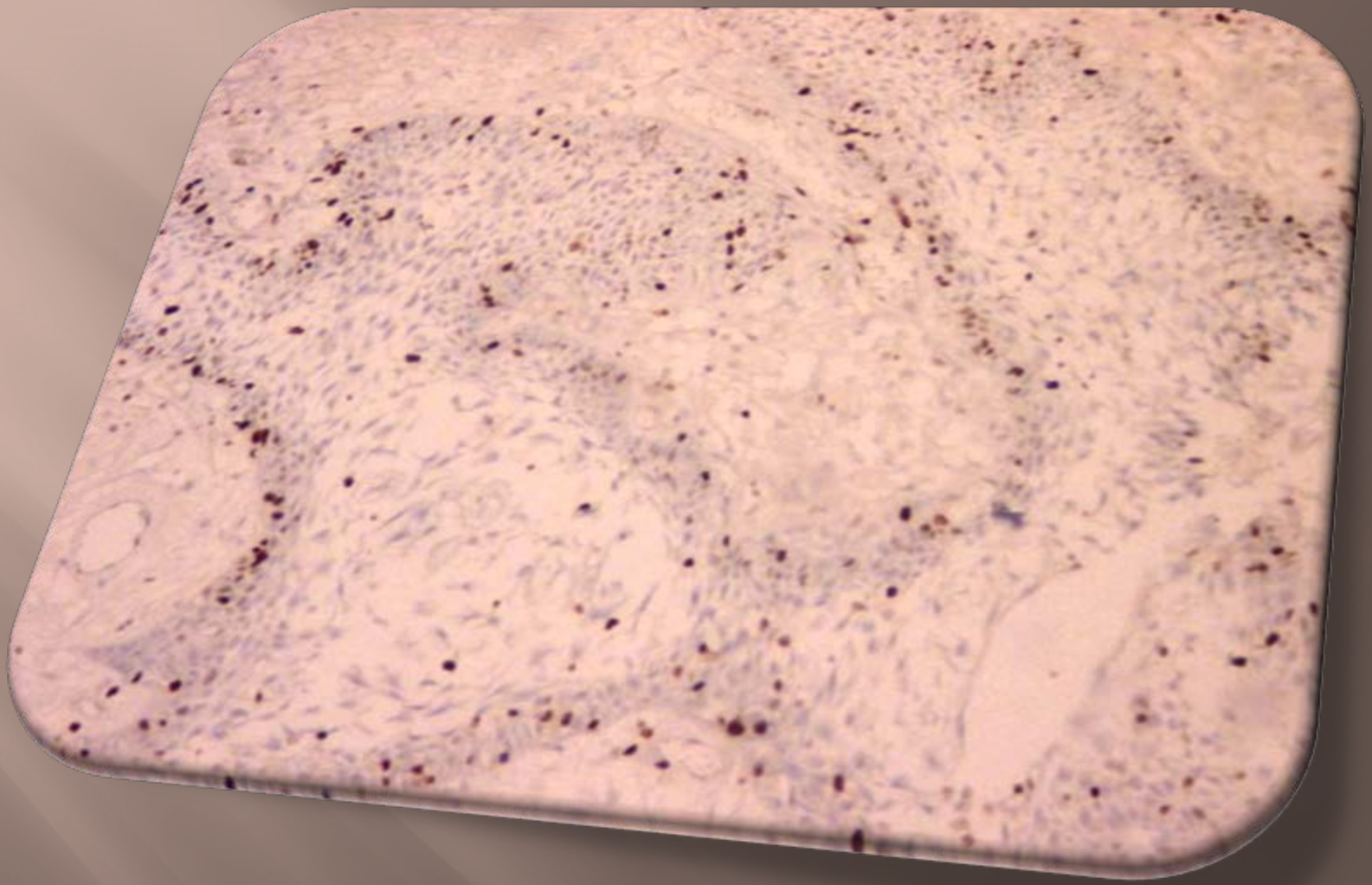
Beta katenin

2.Olgu



Beta katenin

2.Olgu



Ki 67

Histop TANI lgular

1. Kompakt skuamöz epitelden oluşan tabaka ve lobüller
2. Gevşek stellat retikulumu çevreleyen palizatik epitel
3. Anükleer "hayalet hücre
4. Intra
5. pilöid gliozis
6. lkasyon,nekroz,kolesterol yarıkları
7. İHK sal Beta katenin pozitifliği (nükleer ve sitoplazmik)

ADAMANTİNAMATÖZ KRANİOFARİNGİOM
DSÖ GRADE I

KRANİOFARİNGİOM

- Sellar bölgenin benign ancak lokal invaziv tümörleridir.
- DSÖ Grade 1
- Tüm intrakraniyal tümörlerin %4.6'sını,
- Çocukluk çağı intrakraniyal tümörlerinin %5-10'unu oluşturur.
- Adamantinamatöz ve Papiller olmak üzere 2 altipi mevcuttur.

KRANİOFARİNGİOM

Patogenez

- Adamantinamatöz KF'un (AKF) orjininin Rathke poşunun ektopik embriyonik kalıntıları olduğu düşünülmektedir ve odontojenik tümörlerin özelliklerini paylaşmaktadır.
- Papiller KF'un (PKF) patogenezi????

? ön hipofiz epitel hücrelerin metaplastik transformasyonu

? Rathke kleft kisti

- AKF'ın %70 inde Beta katenini kodlayan CTNNB1 geninde mutasyonlar mevcuttur ve bu mutasyonlar Beta katenini stabilize ederek parçalanmasını engeller, nükleusta ve sitoplazmada birikimine neden olur
- Bu mutasyon PKF'da görülmez.

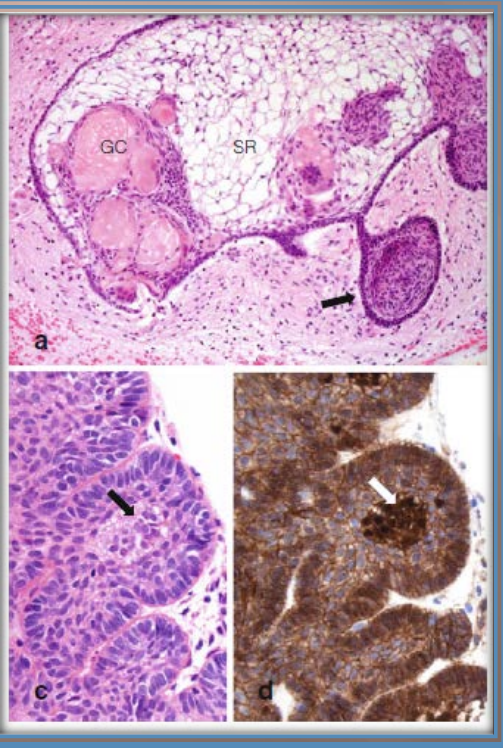
Pituitary (2013) 16:9–17
DOI 10.1007/s11102-012-0418-4

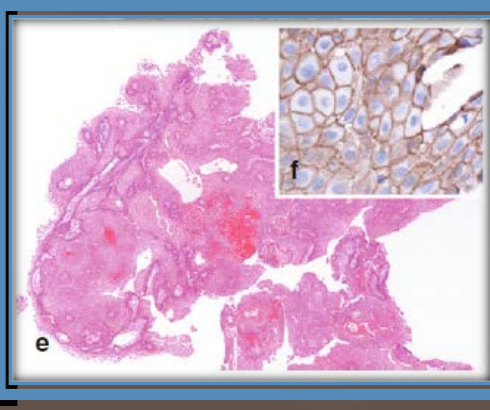
Pathology and pathogenesis of craniopharyngiomas

Sarah J. Larkin · Olaf Ansorge

Table 1 Comparison of pathological and pathogenic features of adamantinomatous and papillary craniopharyngiomas

Feature	Adamantinomatous craniopharyngioma	Papillary craniopharyngioma
Clinical features and epidemiology		
Incidence	0.13 cases per 10^5 person years [41]	
	90 %	10 %
Age	Bimodal, peak incidences 5–10 years and 50–60 years	Almost exclusively adult
Site	Suprasellar; infrequent intrasellar component, extension into neighbouring structures. Rare sites: intranasal, sphenoid sinus, cerebello-pontine angle	Suprasellar or within the 3rd ventricle
Presentation	Visual disturbances	Visual disturbances
	Endocrine deficiencies (e.g. hypopituitarism and diabetes insipidus)	Obstructive hydrocephalus
	Headache	Headache
	Cognitive dysfunction	Cognitive dysfunction
Imaging and pathological features		
General imaging features	Supra/intrasellar, multilobulated and multicystic mass	Usually suprasellar, mostly solid and spherical mass
MR	T1: Solid regions hypo- or iso-intense, cystic regions hyperintense. Strong, heterogeneous enhancement	T1: Hypointense. Cystic regions, if present, also hypointense. Moderate homogeneous enhancement
	Hyperintense on T2	Hyperintense on T2
CT	Solid regions and cyst wall enhance. Calcifications visible	Contrast enhancing with no calcifications
Macroscopic features		
Boundary	Lobular with sharp, irregular interface, adherent to surrounding structures, invasive	Encapsulated, discrete and often solid. No adherence to surrounding structure
Cysts	Cyst contents have dark, 'motor-oil' appearance with cholesterol crystals. Leakage can result in 'chemical meningitis'	When cystic, contents are clear
Calcification	Often present	Absent
Histopathological features		
Architecture	Multicystic, well circumscribed, but with finger-like protrusions into brain parenchyma	Discrete, encapsulated, often solid
Cellular/stromal composition	Peripheral pallisading epithelium	Squamous and well-differentiated, non-keratinizing epithelium
	Stellate reticulum comprising loose aggregates of stellate cells	Fibrovascular core, no stellate reticulum
	Nodules containing anuclear 'ghost cells'/'wet keratin'. Epithelial whorls with nuclear β-catenin expression	Pseudopapillae resulting from epithelial dehiscence, no 'ghost cells'/'wet keratin'. No nuclear β-catenin translocation
Other features	Piloid gliosis common in peritumoral brain. Encasement of blood vessels. Chronic inflammation, xhantogranulomatous reaction, occasional ossification	Scant goblet/ciliated cells in cyst lining, resembling Rathke's cleft cyst. Occasionally small, collagenous whorls
Molecular and pathogenic features		
Wnt pathway	Mutations in *CTNNB1* at S33, S37, S45 and T41	No mutations found
Odontogenic features	Enamelin, amelogenin and enamelysin expressed	Odontogenic markers not expressed





Tedavi

1) Cerrahi eksizyon
- Total…etraf dokulara zarar verme riski
- Subtotal

2) Kist aspirasyonu

3) Adjuvant kemoterapi
- İntrakistik bleomisin uygulaması

4) Radyoterapi
- Subtotal eksizyonlarda
- İntralezyoner

Prognoz

- Geniş serilerde 10 yıllık rekürrenssiz sağkalım %60-93, 10 yıllık overall sağkalım %64-96 bulunmuş.
- KF da nüksle ilişkilendirilen en önemli faktör cerrahi rezeksiyonun genişliğidir.
  - 5cm üzeri tümörlerde
  - Etraf dokuya infiltre olmalarından dolayı AKF'da prognoz daha kötüdür.
  - Yüksek Ki67 ile tümör rekürrensi korele

* KF da malign transformasyon son derece nadirdir ve şu ana kadar bildirilmiş 20 vaka bulunmaktadır.
- Malign transformasyonun nedeni radyoterapi mi?

*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Editor: James I. Ausman, MD, PhD University of California, Los Angeles, CA, USA

Case Report

Sequential pathological changes during malignant transformation of a craniopharyngioma: A case report and review of the literature

Tetsuya Negoto, Kiyohiko Sakata, Takachika Aoki, Kimihiko Orito, Shinji Nakashima[1], Masaru Hirohata, Yasuo Sugita[1], Motohiro Morioka

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Case Report

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Tetsuya Negoto, Kiyohiko Sakata, Takachika Aoki, Kimihiko Orito, Shinji Nakashima[1], Masaru Hirohata, Yasuo Sugita[1], Motohiro Morioka

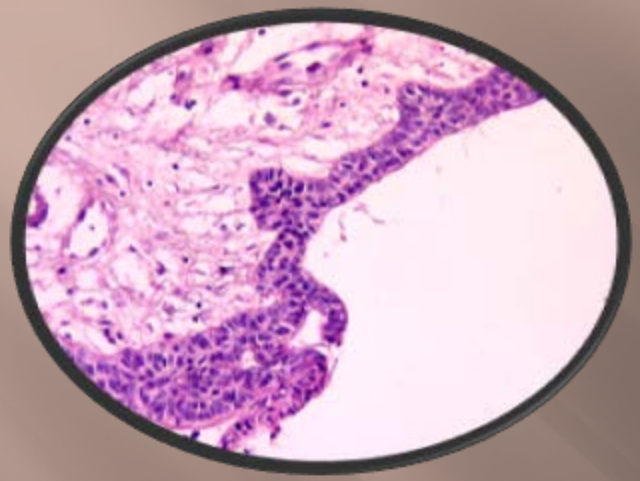
Table 1: Summary of the reported 20 cases of malignant transformation of craniopharyngioma

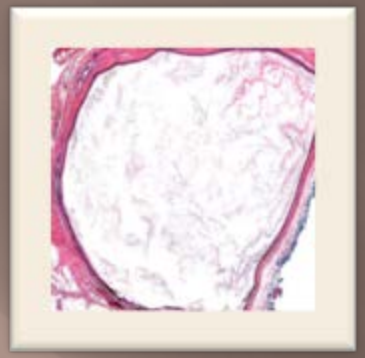
Author, year	Sex	Age (presentation)	Time to MT (year)	Radiation (+/−)	Therapy after MT	Outcome/duration	Histology (authors' description) (at first/at MT)
Akachi *et al.* 1987	F	7	3	+	Surgery(2), RT	Dead/8 m after MT	AC/MT
Nelson *et al.* 1988	F	14	3	+	Surgery	Dead/11 w after surgery	AC/MC
Suzuki *et al.* 1989	M	3	8	+	RT, V P shunt	Dead/2m after MT	AC/SCC
Suzuki *et al.* 1989	M	9	5	+	Surgery	Dead/3 m after MT	AC/SCC
Virik *et al.* 1999	M	24	10	+	Surgery. CT (carboplatin, etoposide), RT	Dead/10 m after MT	AC/UET
Kristopaitis *et al.* 2000	F	27	15	+	Surgery, CT (paclitaxel. earboplatin)	Alive/6m after MT	AC/SCC
Sakai *et al.* 2004	M	3	14	+	Surgery (5), stereotactic radiosurgery (2)	Dead/3 y after MT	-/MT with PP
Plowman *et al.* 2004	F	6	15	+	Surgery, CT(cisplatin, etoposide)	Dead/6m after MT	AC/MT without SF
Yue *et al.* 2006	M	17	0	-	details unknown	details unknown	None※1/SCC
Rodriguez *et al.* 2007	M	31	0	-	Surgery	Dead/6 w after surgery	None※1/OGC
Rodriguez *et al.* 2007	F	58	5	+	Surgery, shunt	Dead/2 m after MT	AC/SCC
Rodriguez *et al.* 2007	M	14	8	+	Surgery	Dead/1 y after MT	AC/MEC
Boongird. *et al.* 2008	F	46	0	-	Surgery	Dead/6 w after surgery	None※1/MC
Ishida *et al.*, 2010	M	6	5	+	Surgery, CT (cydophosphamide, cisplatia, etopoade)	Alive/10 m after MT	AC/MT
Aquilina *et al.*, 2010	M	4	8	+	Surgery (2), CT (carboplatin. cetuximab)	Dead/6 m after MT	-/SCC
Aquilina *et al.* 2010	F	6	7	+	Surgery, CT (paclitaxel .earboplatin), RT	Alive/5 y after MT	AC/SCC
Ujifuku *et al.* 2010	M	32	10	+	Surgery (2)	Dead/43 d after surgery	AC/SCC
Launola *et al.* 2011	F	66	0	-	RT	Dead/15 m after surgery	None*1/MC
Gao *et al.* 2011	F	41	4	-	Surgery	Dead/3 5m Mfrer cuegery	MIX/ABC
Present case	M	12	24	+	Surgery	Alive/12 m after surgery	AC/MT

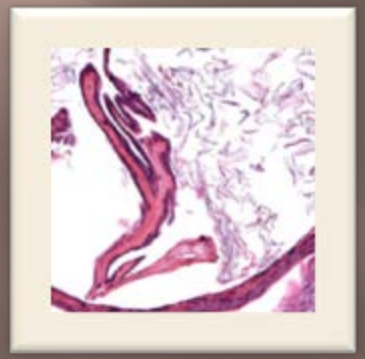
ABC: Ameloblastic carcinoma; AC: Adamantinomatous craniopharyngioma; MC: Malignant craniopharyngioma; MEC: Myoepithelial craniopharyngioma; MIX: Mixed type craniopharyngioma; MT: Malignant transformation; OGC: Odontogenic ghost cell carcinoma; PC: Papillary craniopharyngioma; PP: Pleomorphic pattern; RT: Radiation therapy; SCC: Squamous cell carcinoma; SF: Squamous features; UET: Undifferentiated epithelial tumor; d: Day; m: Month; y: Year; ※1: De novo tumor

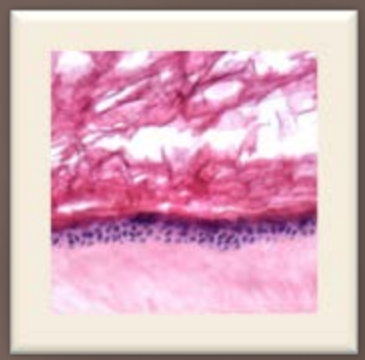
NELERLE KARIŞABİLİR?

- Epidermoid kist?





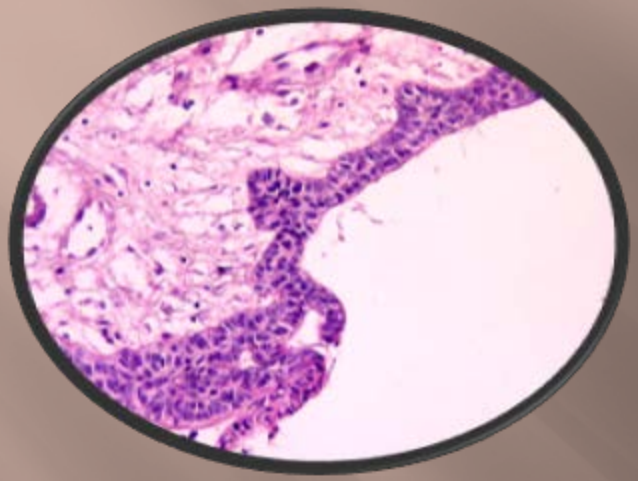


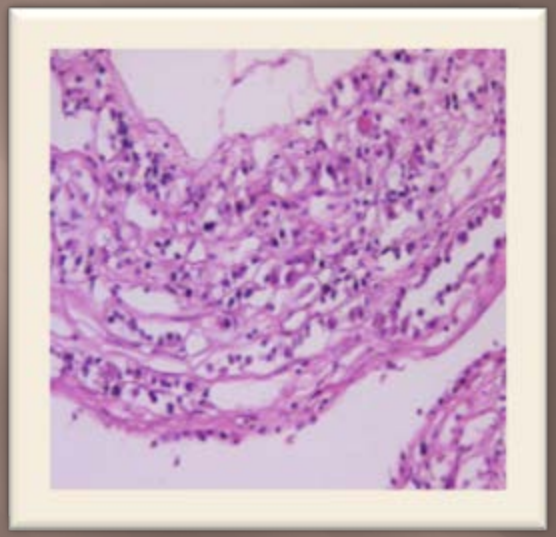


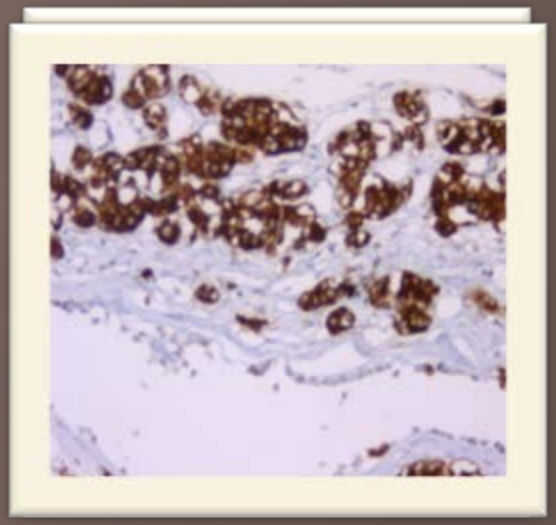
- Keratohyalin granülleri
- Kuru keratin izlenir,
- Islak keratin ve nükleer palizat izlenmez

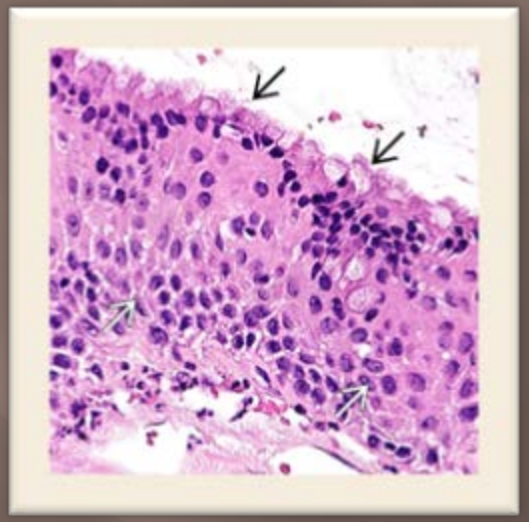
NELERLE KARIŞABİLİR?

Adamantinamatöz KF ▣ Rathke Cleft Kisti ?

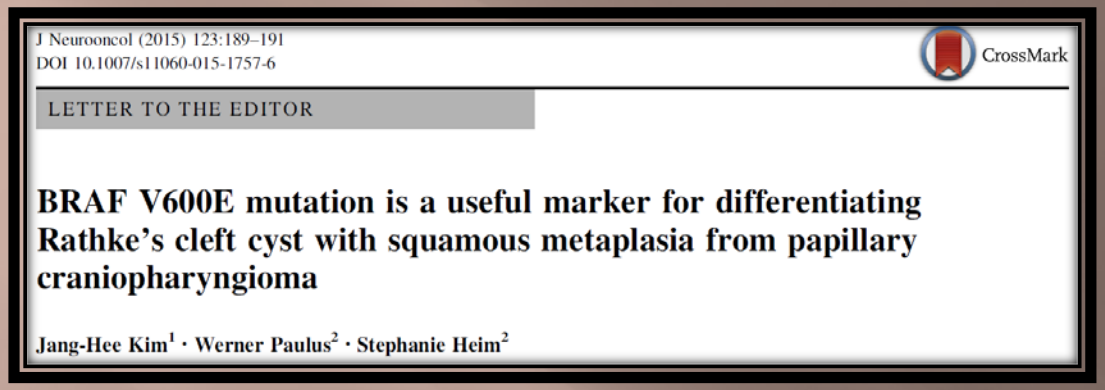








- Suprasellardan ziyade intrasellardır.
- Genelde fokal de olsa silialı ve/veya müsinöz epitel vardır.
- Islak keratin ve kalsifikasyon izlenmez
- Beta katenin nükleuslarda negatif
- CK8 ve CK20 (+)

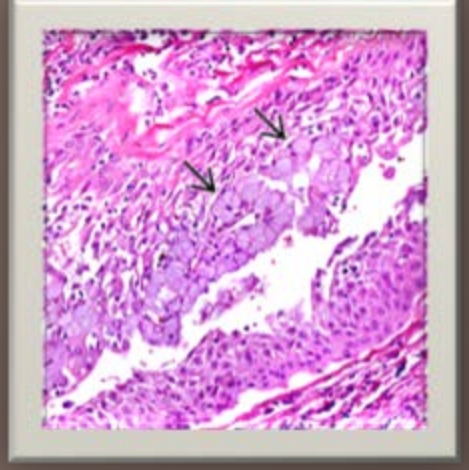

J Neurooncol (2015) 123:189–191
DOI 10.1007/s11060-015-1757-6

CrossMark

LETTER TO THE EDITOR

BRAF V600E mutation is a useful marker for differentiating Rathke's cleft cyst with squamous metaplasia from papillary craniopharyngioma

Jang-Hee Kim[1] · Werner Paulus[2] · Stephanie Heim[2]

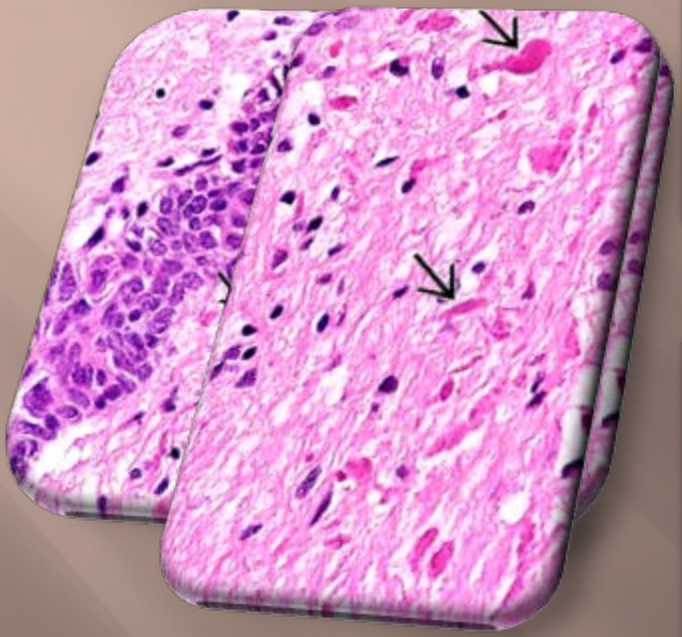


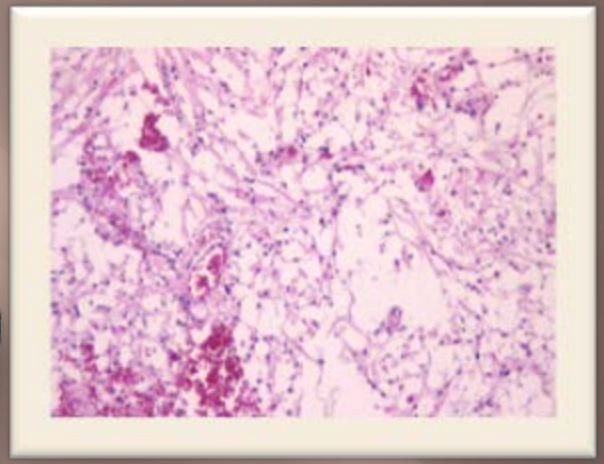
- Skuamöz metaplazi içeren Rathke Cleft Kisti ? X Silialı Papiller KF

❑ Benzer histopatoloji	Benzer histopatoloji
❑ Beta katenin nükleer (-)	Beta katenin nükleer (-)
	Transizyonel form?
	Orijini rathke kleft kisti midir?
❑ Braf V600E mutasyonu (-)	Braf V600E mutasyonu (+)

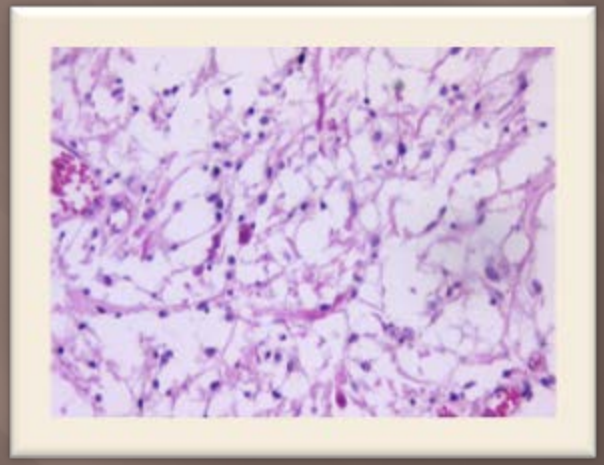
NELERLE KARIŞABİLİR?

- Pilositik astrositoma ?





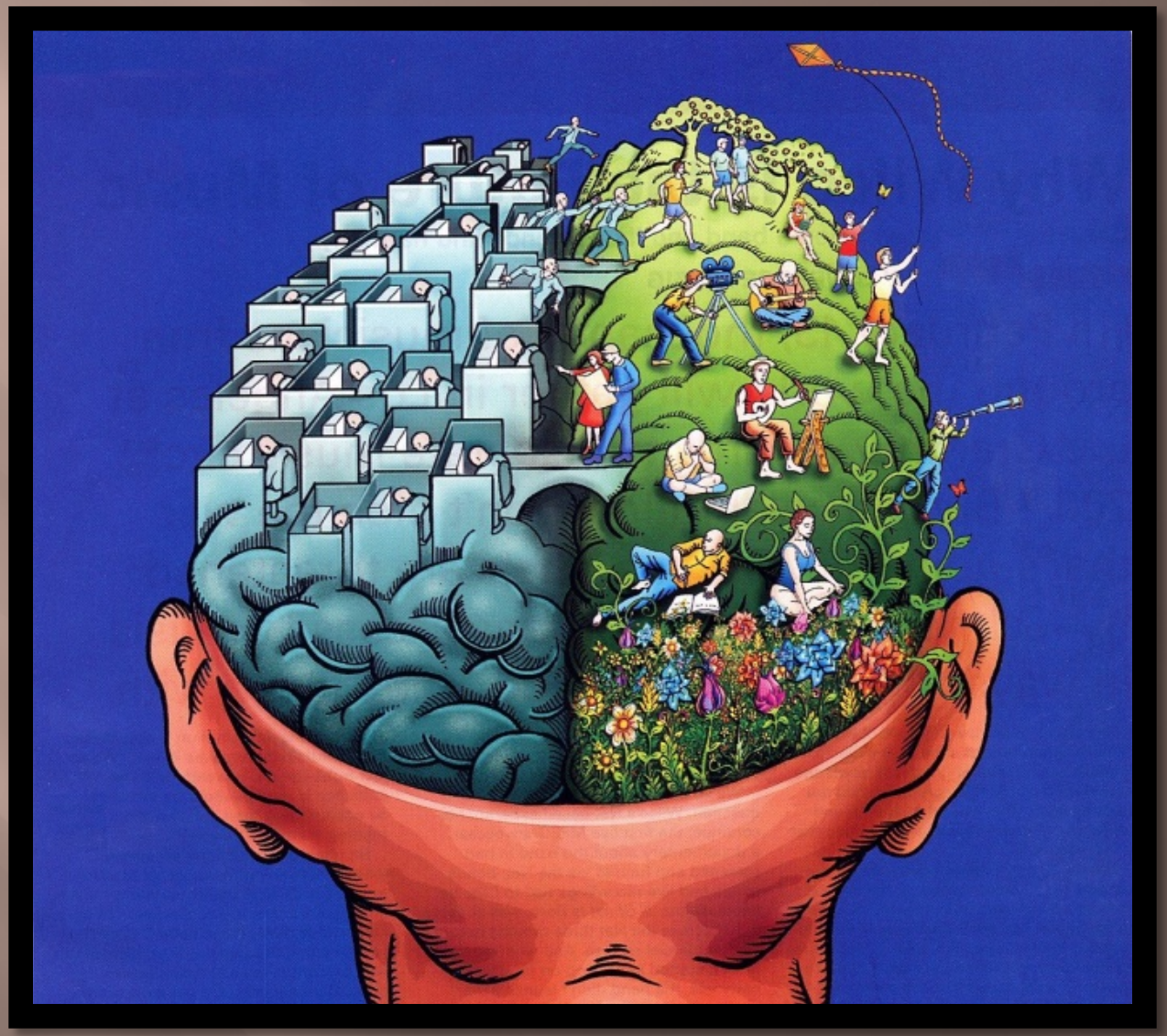
- Piloid gliozisten daha selüler



- Bifazik, mikrokistik

NELERLE KARIŞABİLİR?

- **Sellar bölgenin Ksantogranülomu**
- Reaktif bir proçes
- Kolesterol yarıkları, makrofajlar, dev hücreler, inflamasyon, hemosiderin, nekroz, ve kist
- Epitel yok



Teşekkürler…