# Slide seminar
# Bone Pathology

## Case 5

# Clinical history:

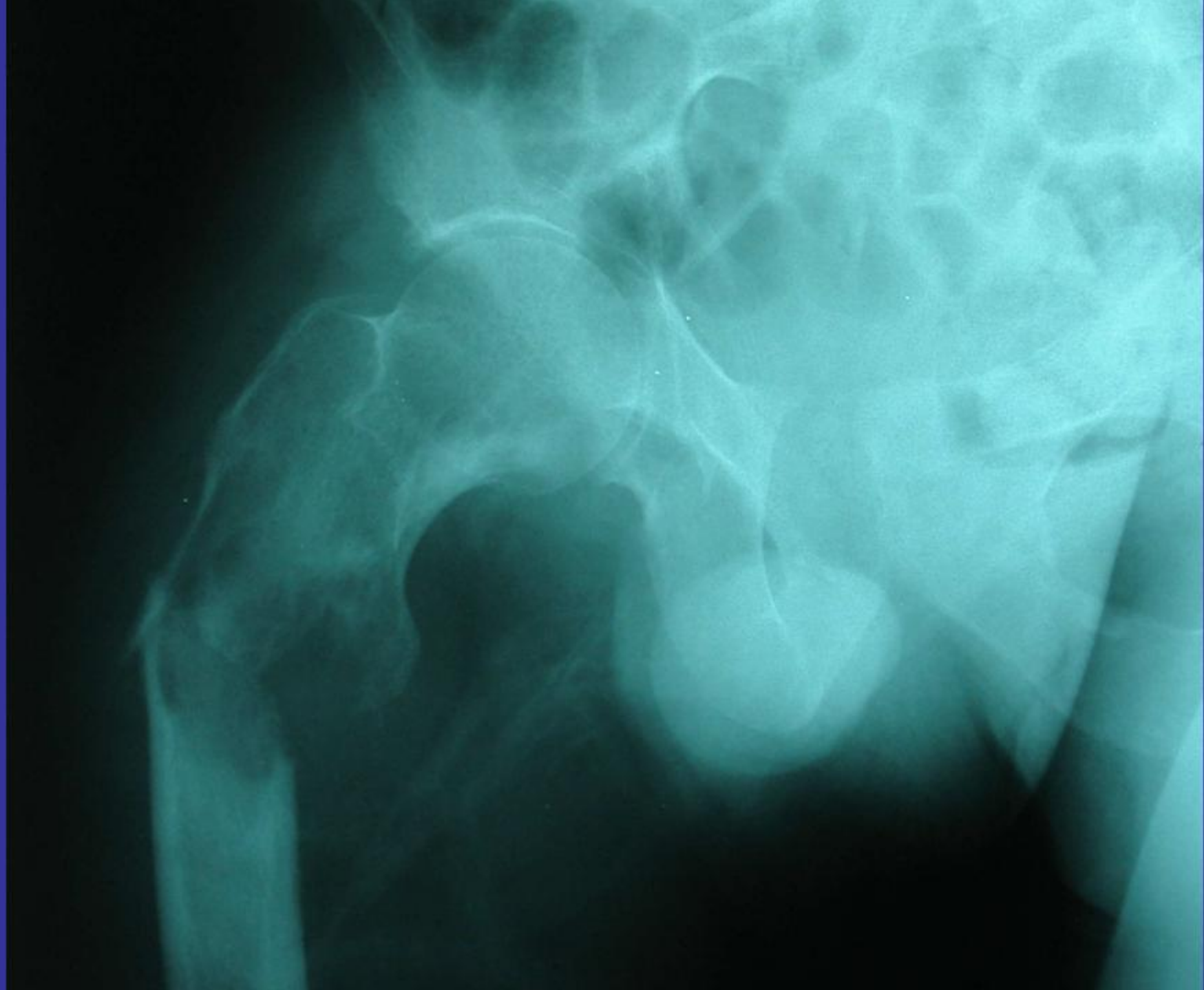
A 49 year-old man admitted in Orthopaedic Department of Constantine Hospital for :

Painfull swelling

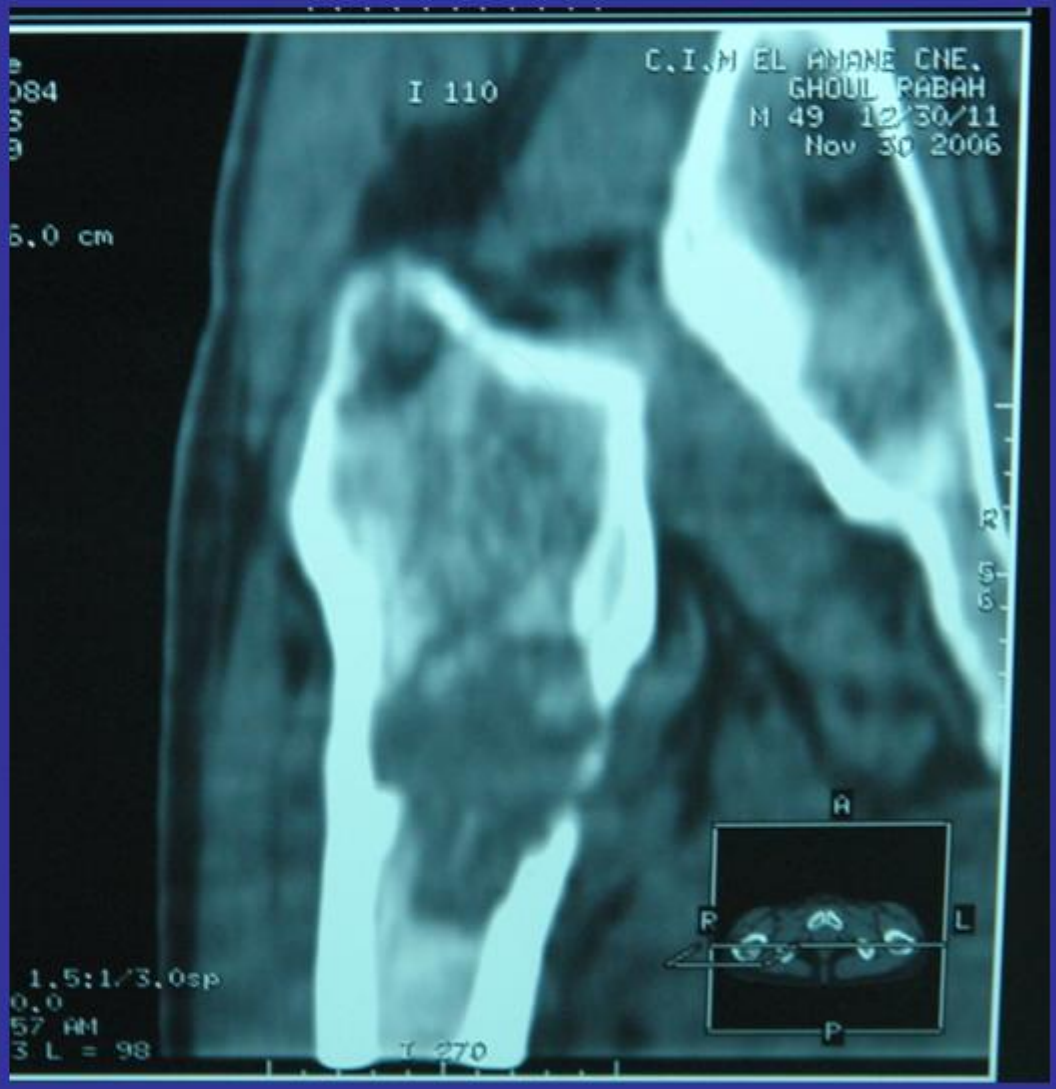
Pathologic fracture of the upper right femoral metaphysis.

# X-Ray and CT Scan:

Large lytic area  51 X 37 mm in upper metaphysis and diaphysis of the right femur accompagned by calcified mass of 3.7 mm in external hill side of the fracture.



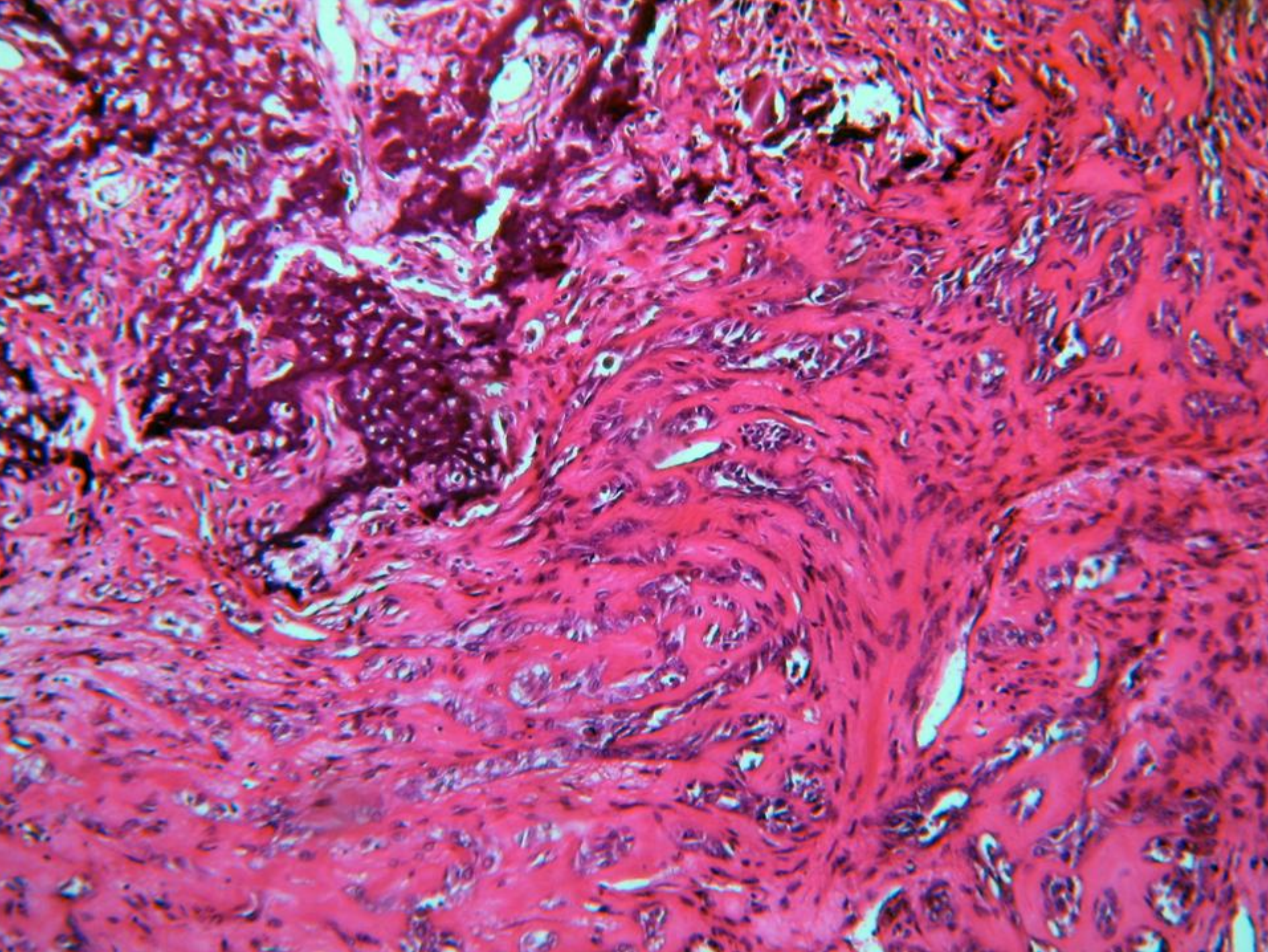
Grossly, the tumor is reddish, hard and focally calcified

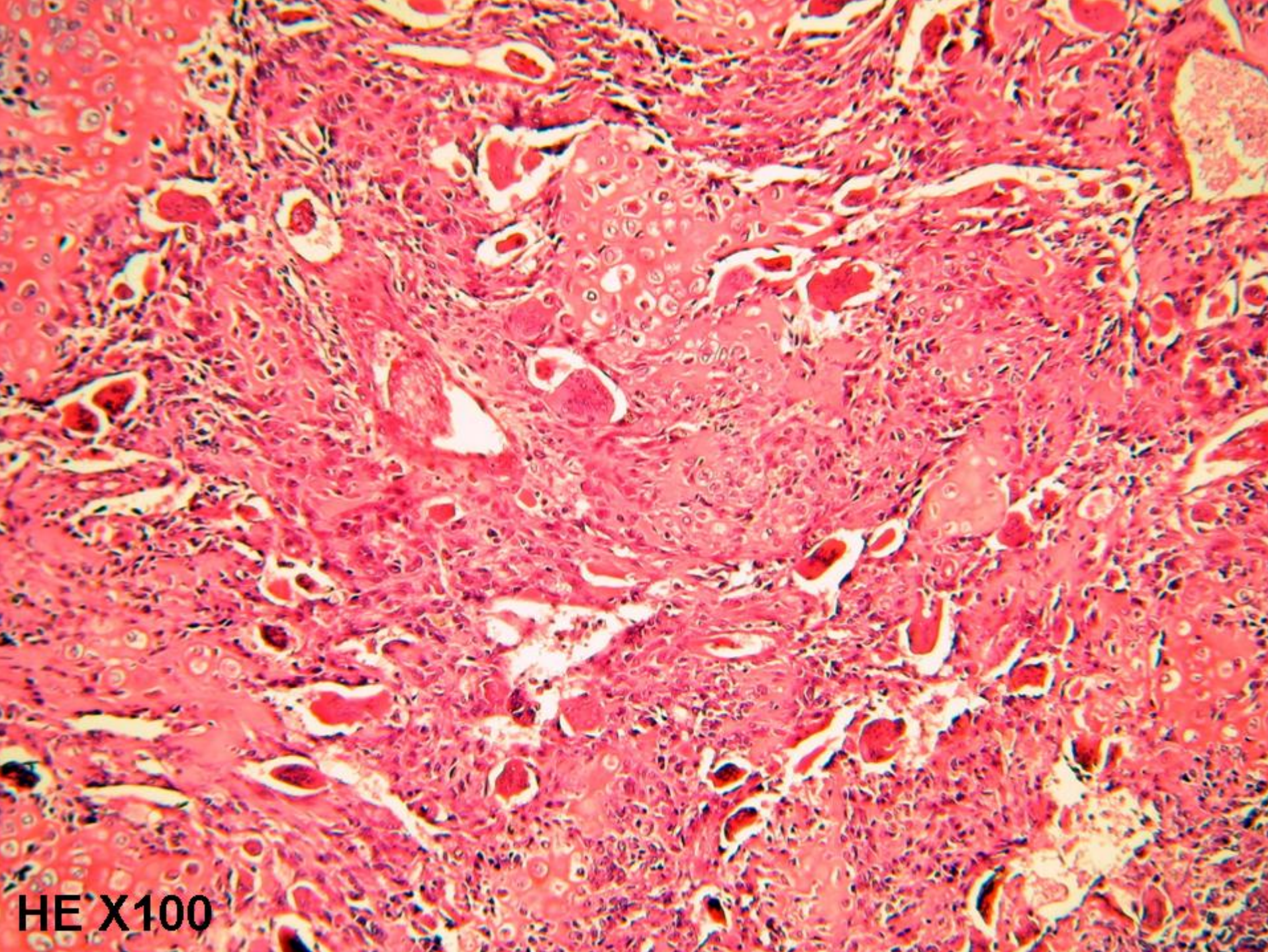


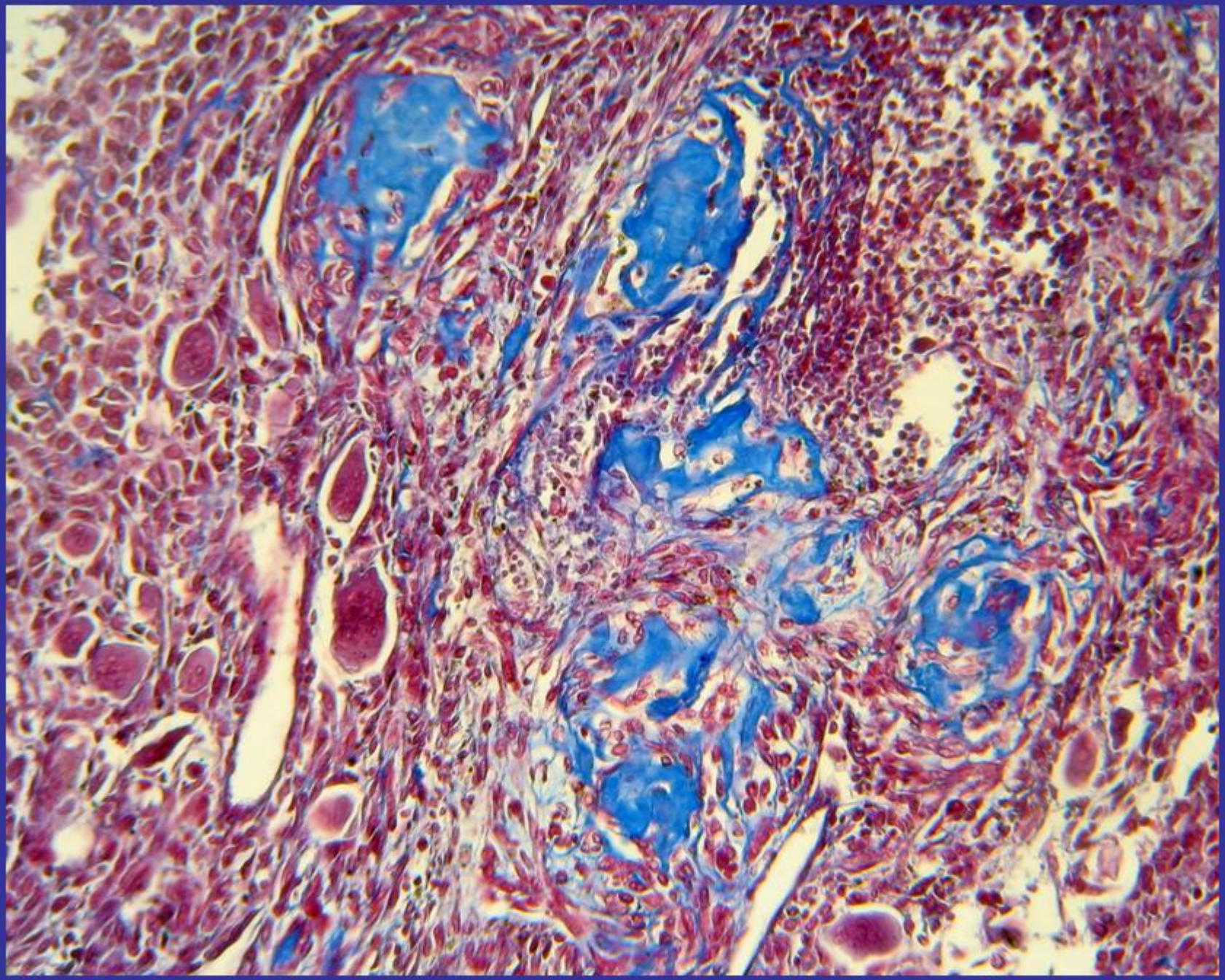
# Histological examination:

Mononuclear background with numerous giant cells and focally an epithelial component represented by tubes, solid nests, clusters, cellular cords and sometimes focal squamous differenciation

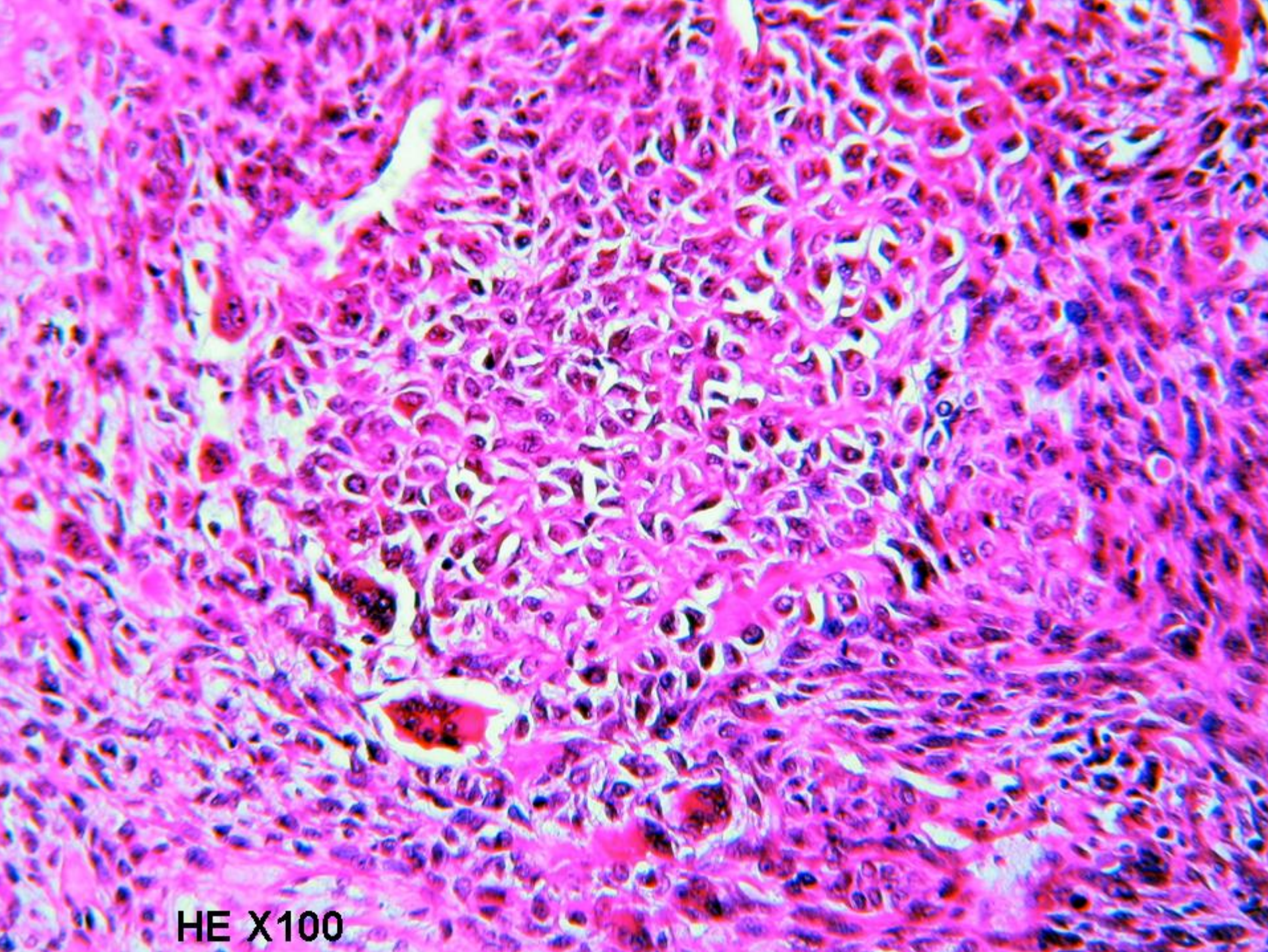
Osteoid and bone formation +++




HE X100



Masson X250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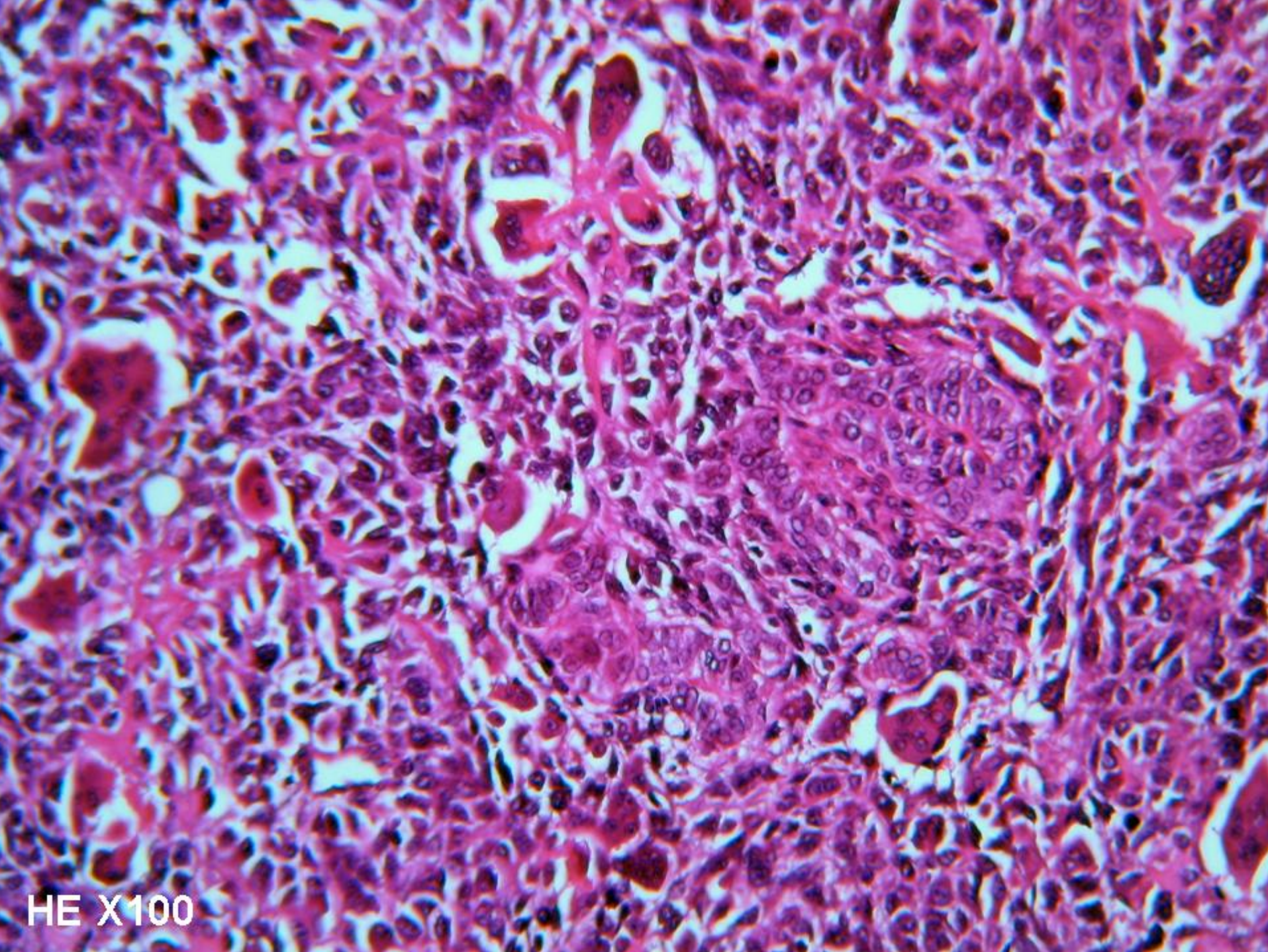
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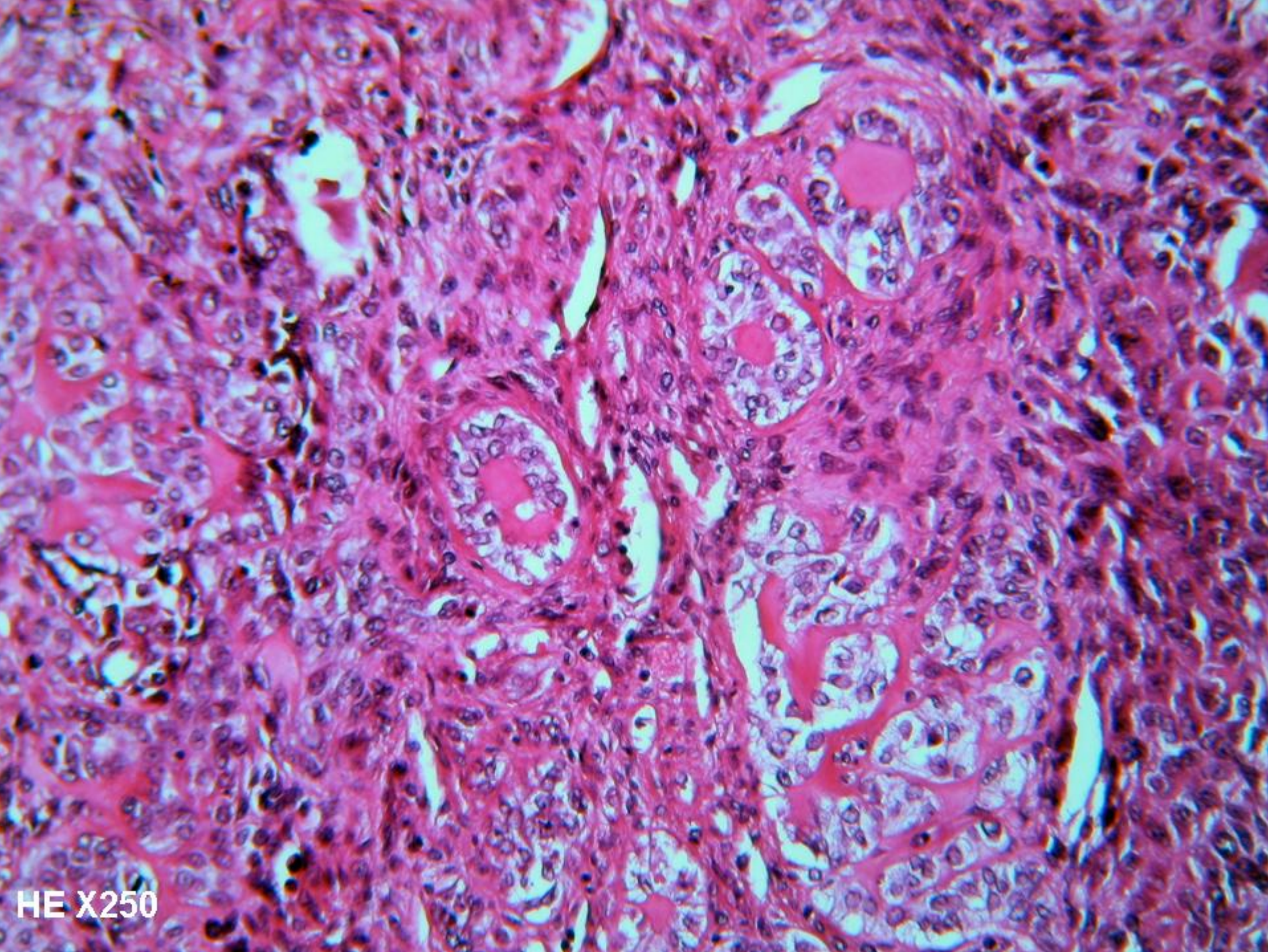
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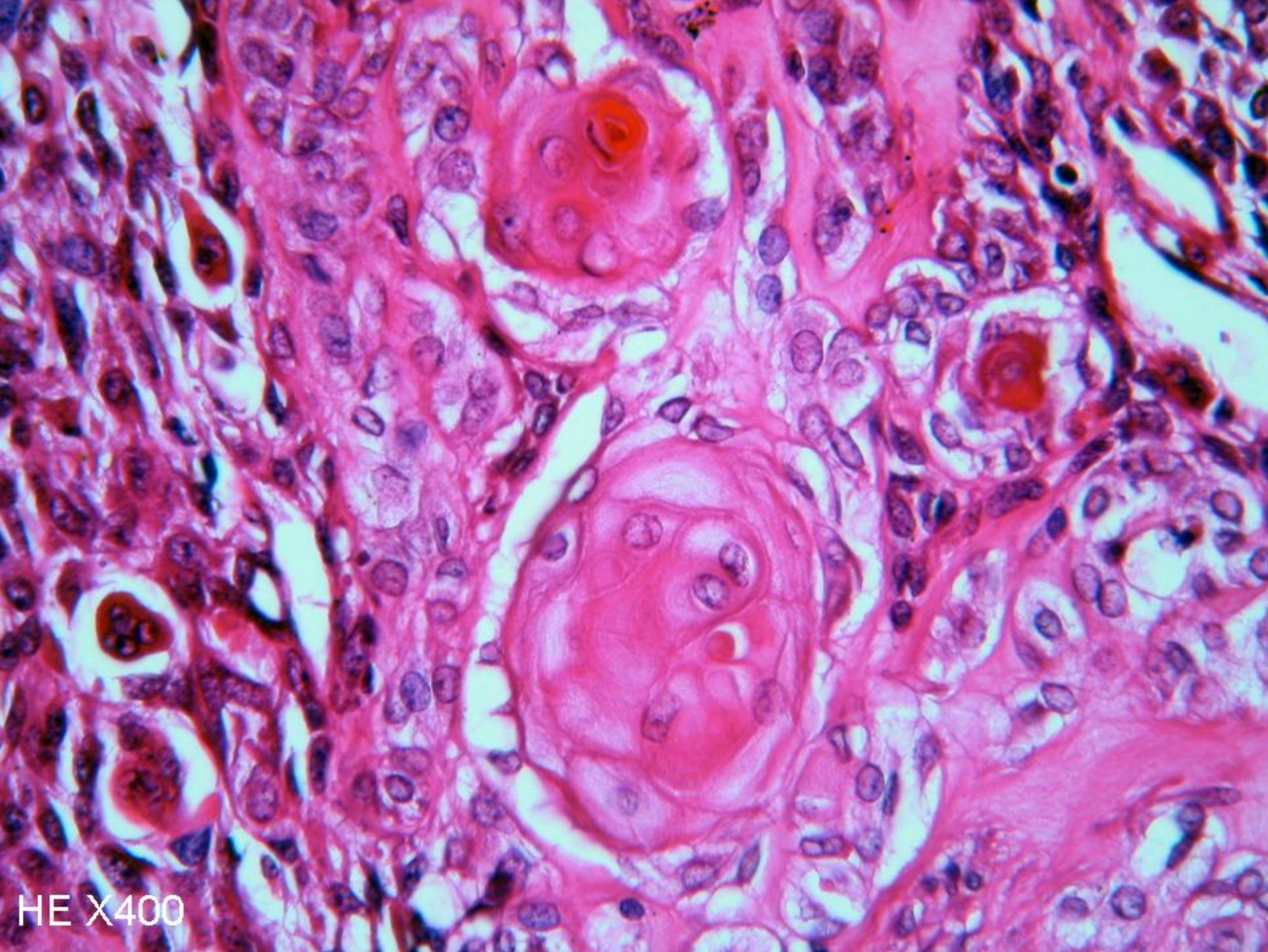
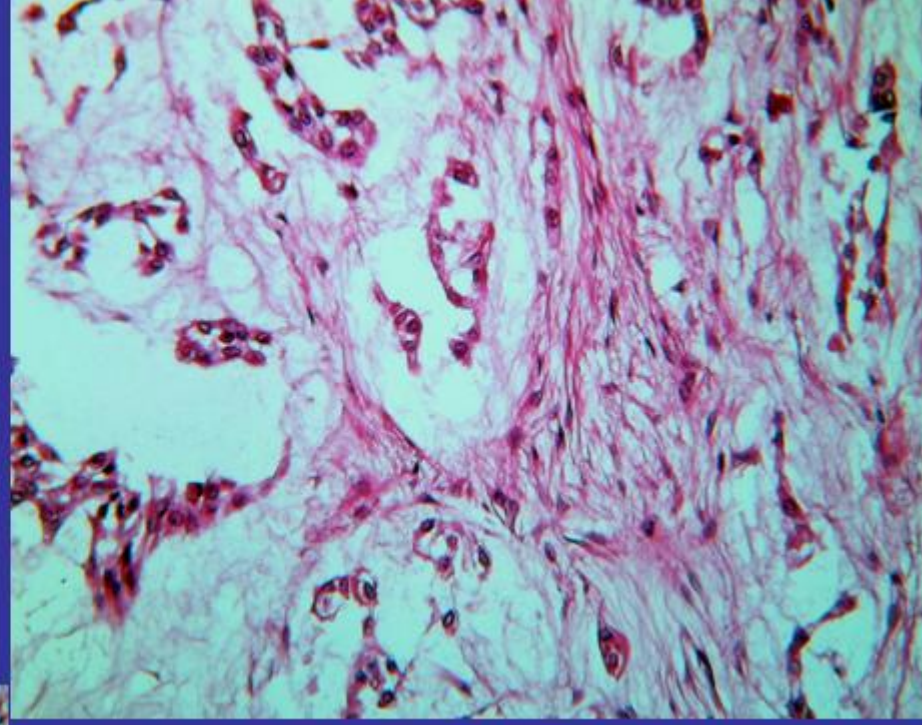
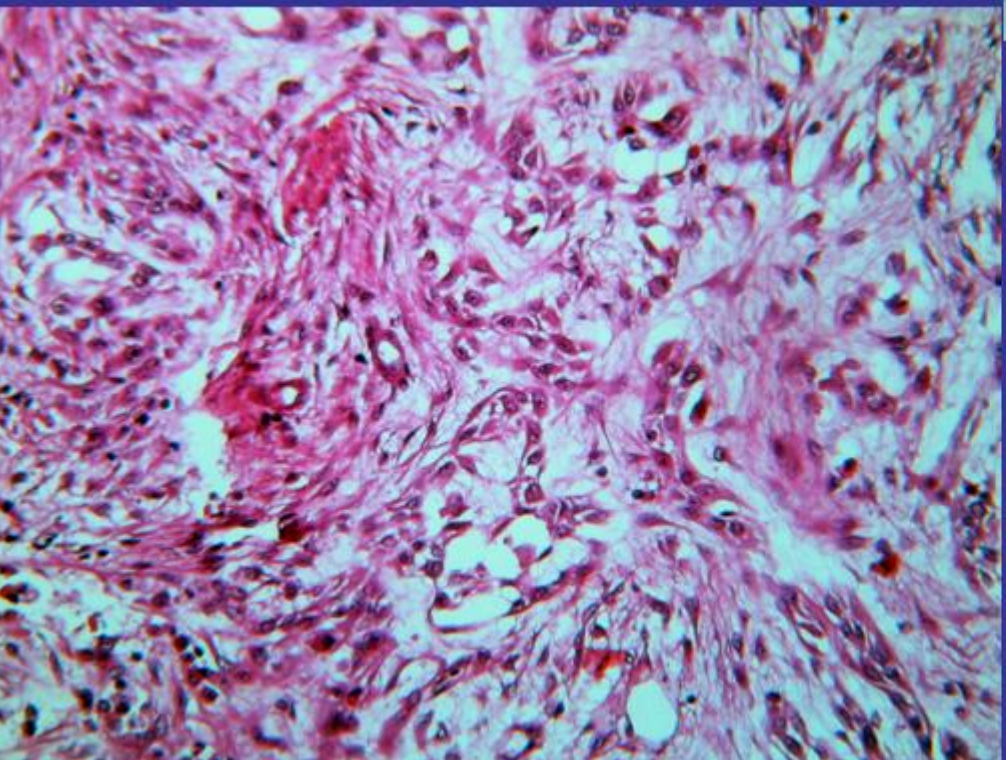
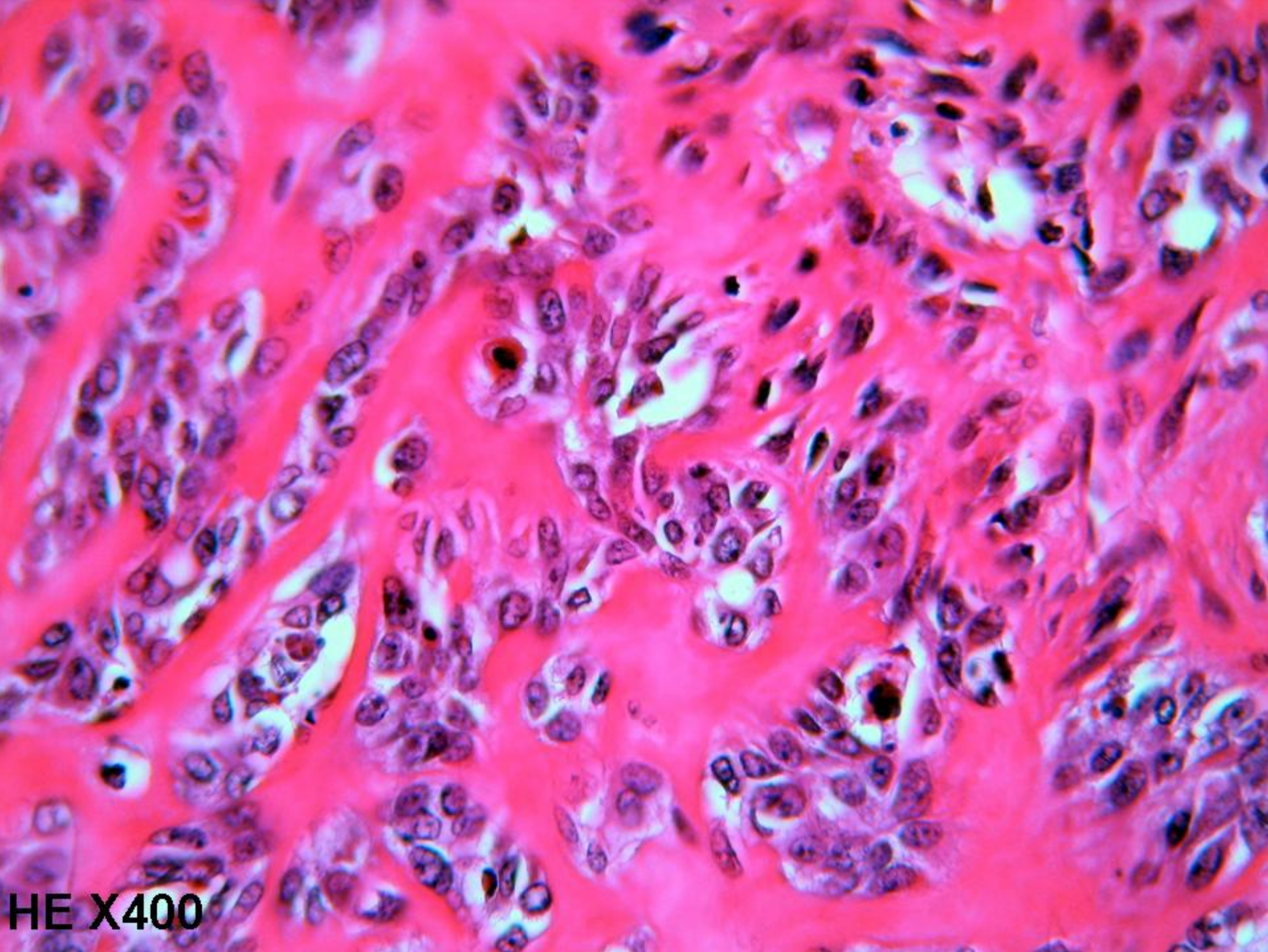

HE X400

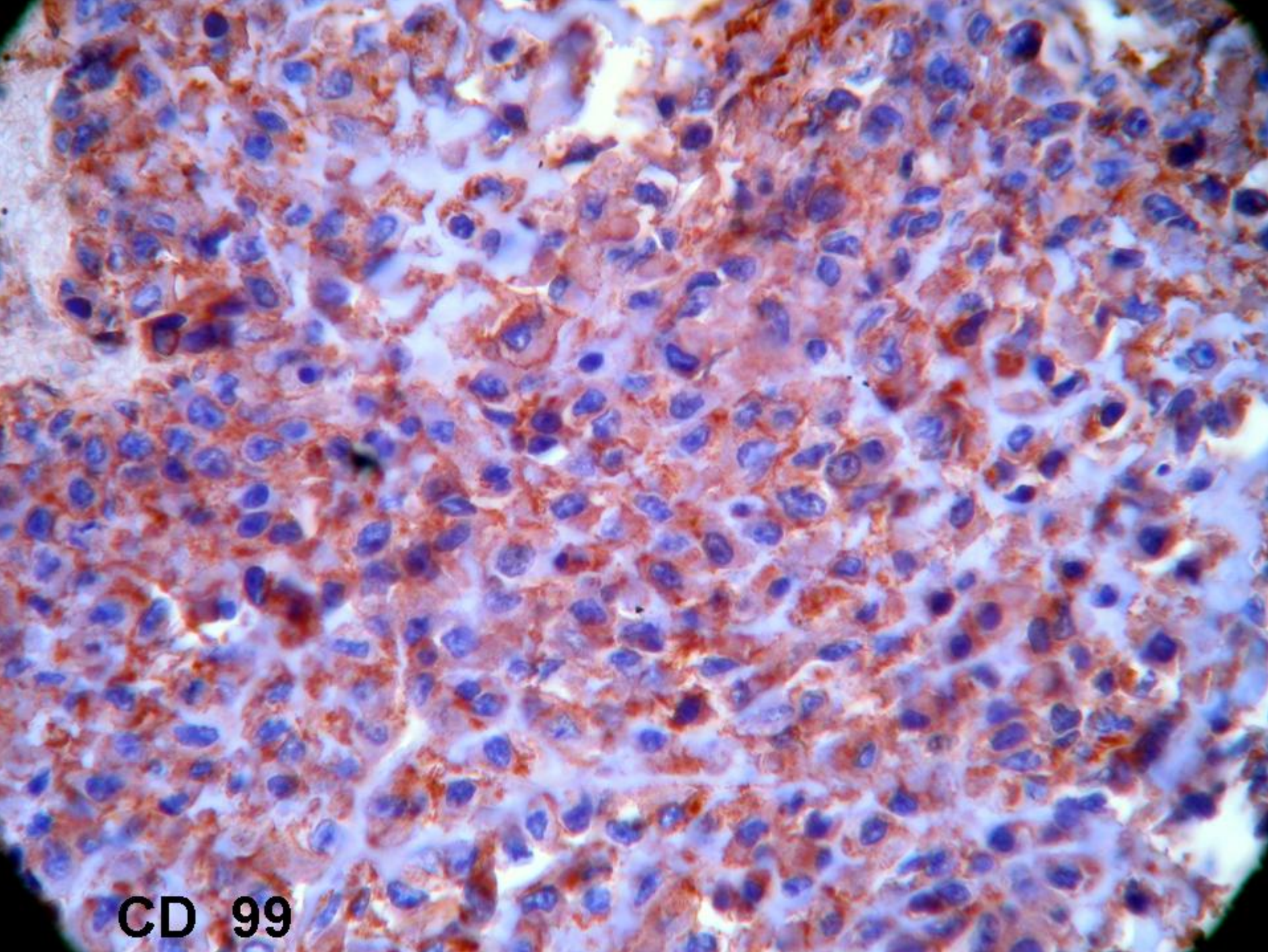


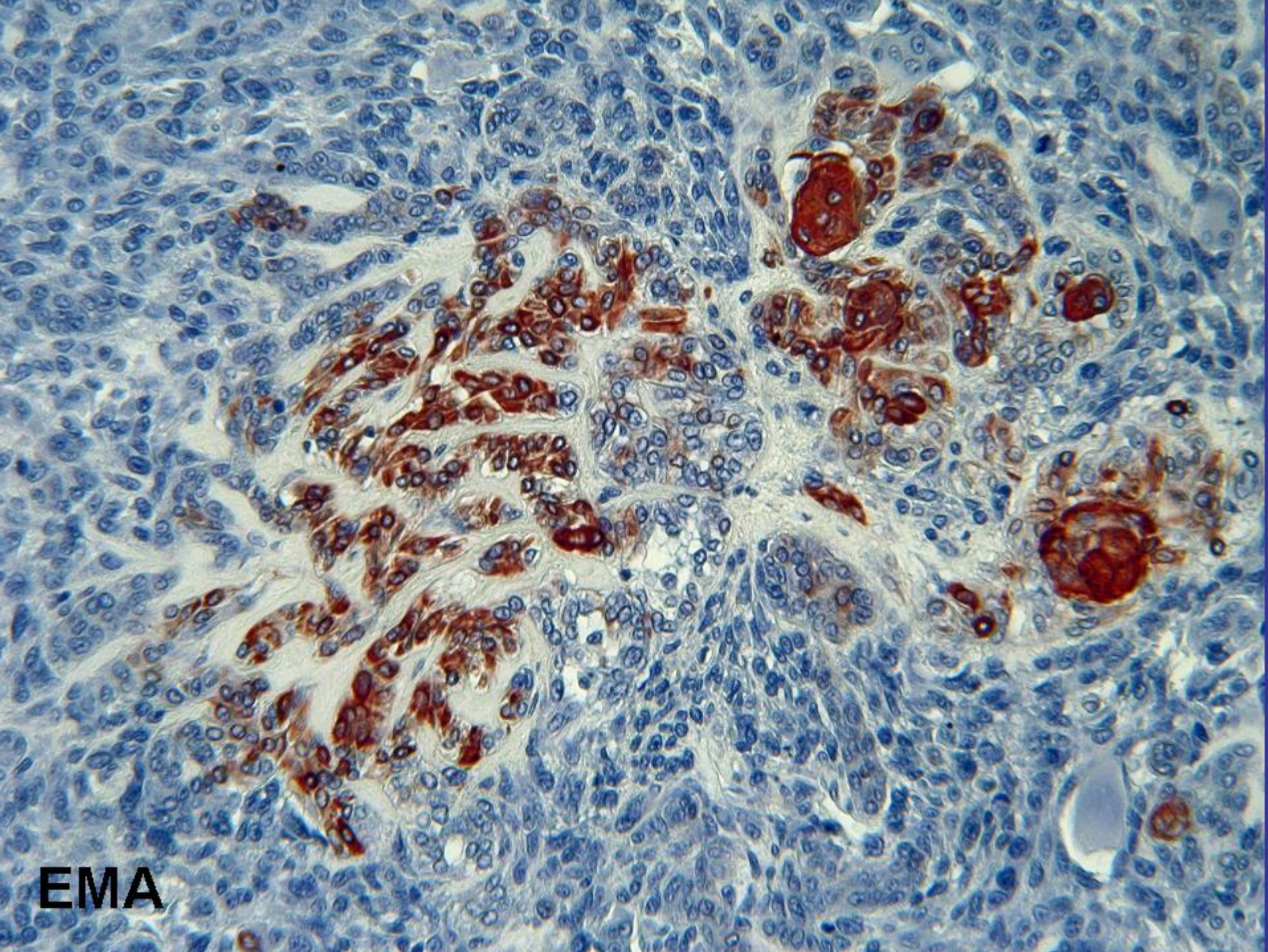


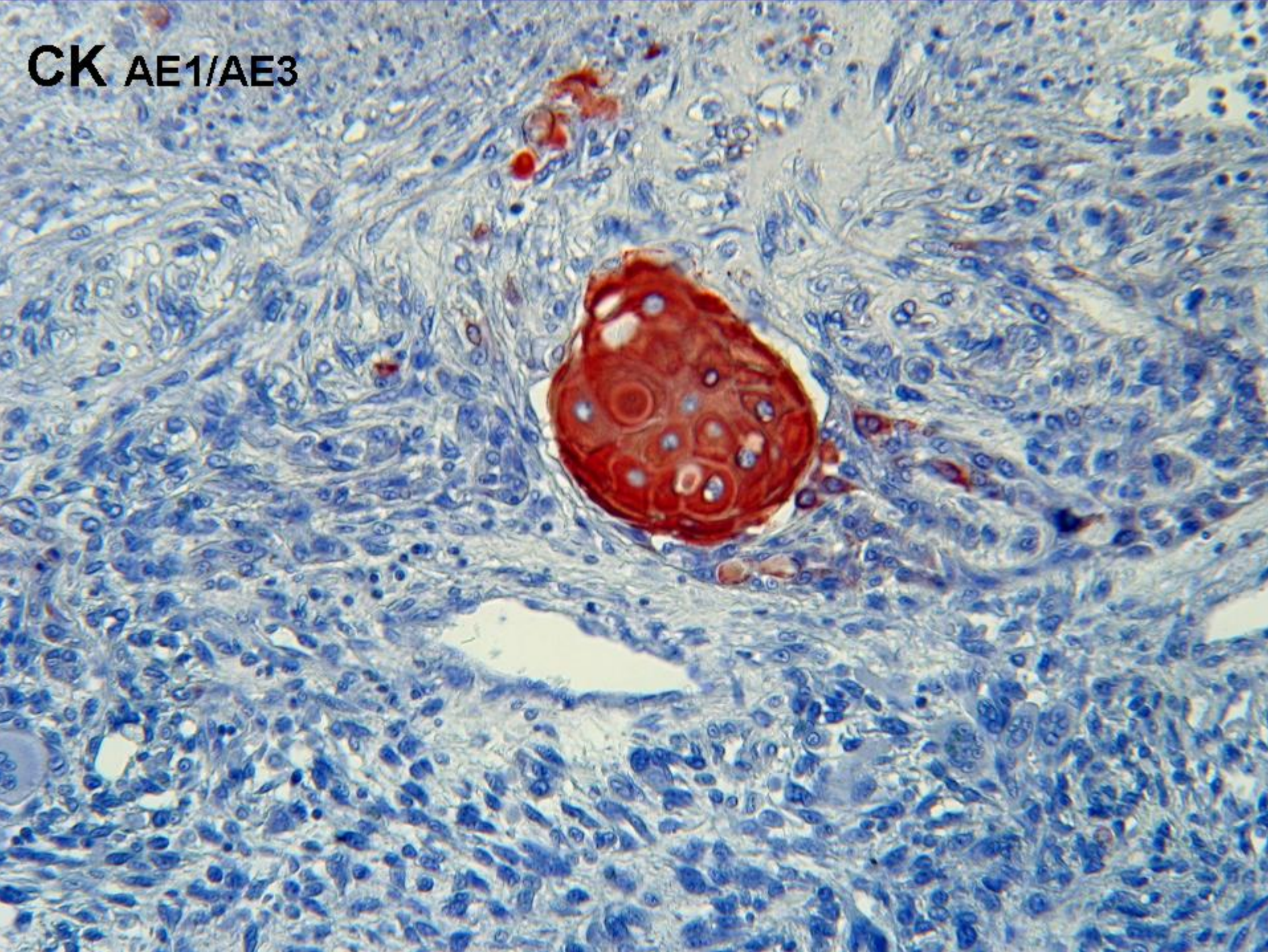

HE X400

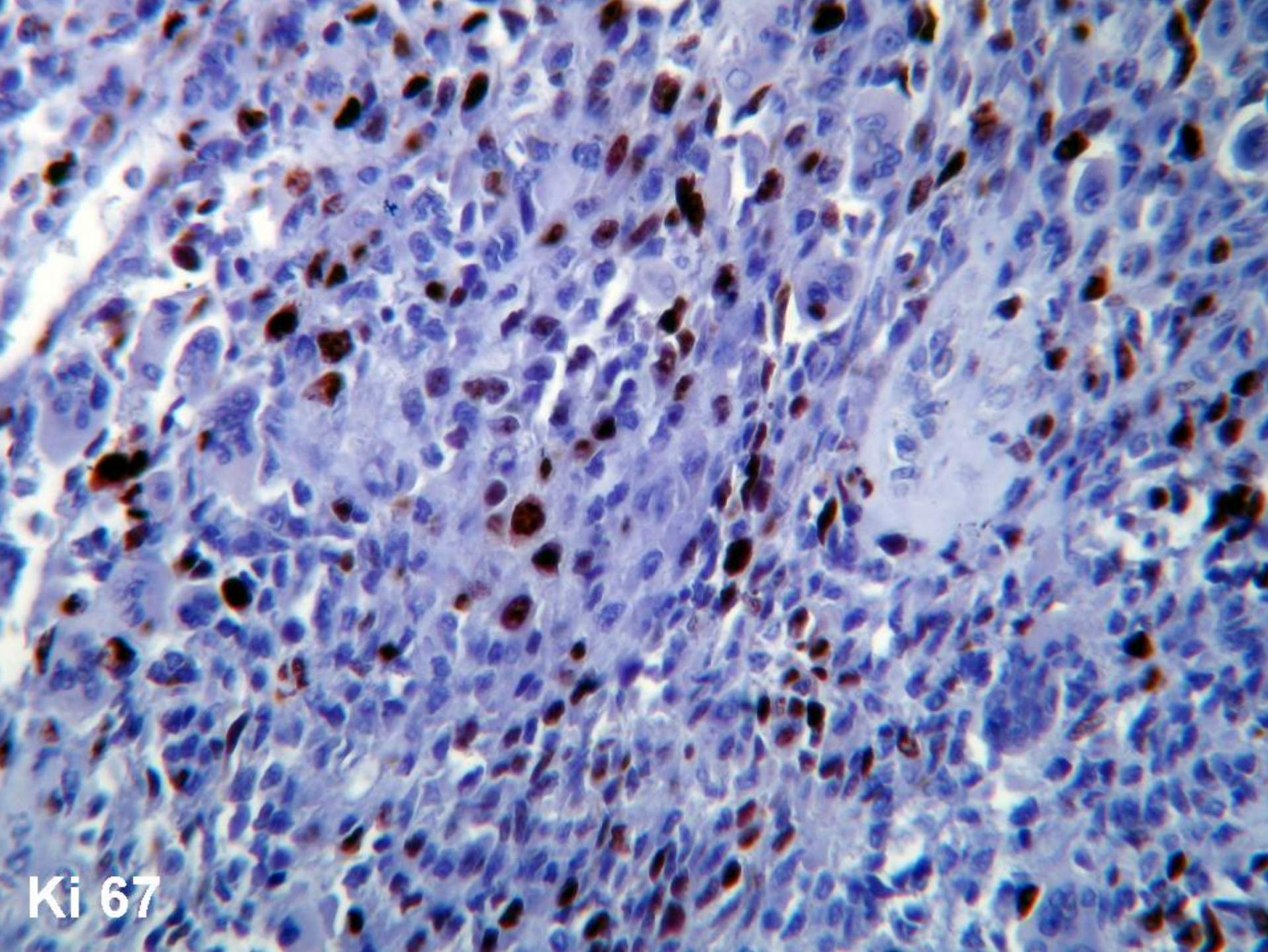
IHC:
Strong expression of :
CD99 ++
Epithelial structures : CK and EMA +

CD 34 -
Neoplastic cells present cytological irregularities and mitoses ( Ki 67 ↑↑↑)


CD 99


EMA


CK AE1/AE3


Ki 67

**Diagnosis :**

***« Giant cell » biphasic synovial sarcoma with massive osteoid and bone formation***

# Comments:

Synovial sarcoma is a well-defined entity which occurs predominantly in the limbs ans especially in the neighbourhood of joints. Arises from pluripotential mesenchymal cells capable of partial or aberrant epithelial differentaition

*Three variants are recognised :*

Biphasic synovial sarcoma, Monophasic synovial sarcoma, Poorly differenciated round cell synovial sarcoma

There are also rare forms, difficult to diagnose and which deserve to be known like :

Cystic forms

Calcifying forms

Forms with giant cells

Forms with myxoid change

Rare forms containing osteoid or bone formation **(can be confused with osteosarcoma)**

In 1952 and 1961, Wright CJ and Geiler G reported and described in detail cases of synovial sarcoma with giant cells.

In 1993, Milchrub S and al. reported 4 cases of synovial sarcoma with extensive calcifications, osteoid and bone formation simulating osteosarcoma.

# Major importance:

Immunohistochemistry

- CD 99 +
- bcl2 +
- CK and EMA +
- β-catenin +
- E-Cadherin +

- CD 34 Negative
- SMA, h Caldesmon, Desmin, CD117 Negative

## Specific chromosomal translocation:

t (X:18)(p11.2;q11.2) : more than 90% of cases

CD 99 expression
Negativity of CD 34 and
Epithelial component positivity for CK and EMA alows to distinguish synovial sarcoma with extensive osteoid and bone formation from osteosarcoma.

## Prognosis

5 year survival rate : 68%
10 year survival rate : 41%

## poor prognosis variables :

Tumour size > 6.7 cm
Poor differentiation ( subtype)
High nuclear atypia, mitoses > 27/10 hpf
Absence of stromal calcification
Nuclear expression of b-catenin and Ki-67 (MIB-1) index > 27%
High proliferation rate and poor prognosis are associated with the SYT-SSX1translocation14.

# References :


1 / Wright CJ: Malignant synovioma. J Pathol Bacteriol 64:585, 1952

2 / Geiler G: Die synovialome: morphologie und pathogenese. Berlin, springer-verlag, 1961

3 / Milchgrub S, Ghandur-Mnaymneh L, Dorfman HD, Albores-Saavedra. Synovial sarcoma with extensive osteoid and bone formation.

4 / Singler S, Baldini EH, Demetri GD, Fletcher JA, Corson JM. Synovial sarcoma: Prognostic significance of tumor size, margin of resection and mitotic activity for survival. J Clin oncol 1996; 14: 1201-1208.

5 / An immunohistochemical vade mecum. Dr Paul W Bishop, BA MB BCh FRCPath Consultant Histopathologist, Wythenshawe Hospital, South Manchester M23 9LT UK - version date May 2008