Case Report

When a Chorangioma Becomes a Burden in Fetal Survival: A Reported Case with an Updated Literature Review

DIANA BURLACU[1,2], AGNES BURLACU[1], RÉKA BELÉNYESSY[3], BÉLA SZABÓ[1,4], TIBOR MEZEI[1,2]

[1]*George Emil Palade University of Medicine, Pharmacy, Science, and Technology of Targu Mures, Romania*

[2]*Targu-Mures Emergency Clinical Hospital, Department of Pathology*

[3]*City Hospital of Odorheiu Secuiesc, Romania*

[4]*Targu-Mures Emergency Clinical Hospital, Obstetrics and Gynecology Clinic*

ABSTRACT: Chorangioma is a rare non-trophoblastic benign vascular neoplasm originating from the primitive chorionic mesenchyme. Usually asymptomatic, it affects approximately 1% of female fetuses. We present the case of a giant placental chorangioma (GPC) in a preterm male pregnancy coexisting with a maternal neuroendocrine carcinoma. A 30-week primigravida was admitted to the Obstetrics and Gynecology Clinic of the Targu-Mures Emergency Clinical Hospital, with abdominal discomfort, and an emergency C-section was performed for fetal congestive heart failure. Medical history revealed an advanced-stage rectal neuroendocrine carcinoma. At 20th gestational week, a well-vascularized placental mass was diagnosed. A 1500g premature male fetus was delivered. Histopathologically, the placental mass revealed an unencapsulated but well-circumscribed tumor with lobular architecture composed of congested vascular capillaries and thin-walled vessels. Diagnosis of giant placental chorangioma (GPC) was rendered. GPC is a challenging condition typically occurring in hypertensive or diabetic primigravidas with female fetuses. Antenatal management is suggested at an early stage for a desirable perinatal outcome.

KEYWORDS: *Chorangioma, placenta, benign, vascular, prematurity.*

Introduction

Chorangioma is a relatively rare, non-trophoblastic, indolent vascular neoplasm originating from chorionic tissue.

With an incidence of approximately 1% of all pregnancies, the origin of this benign vascular tumor is believed to be the primitive chorionic mesenchyme [1].

Most chorangiomas are associated with advanced maternal age, hypertensive or diabetic status of the mother, and pregnancy with a female fetus [2,4].

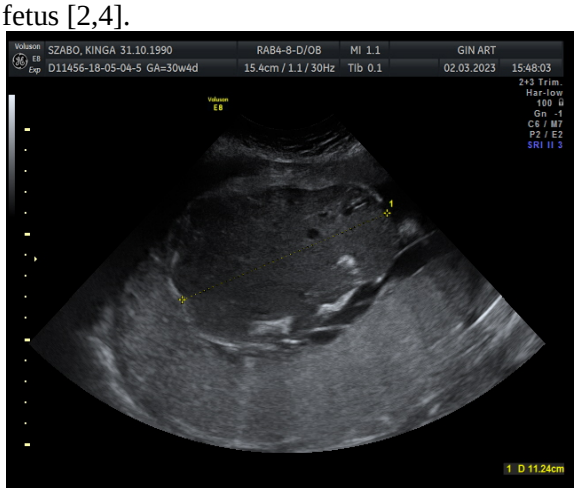

Figure 1. Ultrasound imaging of the tumor taken a few weeks before birth.

Case Presentation

We present the case of a 32-year-old primigravida with regular check-ups.

At 20 weeks of pregnancy a routinely performed ultrasonography revealed a well-defined mass on the placental surface having a slightly different echogenicity from the rest of the placenta.

It was located near the umbilical cord, and Doppler imaging showed rich vascularity (Figures 1 and 2).

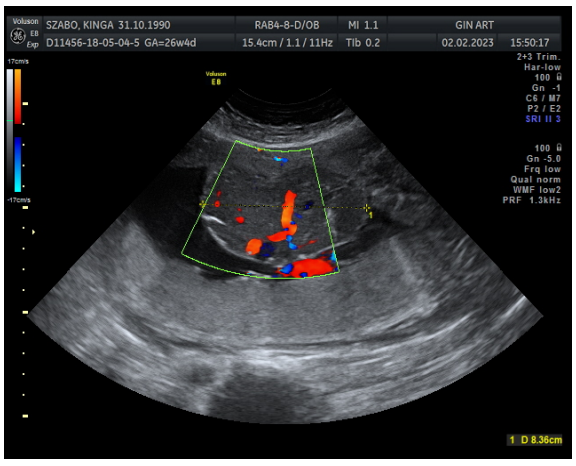

Figure 2. Doppler imaging highlights a relatively rich vascularity.

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A suspicion of chorangioma with insignificant size was raised.

The patient was sent for an MRI scan which confirmed the suspicion of chorangioma.

Knowing that placental chorangioma can cause fetal anemia and thrombocytopenia, the pregnancy was closely supervised by weekly-biweekly ultrasound exams monitoring for eventual Doppler signs of fetal anemia.

By the time fetal anemia was suspected by middle cerebral artery Doppler, ultrasound scans were performed two times per week to schedule the right time of the delivery.

A slight increase in tumor size was noted with signs of developing fetal hydrops.

Fetal cardiomegaly soon appeared, followed by tricuspidal insufficiency and pericardial effusion.

At 30 gestational weeks, she was admitted to Targu-Mures Emergency Clinical Hospital in the Obstetrics and Gynecology Clinic complaining of abdominal discomfort and for further investigations.

Her medical record showed no signs of hypertension or diabetes.

In 2014 she was diagnosed with an advanced-stage rectal neuroendocrine carcinoma with partially resected liver metastasis.

Ultrasound examination described a fetus with biometrics corresponding to its gestational age, of apparently normal morphology, except for cardiomegaly, as mentioned previously.

Fetal echocardiography showed a small discontinuity of the muscular interventricular septum, a small insufficiency of the tricuspid valve, respectively an insufficiency of the mitral valve.

The placenta had posterior insertion at the upper pole.

An emergency C-section was performed, and a 1500g premature male fetus with congestive heart failure and generalized edema was delivered.

The evolution of the newborn was unfavorable shortly after delivery, with progressively altered general condition.

Both severe anemia and thrombocytopenia, with active bleeding at the level of oral and gastric cavities, required repeated transfusions of erythrocyte mass and fresh frozen plasma without obtaining a stable hemodynamic state.

Echocardiographic findings suggested heart failure with severe pulmonary hypertension and pericardial collection located predominantly anterior to the right cavities, without signs of cardiac tamponade.

The premature infant also showed cerebral intraventricular and intraparenchymal hemorrhage, right temporal-parietal-occipital cerebral hemorrhagic infarction with mass effect on the midline structures.

The shortly-developed renal failure with persistent anuria was refractory to treatment.

The comatose state that subsequently ensued with a lack of spontaneous breathing required oro-tracheal intubation.

In the context of massive hemorrhagic syndrome, with pulmonary, digestive, and upper airway bleeding, the patient presented numerous episodes of hemodynamic instability with severe bradycardia and desaturation.

Ultimately leading to a cardio-pulmonary arrest and demise.

The autopsy revealed morphological changes consistent with multiple organ failure, including diffuse alveolar damage (DAD), pulmonary edema with focal alveolar hemorrhage, right temporo-parietal cerebral and right intraparenchymal cerebellar hematomas.

Histological analysis of the placenta revealed the presence of a large tumor detached from the placenta.

The placenta measured 16 x 14 x 2.5cm and weighed 577g without any significant gross changes, except for a large indentation near the umbilical cord insertion.

The tumor, which was submitted within the same recipient, but was not directly connected to the placenta, had a tan color with a fleshy consistency measuring 13.5 x 11.5 x 6.5cm, and weighing 412g.

It showed a glistening and smooth surface (Figure 3).

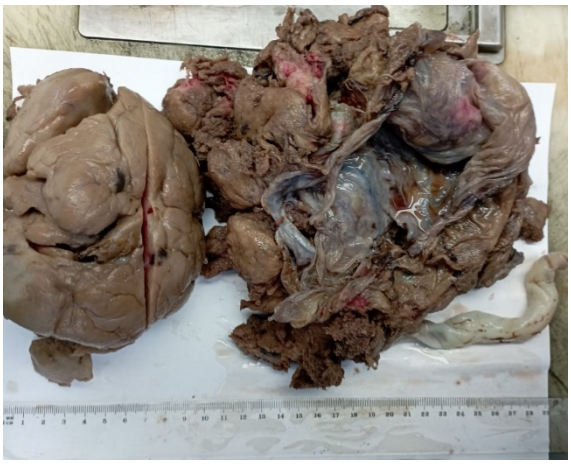

Figure 3. Gross appearance of the placenta, membranes and umbilical cord (right) and the tumor (left).

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Microscopically an unencapsulated but well-circumscribed tissue proliferation was noted with a lobular architecture, mostly composed of congested and thin-walled vascular structures.

The size of these ranged from capillary-sized vessels to small-sized arteries (Figures 4 and 5).

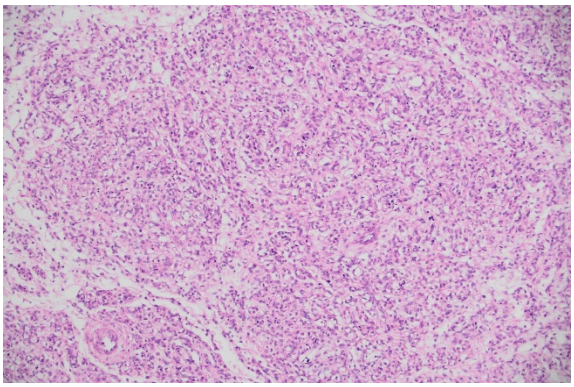

Figure 4. Histological aspect of the tumor on low magnification. A network of capillaries of variable size is seen (H&E stain, 4x objective).

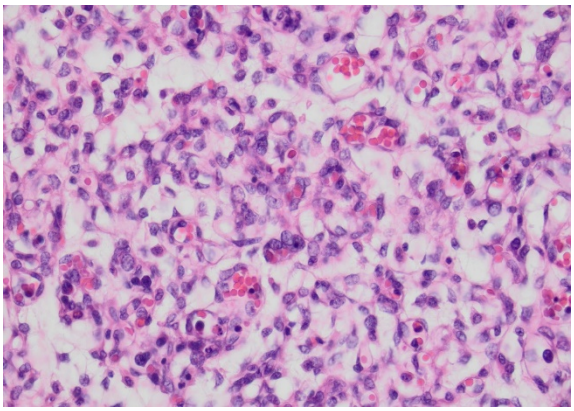

Figure 5. Histological aspect of the tumor on high magnification. Multiple capillary sized vascular channels (H&E, 40x objective).

Occasionally cellular pattern was also noted. Immunohistochemistry revealed CD31-positive endothelial cells lining these vascular channels (Figure 6).

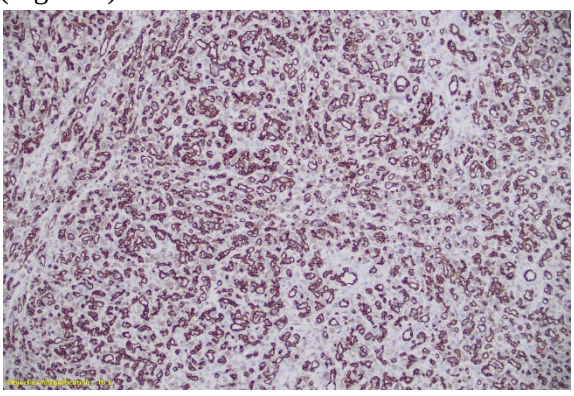

Figure 6. CD31-positive endothelial cells lining these vascular channels (Immunohistochemistry, DAB stain, 10x objective).

Cytokeratin 18 positivity was also noted, suggesting a chorionic plate and anchoring villi origin (Figure 7).

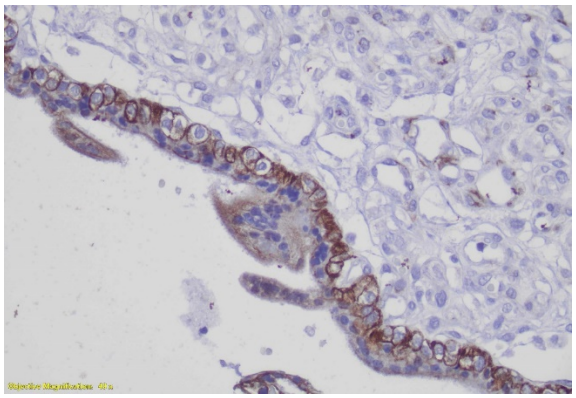

Figure 7. CK 18-positive chorionic plate and anchoring villi (Immunohistochemistry, DAB stain, 40x objective).

No mitotic figures, no significant cytological atypia were encountered.

Based on gross and microscopic morphology, a diagnosis of giant placental chorangioma was rendered.

A written informed consent was obtained from the mother before considering these data for publication.

Discussion

Historical background and clinical diagnosis

Ultrasound examination of abdominal organs was first reported in 1958.

The first chorangioma case ever reported was by Clarke in 1978 [1,2].

Coincidence or not, antenatal ultrasound (US) was also reported for the first time in 1978, thus facilitating the diagnosis and follow-up of pregnant women until delivery and beyond [3].

Furthermore, Bromley and Benacerraff were the first to use Doppler imaging to differentiate a chorangioma by means of ultrasound from a placental teratoma, leiomyoma, or even a blood clot.

All these lesions have similar clinical signs without proper imaging techniques [4].

The arterio-venous shunt characteristic of chorangioma causes low resistance flow on Doppler imaging, hence the characteristic US appearance.

Also, vascular channels of the chorangioma are similar to fetal ones and tend to retain constant morphology, while the US pattern of the blood clot changes over time [5].

Chorangioma is a relatively rare, non-trophoblastic, indolent vascular neoplasm originating from chorionic tissue.

With an incidence of approximately 1% of all pregnancies, the origin of this benign vascular tumor is believed to be the primitive chorionic mesenchyme [6-8].

Most chorangiomas are associated with advanced maternal age, hypertensive or diabetic status of the mother, and pregnancy with a female fetus [9-13].

Our reported case was a young mother without documented hypertension or diabetes, and also pregnant with a male fetus.

Morphological characteristic and immunophenotype

These benign tumors are usually described on the fetal surface of the placenta, in the vicinity of the umbilical cord insertion [14].

Macroscopically, these benign tumors are well-circumscribed, with a congested fleshy cut surface [11,15,16].

Histologically, there are 3 types of chorangiomas described: angiomatous, cellular and degenerate [1].

Of these, the most common is the angiomatous one, characterized by numerous vascular channels lined by endothelial cells, capillaries and blood vessels surrounded by placental stroma [6].

In our case the predominant architecture was angiomatous with a minor cellular component. Since no malignant behavior was reported, some consider them as placental hamartomas, rather than real tumors [1].

Immunohistochemical analysis usually reveals variable reactivity to CD31 of endothelial cells lining the blood vessels.

Additionally, Cytokeratin 18 positivity may suggest the origin of the tumor cells from blood vessels of the chorionic plate and anchoring villi [17,18].

Clinical significance of this tumor is rather related to its size.

Small chorangiomas are usually asymptomatic.

On the other hand, larger chorangiomas may cause serious feto-maternal complications and the possibility of an adverse perinatal outcome.

Chorangiomas greater than 5cm are termed ‘giant’ and careful clinical examination and followup is warranted in such cases.

Giant placental chorangiomas are rare, with a reported frequency ranging from 1:9.000 to 1:50.000 of delivered placentas [9].

Pathophysiology

The exact pathophysiological mechanisms that lead to fetal complications are yet to be determined.

Still, fetal congestive heart failure may develop due to increased blood flow through the vascular channels of chorioangioma acting as an arterio-venous shunt.

Thus, the tumor is a significant burden for the fetal circulatory system, eventually leading to feto-maternal complications, as mentioned above [6,9].

What is of great curiosity is that in our case, the chorangioma acted somewhat analogous to a receptor twin in a twin-to-twin transfusion syndrome.

The tumor, acting as the receptor, due to its arterio-venous shunting, led to fetal high-output heart failure.

Furthermore, fetal platelet sequestration determined hemorrhages and thrombocytopenia, resulting in fetal anemia.

The larger the chorangioma, the more the venous return to the heart is, thus causing tachycardia and, eventually, cardiomegaly [19].

Consequently, as seen in our case report, generalized edema, congestive cardiac failure, and intrauterine growth retardation may be seen in a neonate.

Hence, polyhydramnios, premature delivery, preeclampsia, fetal growth restriction, fetal distress, fetal anemia, cardiomegaly, non-immune hydrops, thrombocytopenia, consumptive coagulopathy, and stillbirth are all possible complications [13,18].

Of all the clinical complications mentioned, the correlation between chorangioma and polyhydramnios in a preterm pregnancy seems the strongest, underlining its significance [7,20,21].

Despite this, polyhydramnios was absent in our case.

Still, other severe complications finally led to the death of the infant.

Chorangioma association with maternal tumors

It is known that maternal malignant tumors may produce placental and fetal metastases [28].

The association between maternal neuroendocrine tumor and chorangioma, however, is less well documented, and, in fact, we did not find any publication documenting the simultaneous appearance of these two tumors.

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Treatment options

Conservative therapeutic approach is preferable in the management of small-sized placental chorangioma, as most of them are asymptomatic.

Therefore, US monitoring at a 6-8-week interval is preferable in such cases.

On the other hand, a giant placental chorangioma with a size greater than 5cm, may require a different approach.

US with Doppler imaging should be performed at 1-2 weeks [6,22].

As chorangioma could lead to various feto-maternal complications before delivery, some interventions are required to maintain fetal viability.

These include interventions such as fetal transfusion, amnioreduction, induced vascular sclerosis with absolute alcohol injection, surgical ligation of placental vessels feeding the tumor, and laser ablation [23-26].

These techniques harbor significant risks, however, they may be a lifesaver for the fetus.

Still, if complications develop earlier, emergency delivery should be considered without hesitation [27].

Large size chorangiomas are usually associated with polyhydramnios, leading to high perinatal morbidity and mortality [1].

Thus, therapeutic amniocentesis, as well as maternal indomethacin treatment, are widely used to treat polyhydramnios.

For fetal lung maturity acceleration, steroid administration is recommended, which is a potent stimulant for alveolocyte maturation and surfactant production [1].

Differential diagnosis

The differential diagnosis of a chorangioma includes placental teratomas, blood clots or leiomyomas.

Compared to chorangiomas, leiomyomas tend to be located on the maternal surface.

The well-delineated aspect of a chorangioma is easily distinguishable from a partial mole due to its diffuse pattern [1].

Also, chorangiosis and chorangiomatosis are worth to be mentioned as differential diagnoses, since they are presented as focal or diffuse proliferation of villous angioblastoma with villi, which are not seen in a chorangioma case [15].

Conclusions

Giant placental chorangioma (GPC) is considered a rare indolent tumor that could lead to serious feto-maternal complications if left untreated.

A diagnosis of placental chorangioma may be suspected on clinical grounds, including US imaging, but the final diagnosis can only be confirmed histologically.

We reported a GPC case in a preterm pregnancy that significantly differs from the usual characteristics of such cases.

Namely male fetal sex, absence of maternal hypertension or diabetes, absence of polyhydramnios, and, finally, remarkable tumor size.

The unfortunate fetal outcome of this case, caused by severe congestive heart failure, highlights the importance of timely antenatal diagnosis.

Appropriate management is necessary to prevent possible complications and to ensure a favorable perinatal outcome.

To our best knowledge, this is the first case reporting the coexistence of a maternal neuroendocrine carcinoma associated with a benign placental tumor.

Acknowledgements

BD drafted the paper and performed the design of the study.

AB contributed to the case description and literature review.

SZB and RB performed the clinical management of the case and interpretation of the clinical data.

MT contributed to pathological (gross and histology), and immunohistochemical assessment, critically revised the manuscript, and conferred the final agreement for publication.

All authors contributed to the article and approved the submitted version.

Disclosure Statement

The authors report there are no competing interests to declare.

Conflict of Interest Statement

The authors declare that the research was conducted without any commercial or financial relationships that could be construed as a potential conflict of interest.

References

1. Kodandapani S, Shreshta A, Ramkumar V, Rao L. Chorioangioma of placenta: a rare placental cause for adverse fetal outcome. Case Rep Obstet Gynecol, 2012, 2012:913878.
2. Asokan S, Chadalavada K, Gardi R, Sastry V. Prenatal diagnosis of placental tumor by ultrasound. J Clin Ultrasound, 1978, 6:180-181.
3. Campbell S. A short history of sonography in obstetrics and gynaecology. Facts Views Vis Obgyn, 2013, 5:213-229.
4. Bromley B, Benacerraf BR. Solid masses on the fetal surface of the placenta: differential diagnosis and clinical outcome. J Ultrasound Med, 1994, 13:883-886.
5. Sepulveda W, Aviles G, Carstens E, Corral E, Perez N. Prenatal diagnosis of solid placental masses: the value of color flow imaging. Ultrasound Obstet Gynecol, 2000, 16:554-558.
6. Yadav M, Maheshwari M, Sharma S, Godha Z, Garg P, Sharma G. Chorioangioma of Placenta: A Rare Case of Near-Miss Mortality. J Obstet Gynaecol India, 2017, 67:224-226.
7. Zacharis K, Kravvaritis S, Charitos T, Chrysafopoulou E, Fouka A. A rare case of a giant placental chorioangioma with favorable outcome. Pan Afr Med J, 2020, 36: 214.
8. Miliaras D, Anagnostou E, Papoulidis I, Miliara X. Non-trophoblastic tumor of the placenta with combined histologic features of chorangioma and leiomyoma. Placenta, 2011, 32:102-104.
9. Kataria N, Singh A, Bedi PK. Giant Placental Chorangioma: A Rare Case Report. J Clin Diagn Res, 2016, 10: ED03-4.
10. Guschmann M, Henrich W, Dudenhausen JW. Chorioangiomas--new insights into a well-known problem. II. An immuno-histochemical investigation of 136 cases. J Perinat Med, 2003, 31:170-175.
11. Vig T, Tirkey RS, Jacob SE, Manoj Kumar R, Yenuberi H, Rathore S, et al. Placental chorioangioma with an emphasis on rare giant placental chorioangioma and associated maternal and perinatal outcome: Clinicopathological study in a single centre. J Family Med Prim Care, 2022, 11:5116-5122.
12. Wou K, Chen MF, Mallozzi A, Brown RN, Shrim A. Pregnancy outcomes and ultrasonographic diagnosis in patients with histologically-proven placental chorioangioma. Placenta, 2011, 32:671-674.
13. Wu Z, Hu W. Clinical analysis of 26 patients with histologically proven placental chorioangiomas. Eur J Obstet Gynecol Reprod Biol, 2016, 199:156-163.
14. Kesrouani AK, Safi J, El Hajj MA. Rapid evolution of placental chorioangioma: natural progression and outcome. J Ultrasound Med, 2013, 32:545-547.
15. Amer HZM, Heller DS. Chorangioma and related vascular lesions of the placenta-a review. Fetal Pediatr Pathol, 2010, 29:199-206.
16. Fan M, Skupski DW. Placental chorioangioma: literature review. J Perinat Med, 2014, 42:273-279.
17. Lifschitz-Mercer B, Fogel M, Kushnir I, Czernobilsky B. Chorangioma. A cytoskeletal profile. Int J Gynecol Pathol, 1989, 8:349-356.
18. Lež C, Fures R, Hrgovic Z, Belina S, Fajdic J, Münstedt K. Chorangioma placentae. Rare Tumors, 2010, 2: e67.
19. Dhar H. Giant placental chorioangioma with intrauterine fetal death. JNMA J Nepal Med Assoc, 2013, 52:384-387.
20. Zanardini C, Papageorghiou A, Bhide A, Thilaganathan B. Giant placental chorioangioma: natural history and pregnancy outcome. Ultrasound Obstet Gynecol, 2010, 35:332-336.
21. Hong L, Hua L, Yao P, Zhang L. A rare giant placental chorioangioma with favourable outcome: a case report. J Obstet Gynaecol, 2022, 42:351-353.
22. Abdalla N, Piórkowski R, Stanirowski P, Pazura M, Cendrowski K, Sawicki W. Can ultrasound be helpful in selecting optimal management methods for pregnancies complicated by placental non-trophpblastic tumors? J Ultrason, 2017, 17:116-122.
23. Zalel Y, Weisz B, Gamzu R, Schiff E, Shalmon B, Achiron R. Chorioangiomas of the placenta: sonographic and Doppler flow characteristics. J Ultrasound Med, 2002, 21:909-913.
24. Wanapirak C, Tongsong T, Sirichotiyakul S, Chanprapaph P. Alcoholization: the choice of intrauterine treatment for chorioangioma. J Obstet Gynaecol Res, 2002, 28:71-75.
25. Haak MC, Oosterhof H, Mouw RJ, Oepkes D, Vandenbussche FP. Pathophysiology and treatment of fetal anemia due to placental chorioangioma. Ultrasound Obstet Gynecol, 1999, 14:68-70.
26. Quintero RA, Reich H, Romero R, Johnson MP, Gonçalves L, Evans MI. In utero endoscopic devascularization of a large chorioangioma. Ultrasound Obstet Gynecol, 1996, 8:48-52.
27. Escribano D, Galindo A, Arbués J, Puente JM, De la Fuente P. Prenatal management of placental chorioangioma: value of the middle cerebral artery peak systolic velocity. Fetal Diagn Ther, 2006, 21:489-493.
28. Glanc P. Fetal and placental metastases associated with maternal cancers. Abdominal Radiology, 2023, 48:1784-1792.

Corresponding Author: Diana Burlacu, PhD student, 3rd year resident in Pathology Department of Targu-Mures Emergency Clinical Hospital, e-mail address: dianaburlacu@yahoo.com

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