

Placental mesenchymal dysplasia

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Summary

- Normal placenta
- Definition of PMD (Placental mesenchymal dysplasia)
- Epidemiology of PMD
- Ultrasonography
- Gross examination of placenta with PMD
- Microscopic findings in PMD
- Differential diagnosis
- Take-home messages

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Normal term
placenta

- Discoid shape
- Diameter: 15-20cm
- Thickness : 1,5-3cm
- Weight : 450-600 grams
- Fleshy, spongy to feel
- The main components :
 - **Placental disk: fetal/maternal surface**
 - **Umbilical cord**
 - **Extraplacental free membranes**



Placental disk

- Fetal surface (chorionic plate):
 - Smooth
 - Transparent
 - covered by amnion with UC attached close to the center
 - umbilical vessels radiating from UC

Placental disk (2)

- Maternal surface:

- Irregular
- Divided into 15-20 reddish brown, convex areas (cotyledons), covered by shreds of decidua
- After birth, the placenta should be carefully inspected for **missing cotyledons**
- If cotyledons remain attached to the uterine wall – **severe bleeding**



Umbilical cord

- Length = 54-61 cm
 - two umbilical arteries (deoxygenated blood to the placenta)
 - one umbilical vein (oxygenated blood: placenta -> fetus)
- loose mesenchyme with intercellular ground substance (Wharton's jelly).
- covered by amniotic epithelium



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Placental
membranes

- **amnion** (the innermost lining of the amniotic cavity composed of a single layer of flat epithelial cells)
- **chorion** (connective tissue that carries the fetal vasculature)

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Villous parenchyma

The structure of the villus changes dramatically over the period of a normal gestation.

Immature 1st trimester villi are large and covered by two layers of trophoblast:

- an *inner layer of cytotrophoblast*
- an *outer layer of syncytiotrophoblast*.

The villous stroma is very loose and blood vessels are small and centrally placed.

Hofbauer cells (placental macrophages) are numerous.

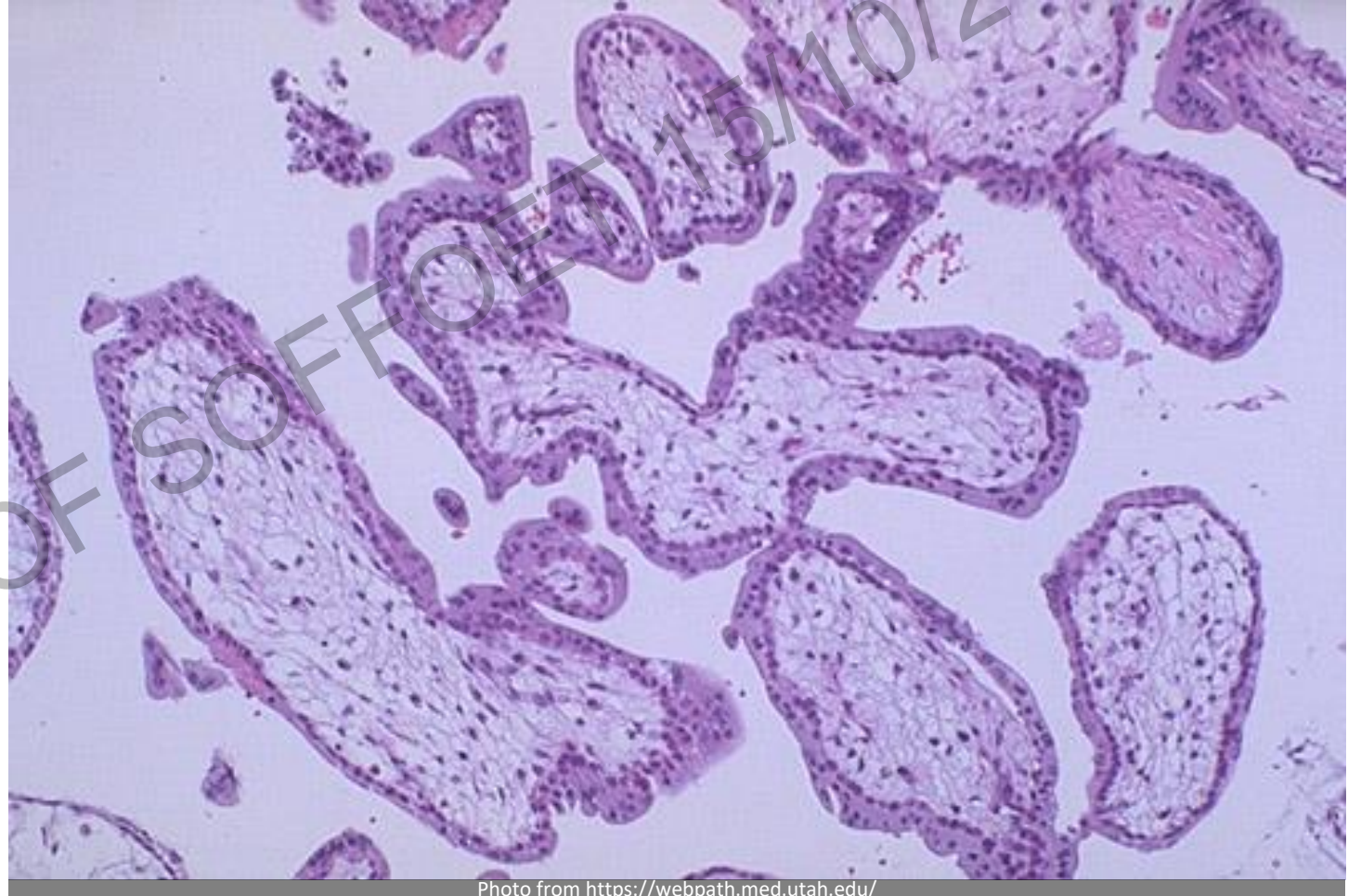


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Villous parenchyma (2)

2nd trimester villi:

- smaller
- the syncytiotrophoblast layer is thinner and the nuclei are less evenly dispersed
- the cytotrophoblast is discontinuous and difficult to identify
- the villous stroma is more compact and collagenized
- the capillaries are larger, more numerous and located peripherally.

Villous parenchyma (3)

2nd trimester villi:

- Clumps of **pink fibrin** begin to appear between the chorionic villi
- "**syncytial knots**" = clusters of syncytial nuclei – used to evaluate villous maturity (increase with gestational age)

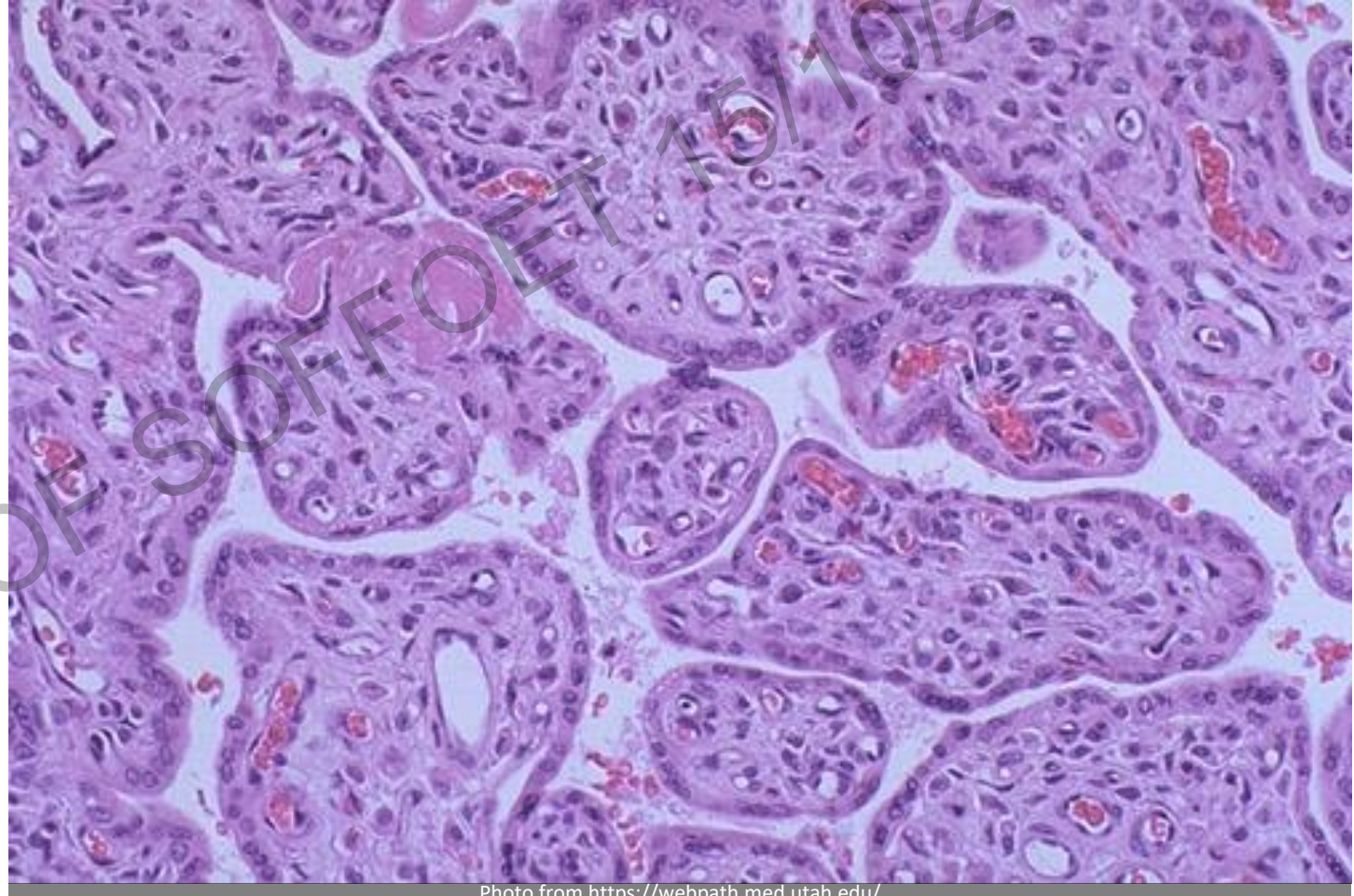


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Villous parenchyma (4)

3rd trimester villi:

- Even smaller
- **proeminent syncytial knots** and **intervillous fibrin**
- reduced stroma - thin strands compressed between dilated fetal capillaries
- The capillaries fuse with the thinned syncytiotrophoblast layer = *vasculosyncytial membranes* (maternal- fetal circulation exchange)

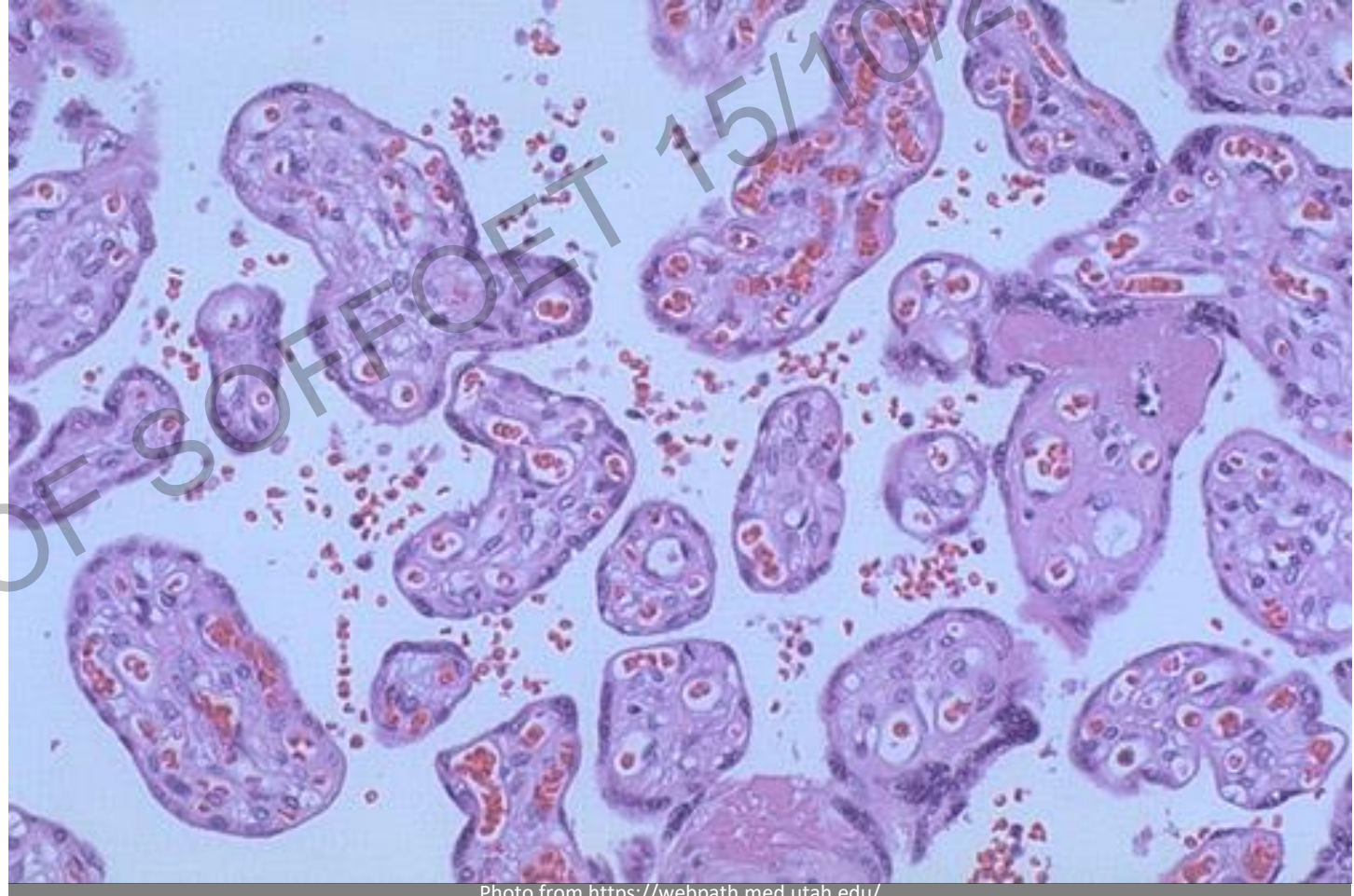


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Placental mesenchymal dysplasia (PMD)



= rare benign vascular malformation characterized by placentomegaly with grape-like fluid-filled vesicles resembling molar tissue at gross placental examination

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Epidemiology

- unknown incidence: estimated to 0.02%
- first described by Moscoso in 1991 as **stem villous hyperplasia** with elevated maternal serum alpha-fetoprotein and enlarged placentas with ultrasound features that suggest a partial mole.
- Slightly elevated serum titers of beta-hCG have also been reported

PMD may coexist with a normal fetus or may be associated with **Beckwith-Weidemann syndrome (BWS)** (1/3 cases)

BWS syndrome:

- chromosome 11 abnormalities
- large for gestational age fetus
- enlargement of internal organs (organomegaly)
- abdominal wall defects (umbilical hernia, omphalocele or diastasis recti)
- macroglossia
- distinctive grooves in ear lobes
- facial abnormalities
- increased risk of developing childhood cancers: Wilms tumor, hepatoblastoma or neuroblastoma

Ultrasonography

- ✓ Enlarged uterus
- ✓ Thickened placenta with anechoic regions

- ≠ complete mole
- ≠ partial molar pregnancy
- ≠ chorioangioma

Placental mesenchymal dysplasia

- hypoechoic/multicystic areas
- with normal areas of placenta

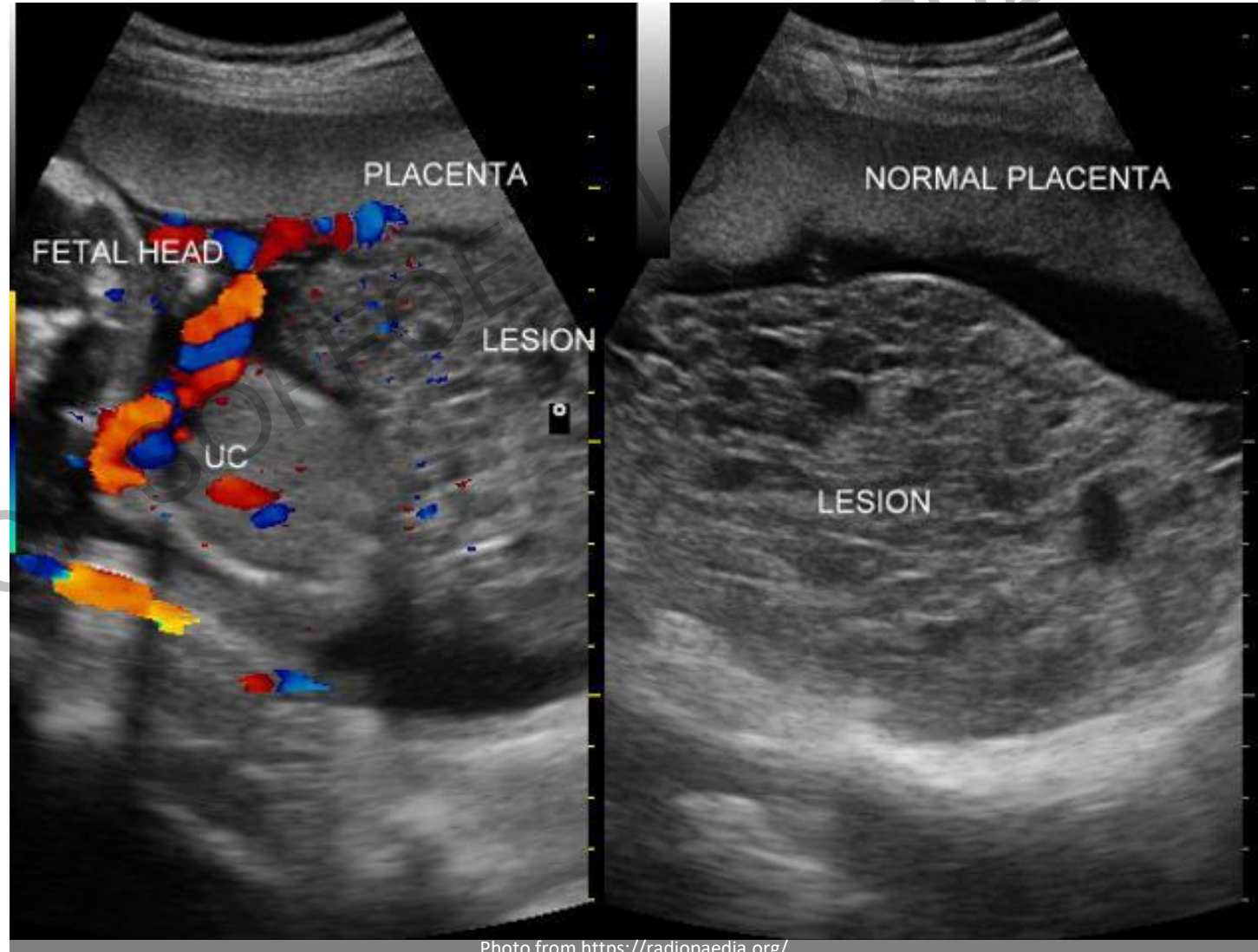


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Complete/Partial mole

- intrauterine mass with cystic spaces with/without fetal components
- "snowstorm" or "bunch of grapes" appearance

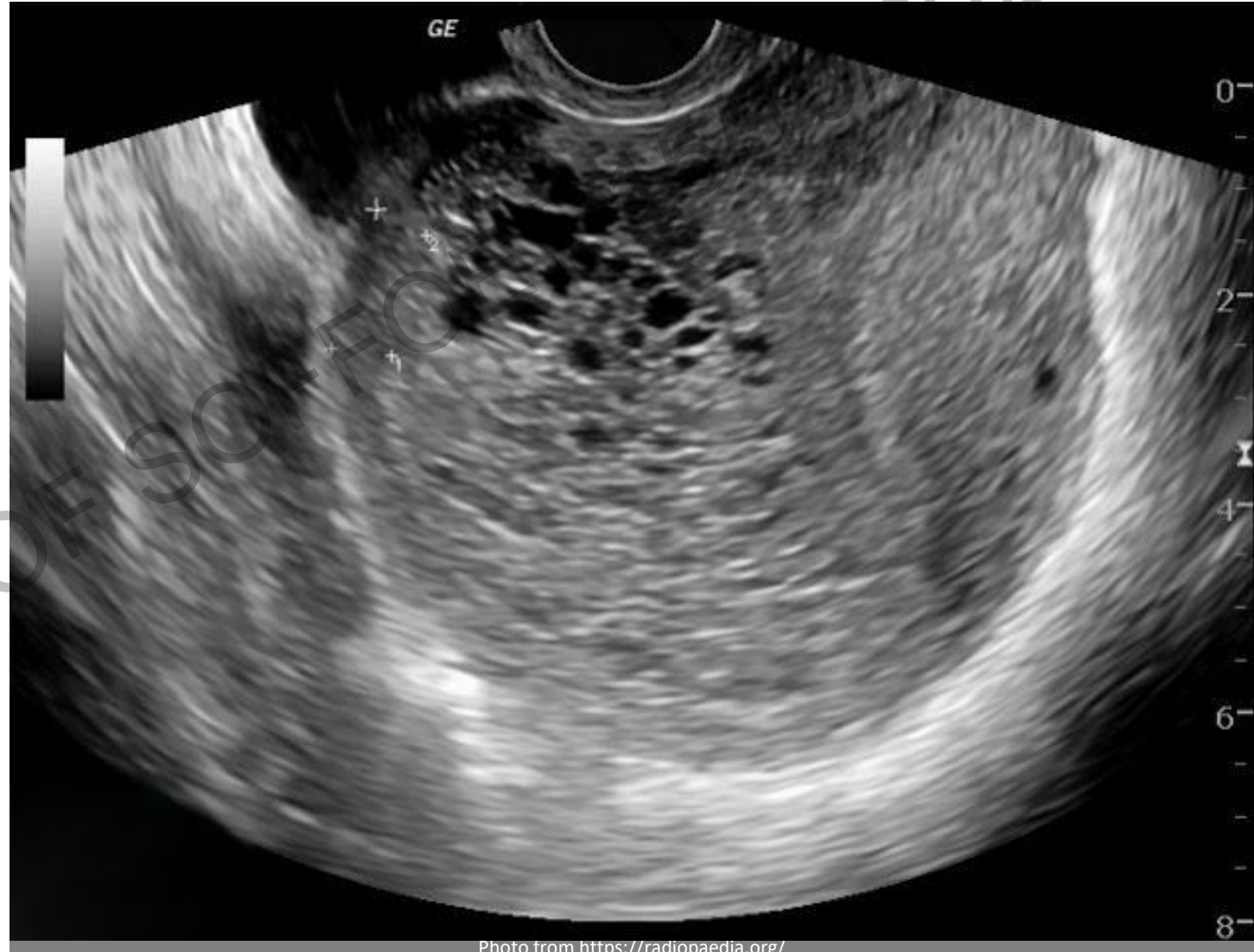
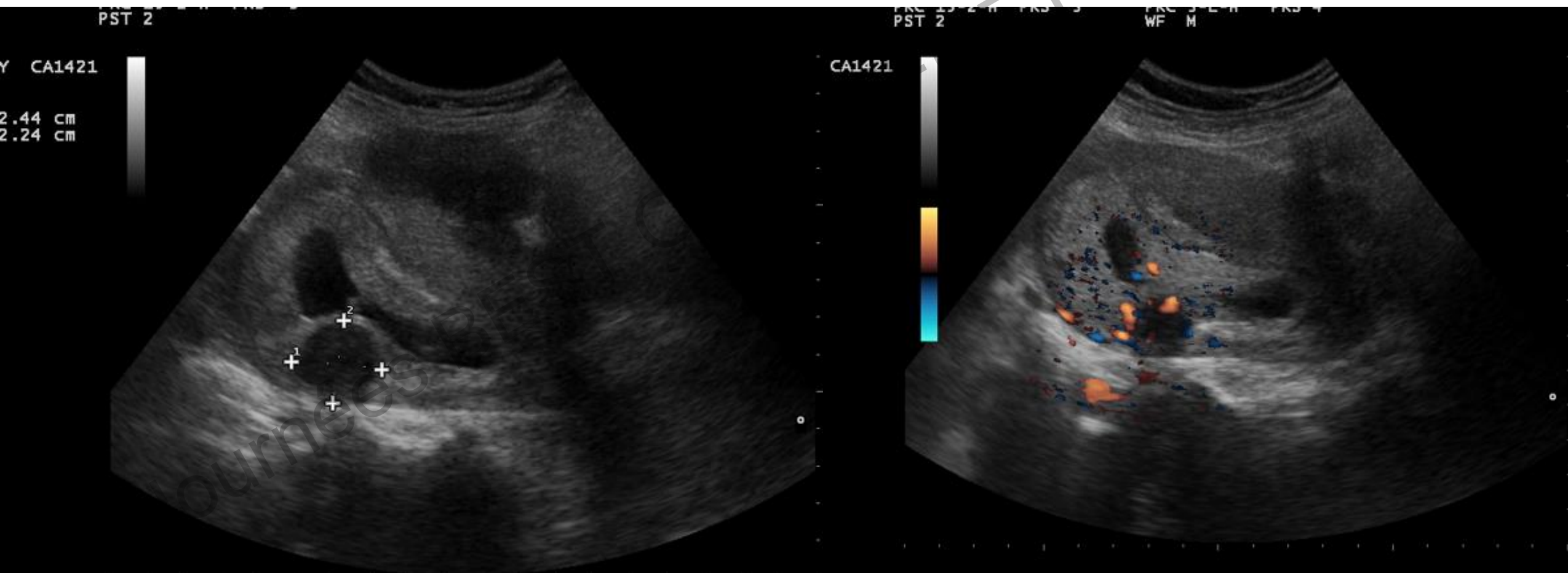


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Chorangioma

A well circumscribed hypoechoic mass at the fetal surface of the placenta, adjacent to the cord insertion, bulging into the amniotic cavity. The mass has internal vascularity and adjacent feeding vessels.



Case presentation

32 years old woman (27 weeks pregnant) known with IUGR presented to the hospital for painful uterine contractions.

She delivered a 550g non-viable fetus.

Gross examination of the placenta

- markedly enlarged placenta 730g
- **maternal surface:** “bed of yarn” appearance
- **chorionic plate:** multiple tortuous vessels, some of them thrombosed
- **the cut surface:** numerous cystically dilated vesicles, ranging from 0.1 to 5 cm, filled with serous fluid

Macroscopic appearance of PMD:

- enlarged placenta: 10/9/4 cm
- 645 grams
- grape-like vesicles max \varnothing = 5 cm
- UC : 57 cm length, 2A+1V
- fetal membranes are translucent, slightly thickened.



- **Maternal surface :**
numerous small vesicles



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▪ **On the section surface:**

multiple vesicles with
serous fluid and tortuous vessels



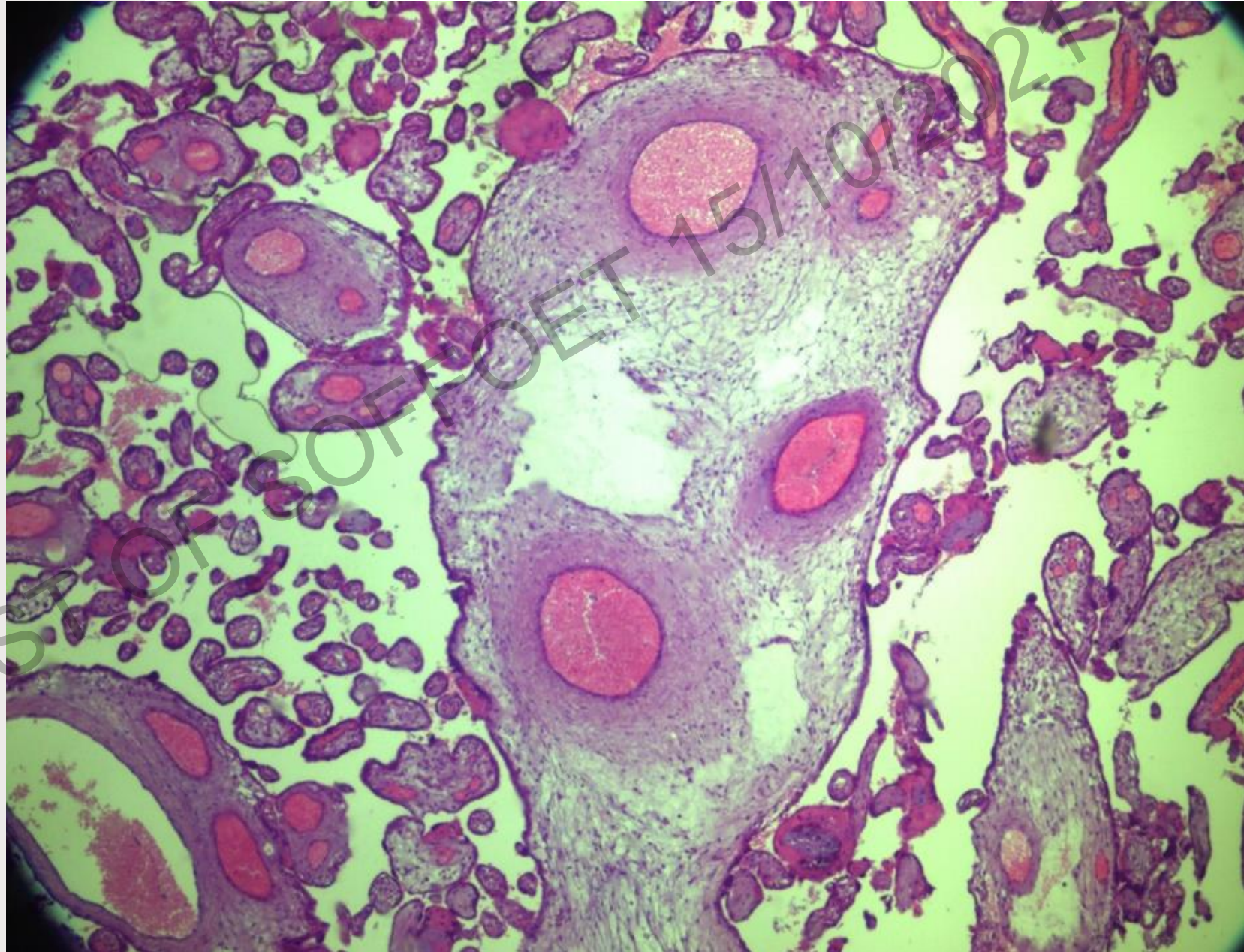
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Histopathology findings:

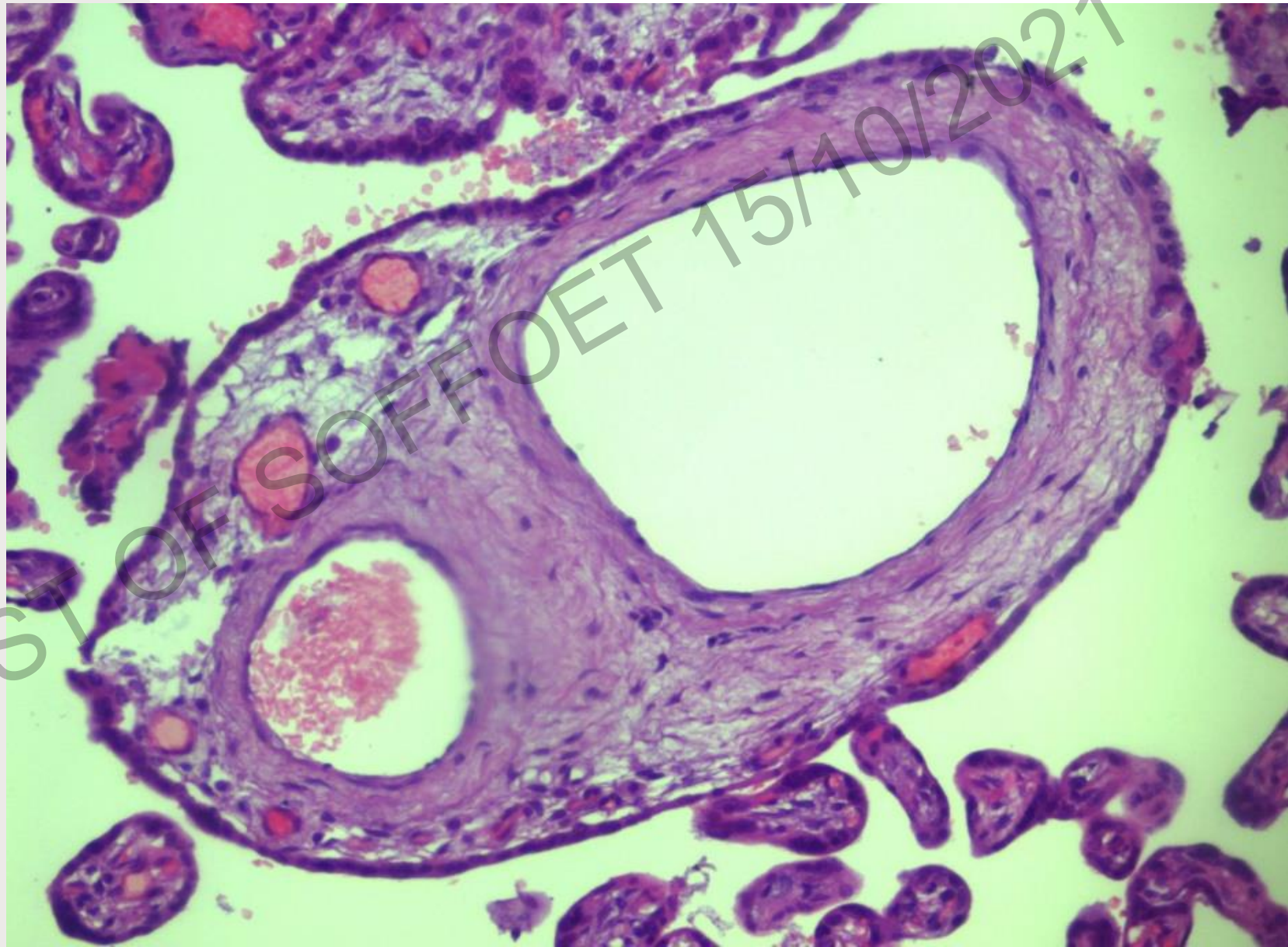
- ❖ **markedly edematous villi** interspersed with normal sized villi
- ❖ **cistern-like formation** (broken strands of fibrillar material)
- ❖ **loose myxoid stroma** rich in hyaluronic acid
- ❖ **absence of trophoblastic proliferation** or trophoblastic inclusions
- ❖ **hydropic changes: stem villi** (mesenchymal dysplasia) vs terminal villi are (partial moles).

- Heterogeneity in villous size
- hydropic stroma
- dilated thick vessels
- cistern-like formation



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- Huge villi with different sized vessels, covered by a layer of trophoblasts



Differential diagnosis

➤ **Hydatidiform mole: partial/complete**

- Benign neoplasm of chorionic villi
- Macroscopic appearance similar to PMD : grape-like vesicles of different sizes
- Trophoblastic proliferation (both syncytial and cytotrophoblastic cells) with mitotic activity

Differential diagnosis(2)

- Complete mole: the whole conceptus is transformed into vesicles
 - ❑ no embryo present
 - ❑ caused by fertilization of an anucleated ovum (without chromosomes) with spermatozoid which duplicate – 46cz (paternal origin only)
 - ❑ Increases the risk of choriocarcinoma



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Differential diagnosis(3)

- Partial mole: only a part of trophoblast is affected by hydropic changes
 - ❑ usually, viable fetus
 - ❑ caused by fertilization of an ovum by 2 spermatozoids - >69cz



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Differential diagnosis (4)

➤ Chorangioma

☐ = non-neoplastic condition

☐ Hamartoma consisting of blood vessels

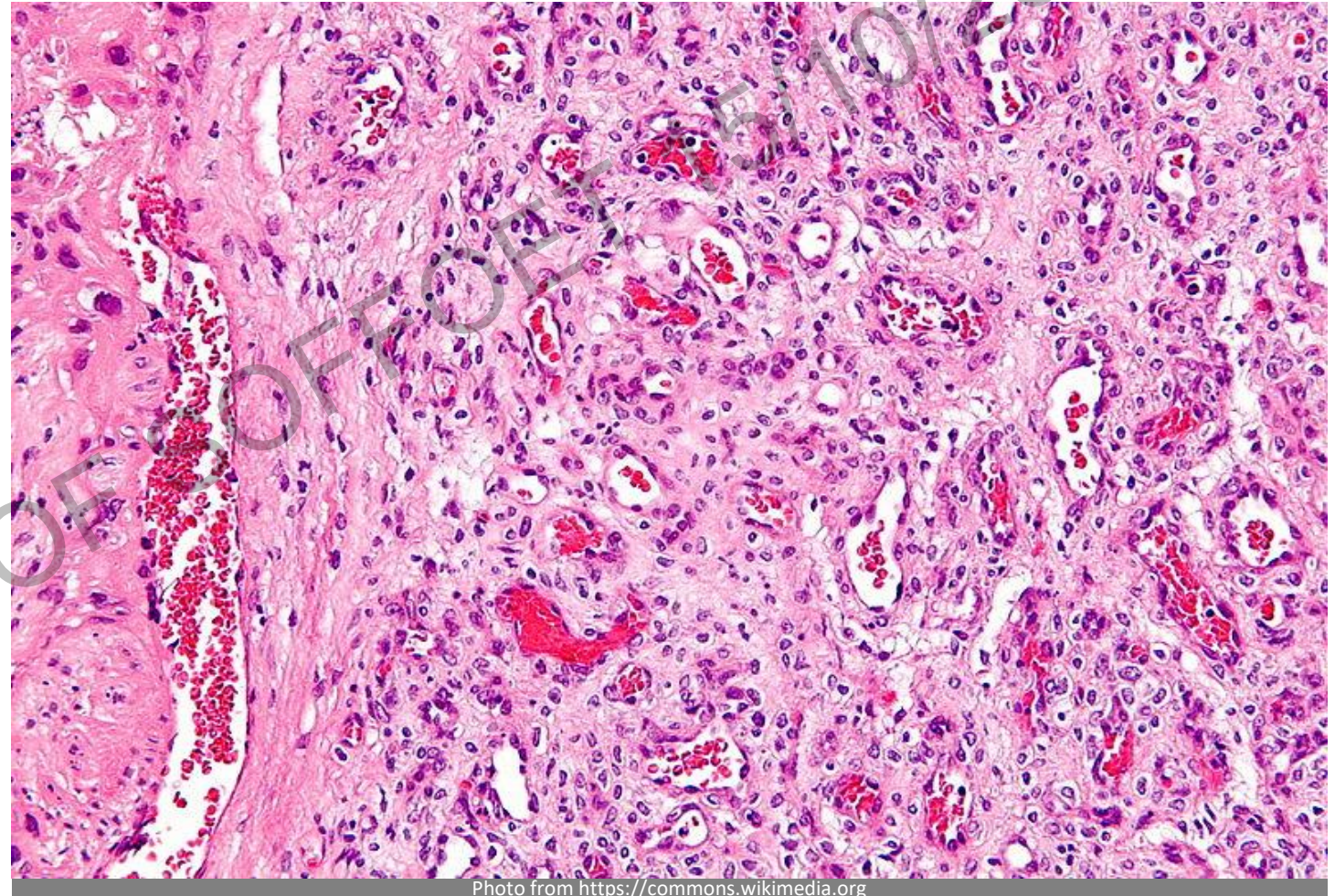


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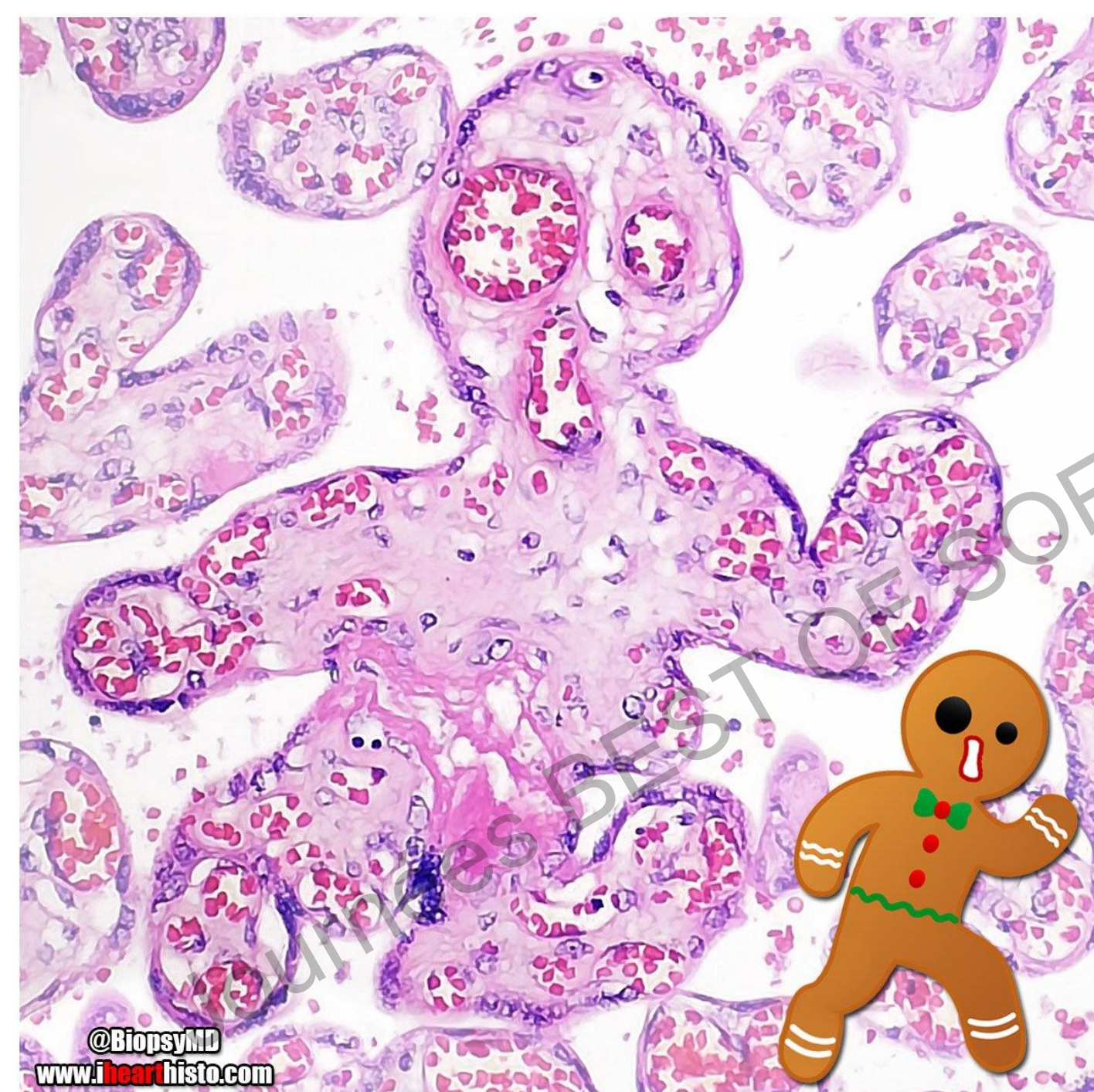
Take-home messages:

1. PMD is a benign vascular disorder of placenta
2. Clinically mistakenly as molar pregnancy or chorangioma
3. Placentomegaly with grape-like vesicles
4. Absence of trophoblastic proliferation
5. Close attention to fetal morphology for early recognition of fetal complications

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