

Retinoblastoma



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Epidemiología



- Tumor maligno Poco frecuente
- 1 en 20, 000 nacimientos
- Cambio en Mortalidad
 - Actualmente 95% Curación



- < 3 años
- Distribución universal
- Excepcional en adultos

Genética



Pérdida de
Alelos

Gen RB1

Cromosoma
13 q



Mutación Somática

- NO hereditario

Mutación germinal

- 5-10% que la portan NO desarrollan Rb
- 25-35% Unilateral
- 65-75% Bilaterales
- > Predisposición a cáncer
NO ocular

Diferenciación / Aspecto citológico



Retinoblastoma

Retinocitoma

Clínica



Leucocoria
90%

Estrabismo

**Masa
fungante**



Proptosis

Buphtalmos

Datos
neurológicos

Adenopatías





Figure 54-20 ■ Retinoblastoma. Clinical photograph shows proptosis, with eyelid and conjunctival inflammation from extraocular extension of retinoblastoma.



Figure 54-21 ■ Retinoblastoma. Fundus photograph shows a large mass involving the macular area and small tumors in the inferior retina.

Patrones macroscópicos



Endofítico



Exofítico



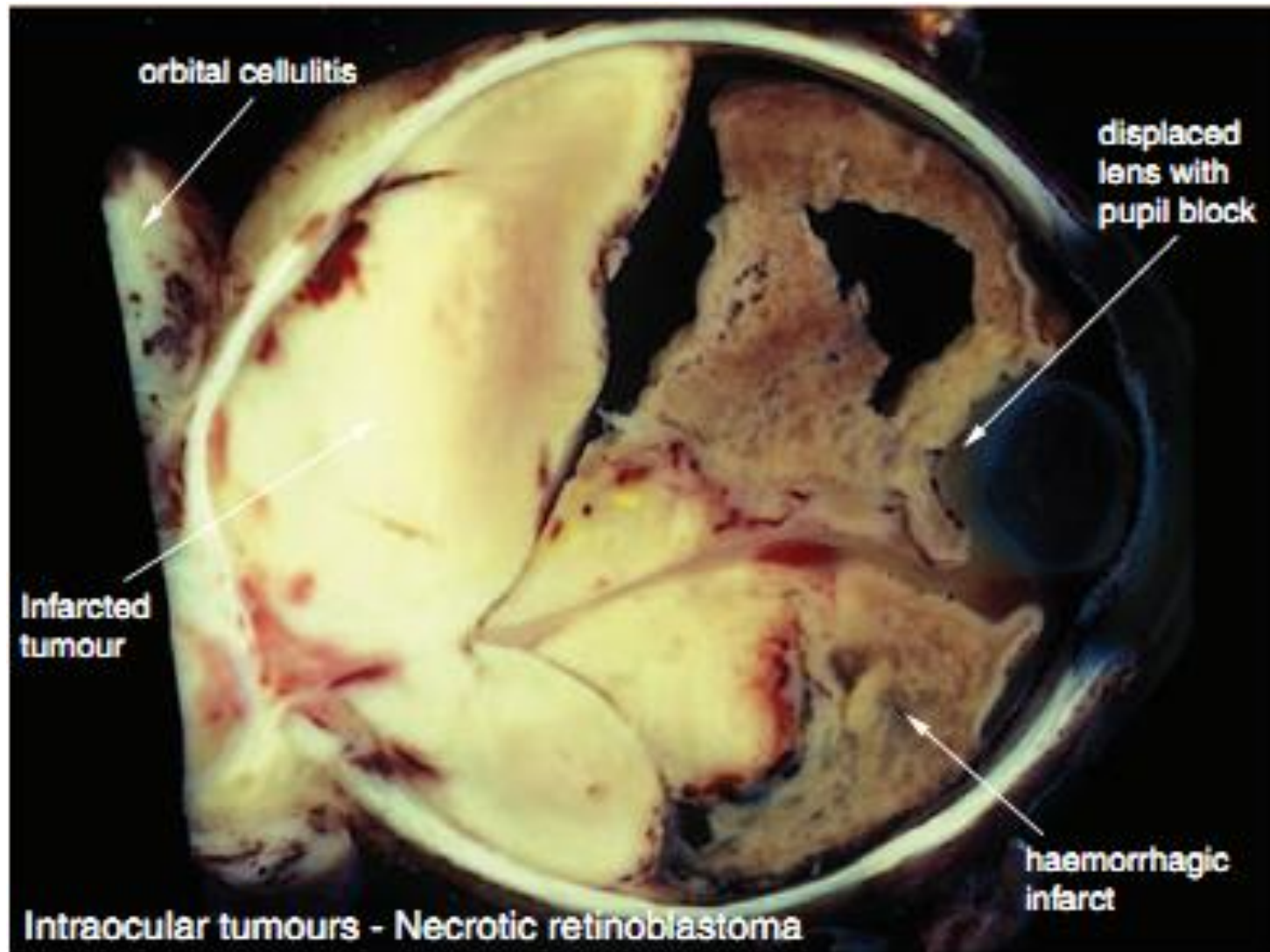
Difuso



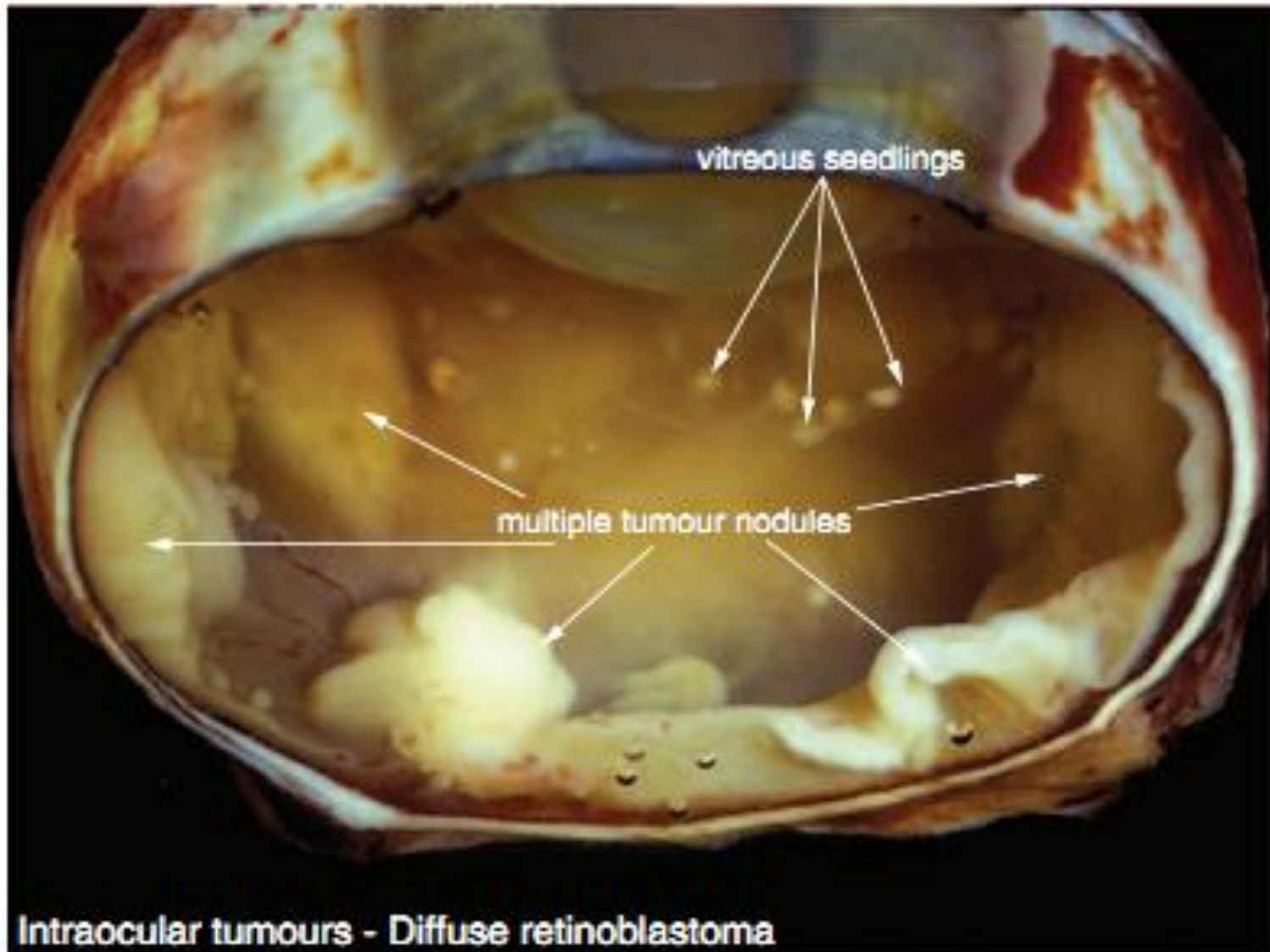
Mixto



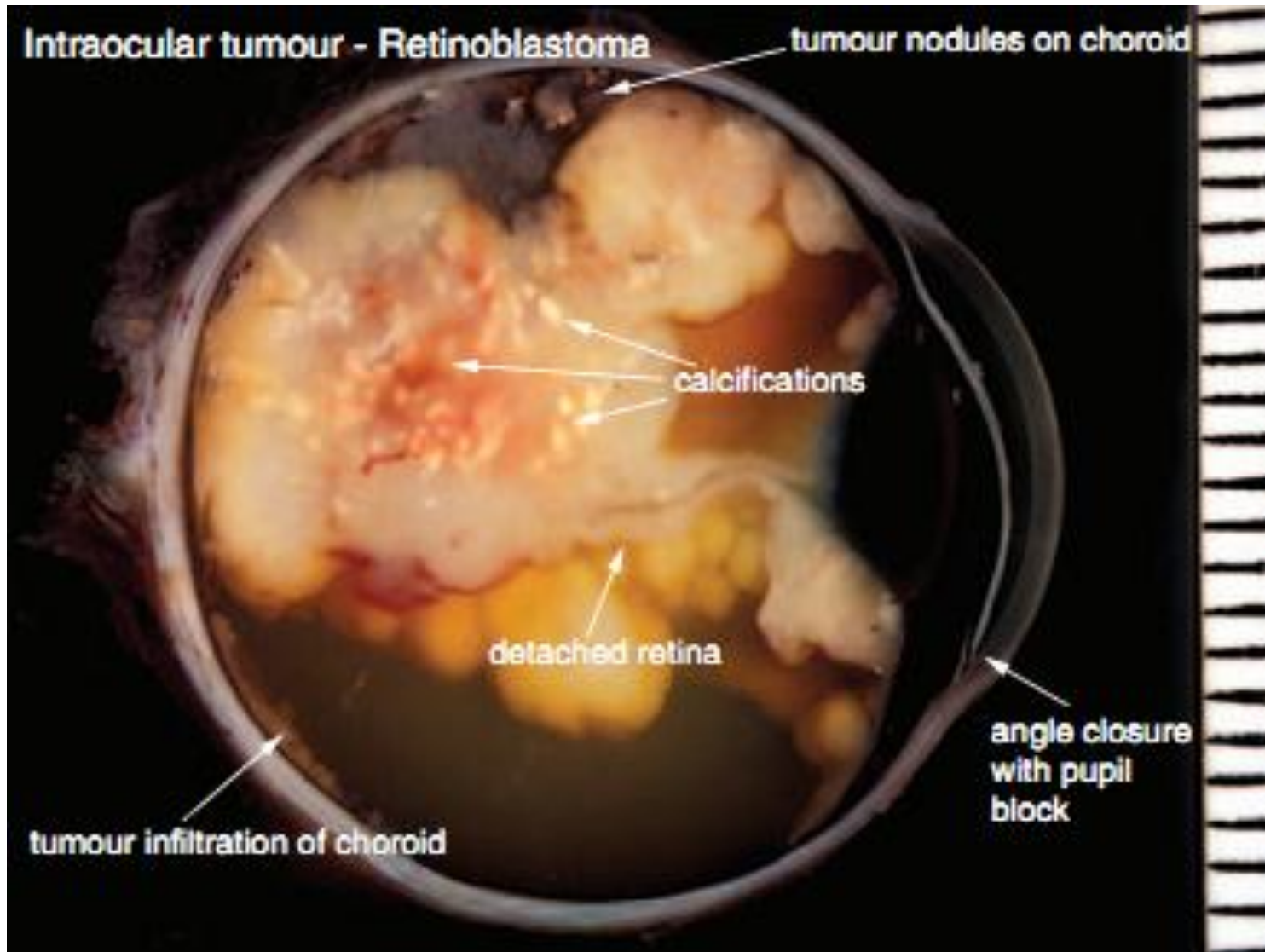
Regresión completa



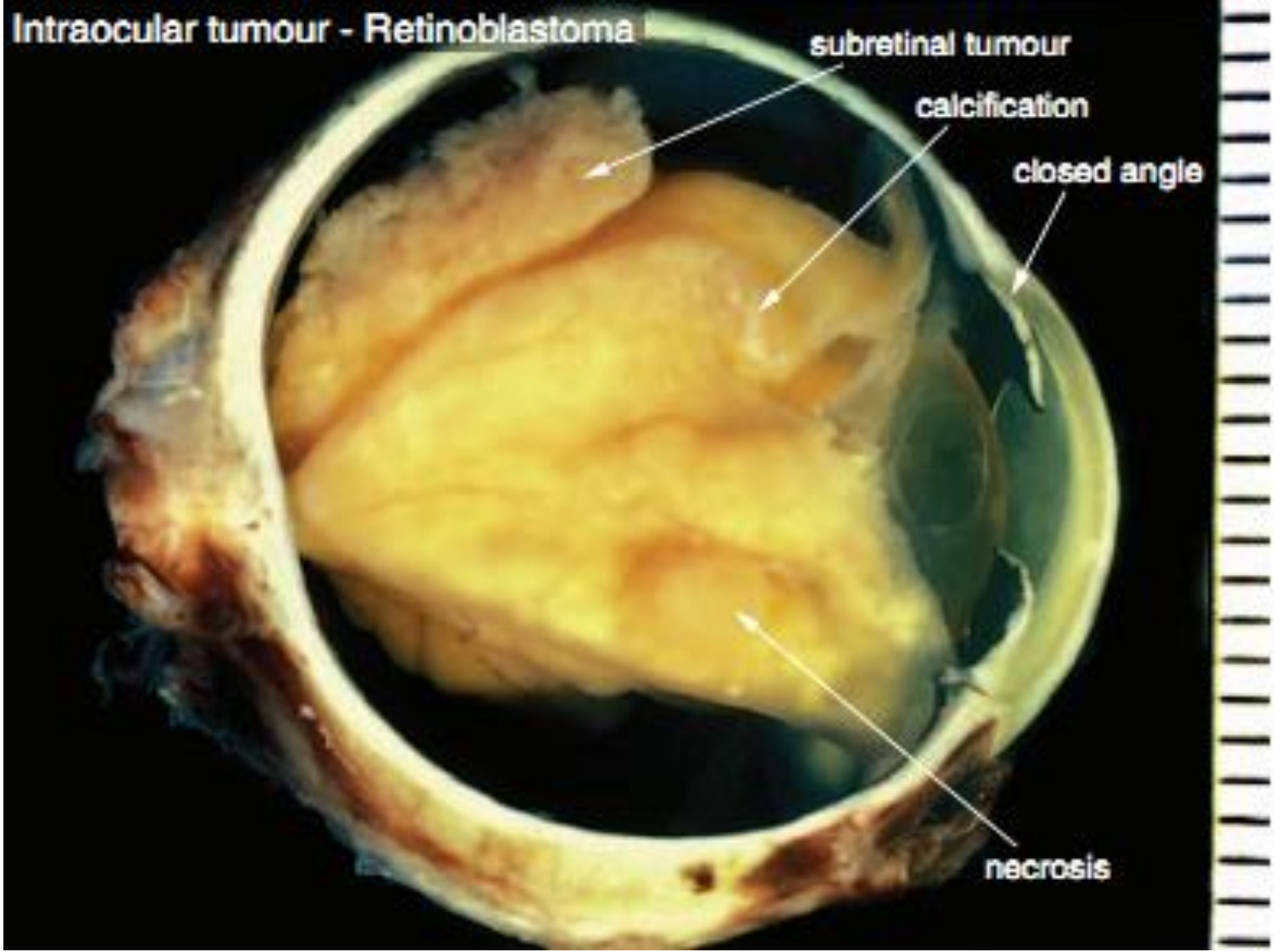
Intraocular tumours - Necrotic retinoblastoma



Intraocular tumours - Diffuse retinoblastoma



Intraocular tumour - Retinoblastoma



subretinal tumour

calcification

closed angle

necrosis



Histopatología



Tumor
neuroblástico

Células
citoplasma
escaso

Núcleo
basofílico
pleomórfico

Mitosis

Crecimiento /
Angiogénesis



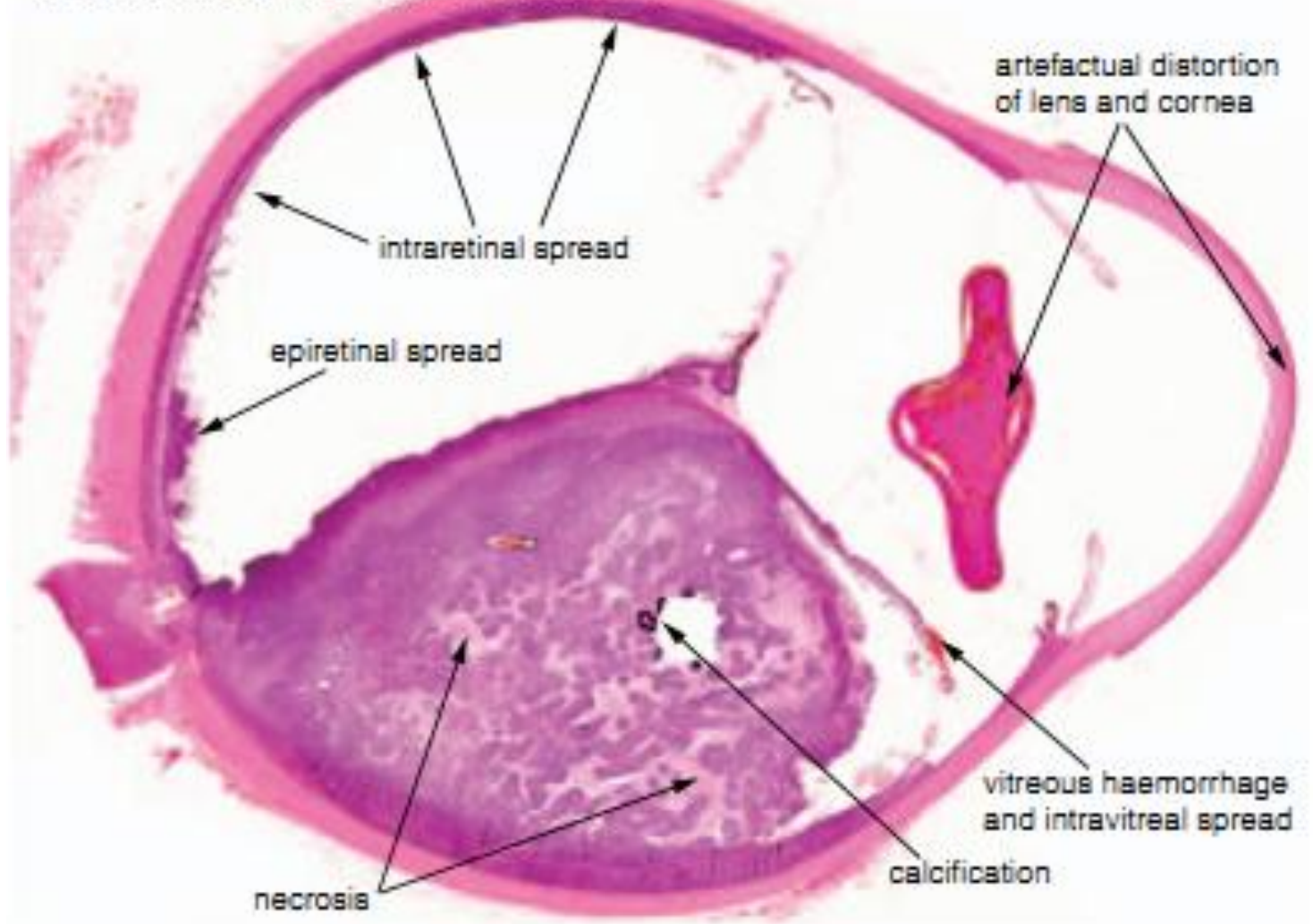
Necrosis
coagulativa

Focos de
calcificación

Rodean a
los vasos

Esferulas
neoplásicas
en vítreo

Intraocular tumour - Retinoblastoma





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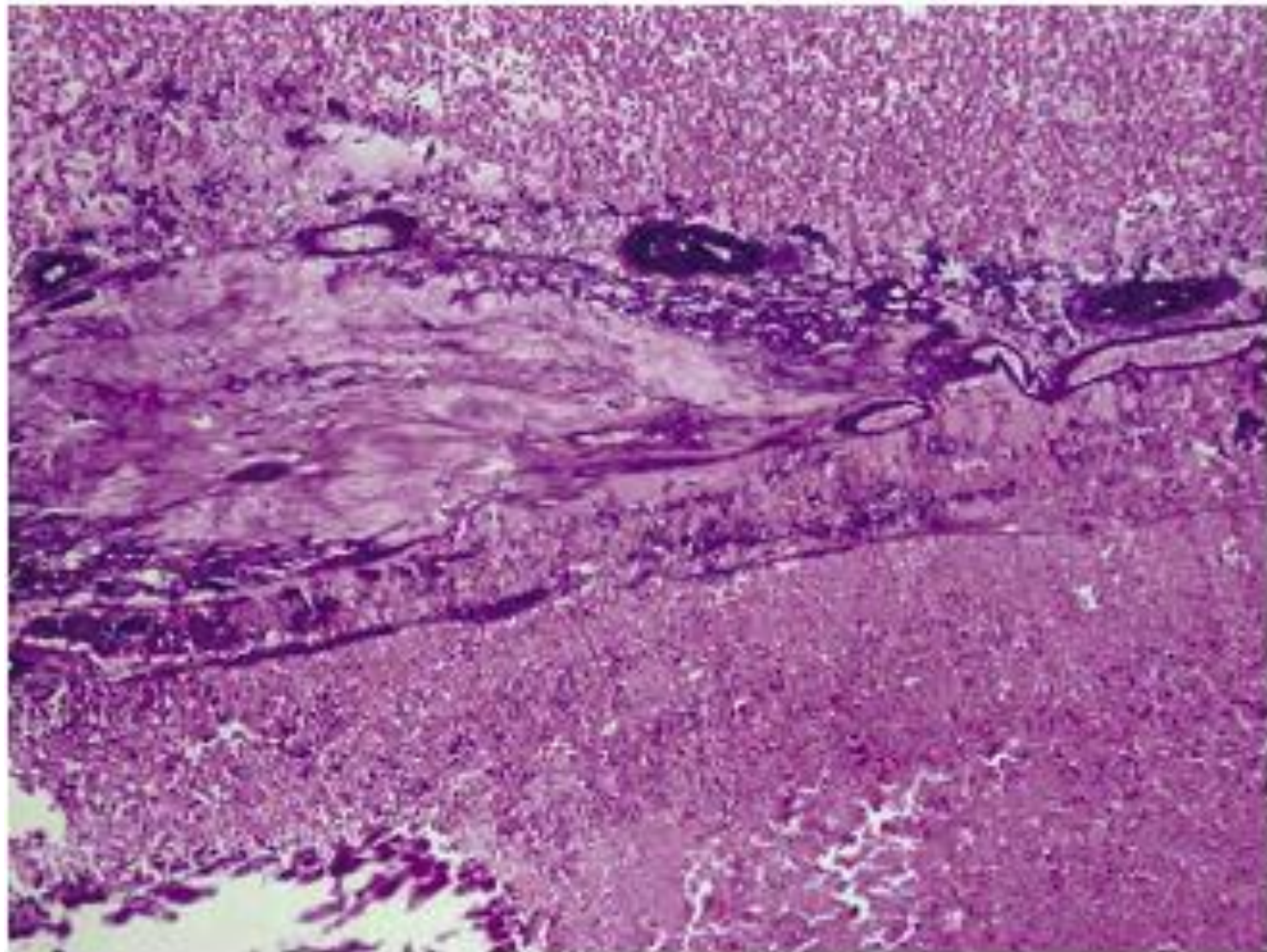
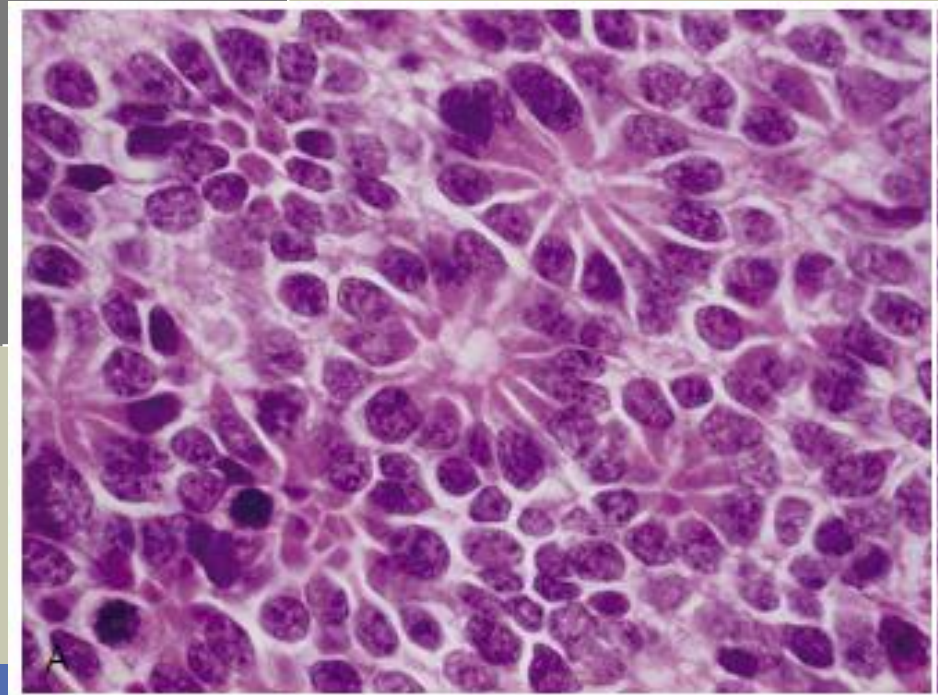


Figure 54-22 ■ Retinoblastoma. Basophilic staining involving tumor blood vessels. Note the extensive tumor necrosis and a focus of calcification.



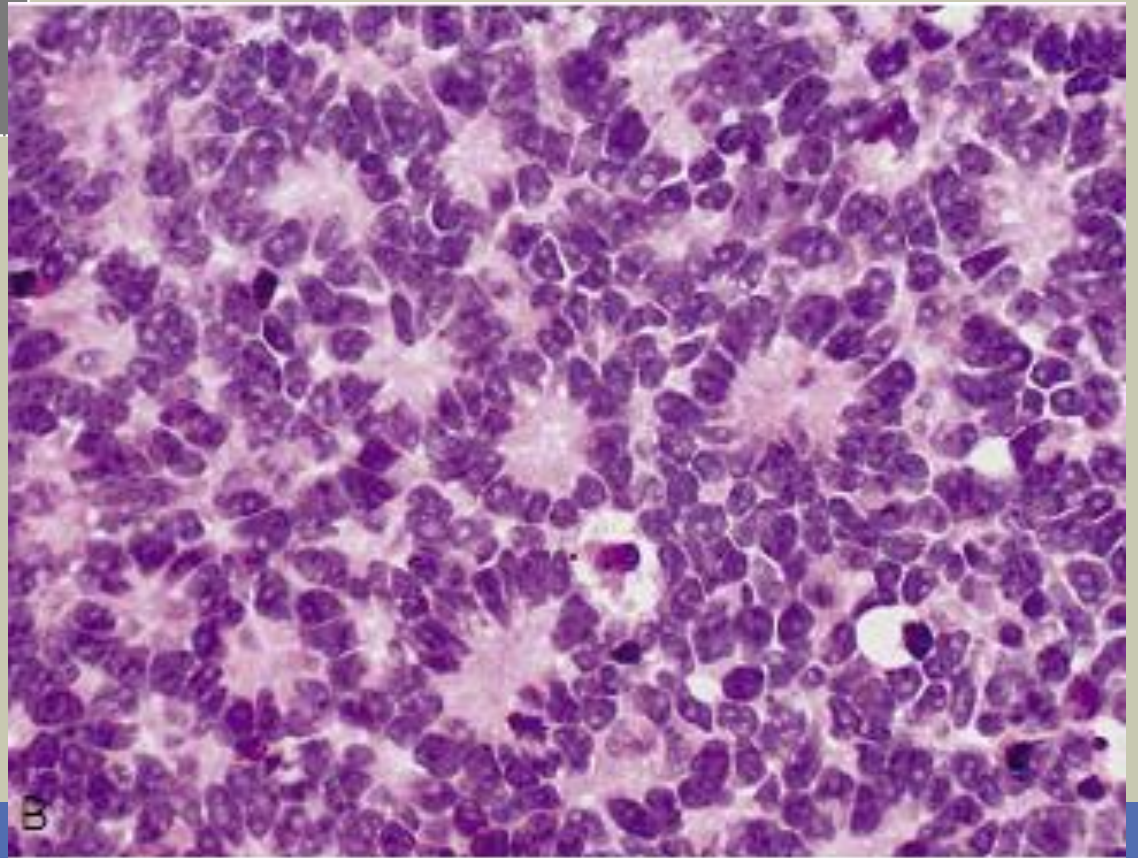
Rosetas Flexner- Wintersteiner, cél. Cubicas que rodean una luz

- Retinoblastoma
- Meduloepitelioma
- Pinealoblastoma





Rosetas Homer- Wright



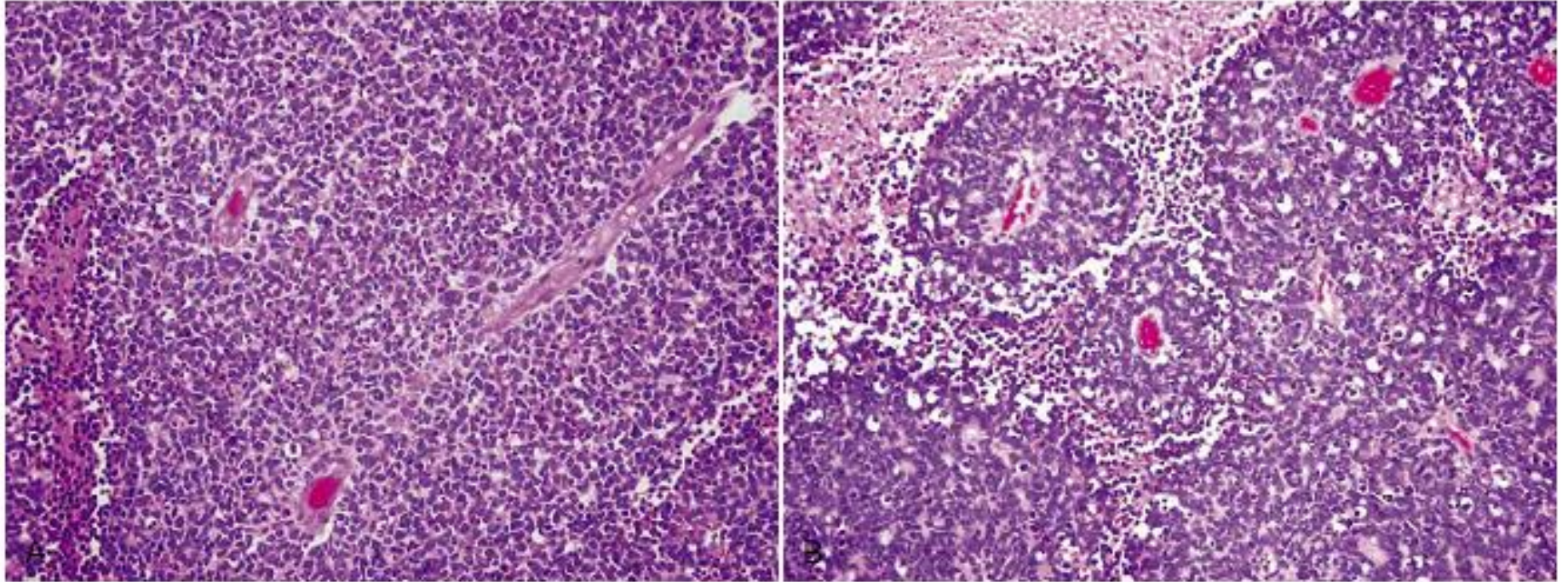


Figure 54-24 ■ Retinoblastoma. **A**, Undifferentiated retinoblastoma with tumor cells in a diffuse pattern and a necrotic area. **B**, Undifferentiated retinoblastoma forms pseudorosettes.

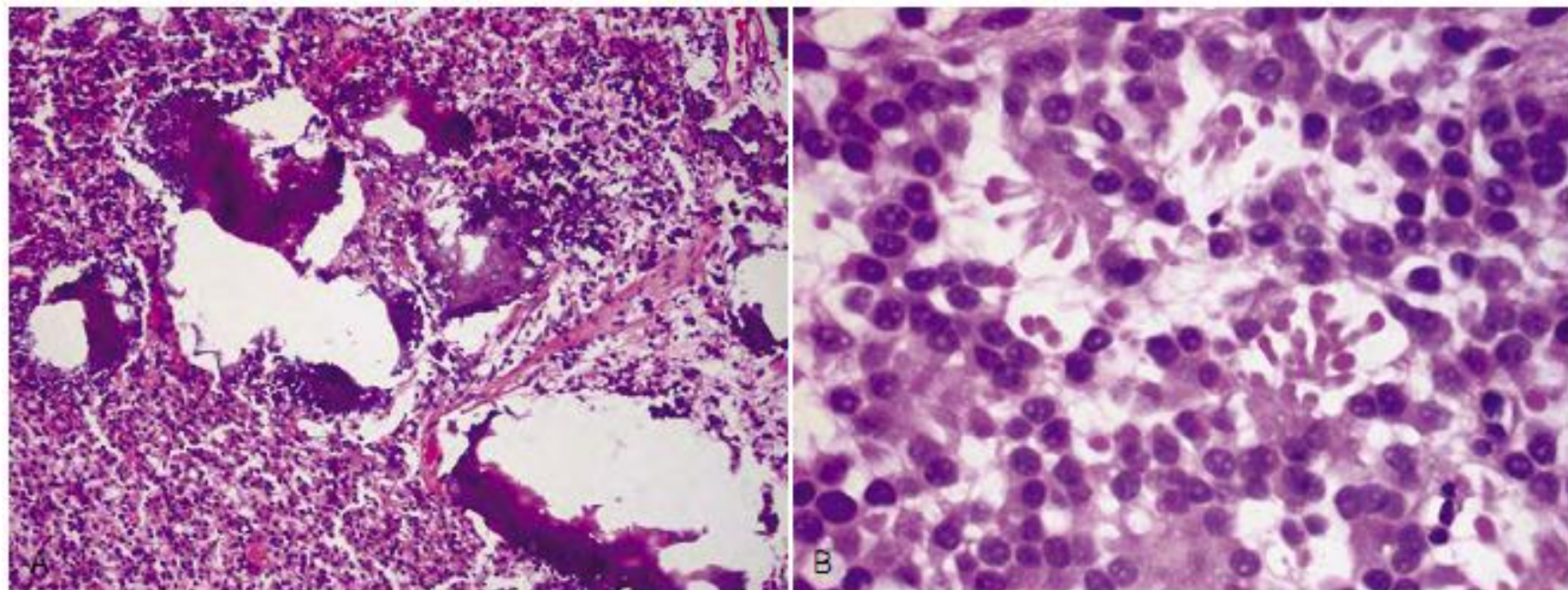


Figure 54-26 ■ Retinocytoma. **A**, Retinocytoma shows multiple foci of calcification. **B**, The retinal mass reveals fleurettes composed of benign-appearing cells.

Metástasis / Invasión



Crecimiento
rápido

Invasión

SNC : Disco
óptico

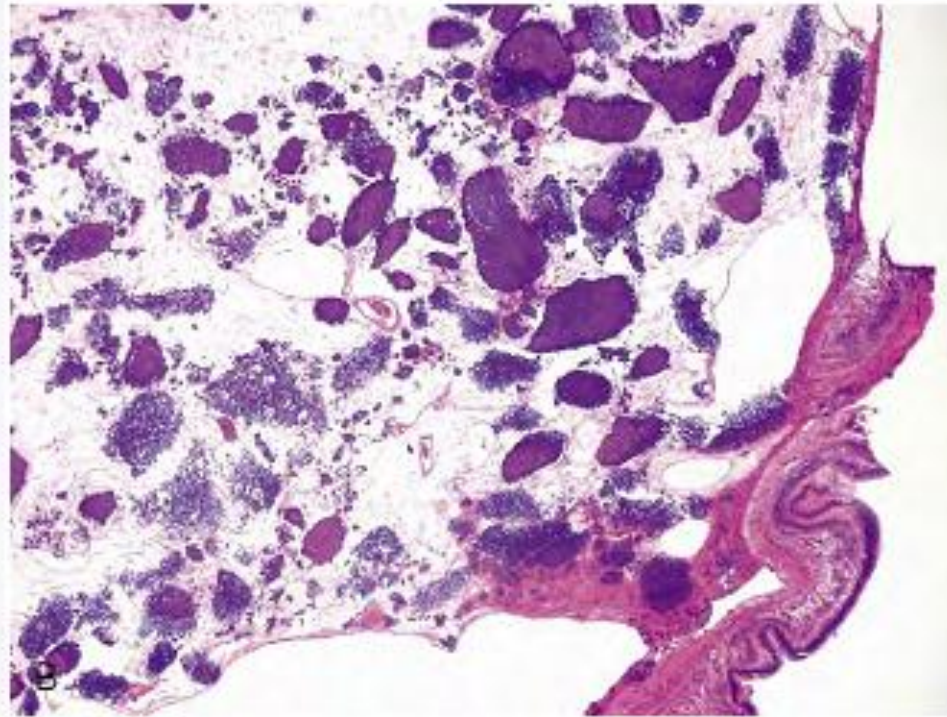
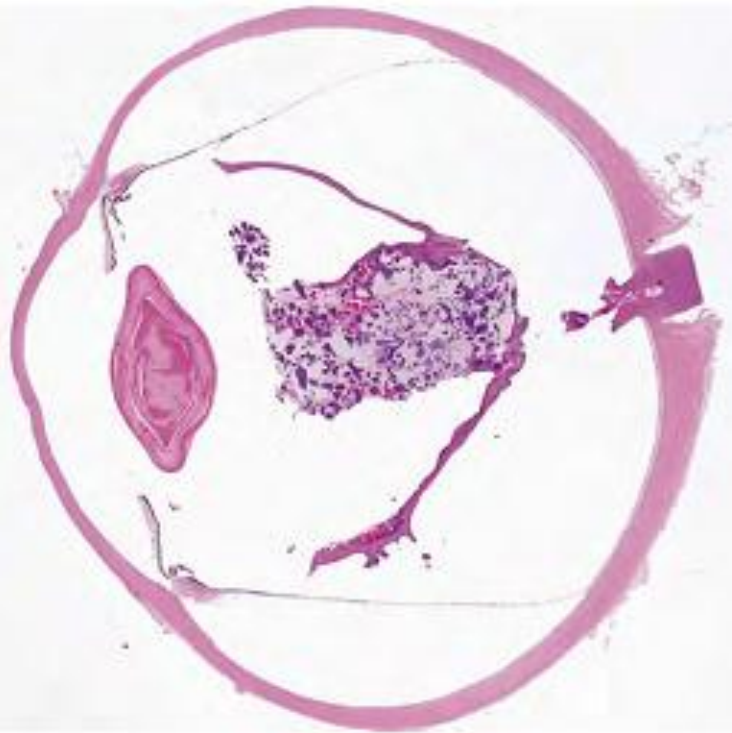


Figure 54-27 ■ Retinoblastoma. A, Retinoblastoma after chemoreduction therapy displays a calcified mass with areas of necrosis. B, Higher magnification reveals calcification.

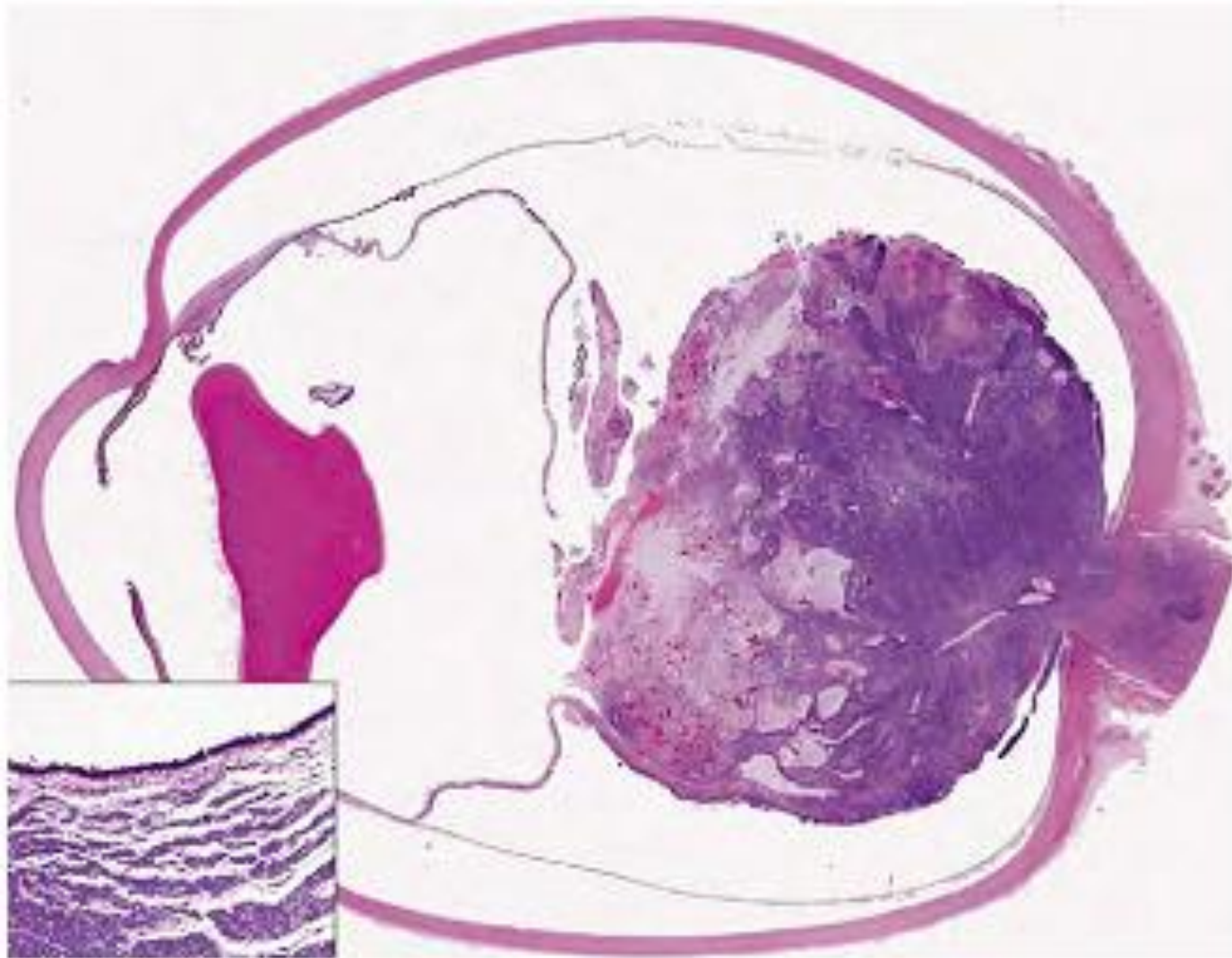


Figure 54-28 ■ Retinoblastoma. Histologic section of an eye with intraocular retinoblastoma and invasion of the optic nerve that extends into the post-lamina cribrosa. Note choroidal invasion in an eye with retinoblastoma (*inset*).



**Síntomas - Extensión
extraocular 6 meses**

Vías de diseminación

- Nervio óptico
- Leptomeninges
- Hematógena (pulmones y hueso)
- Linfática



Metástasis

1er año

Gral NO
rosetas

Pronóstico



**Factor >
Importante**

Invasión :

- Tunicas ojo
- Nervio óptico

Tratamiento



- Enucleación
- Radioterapia
- Quimioterapia