DOI: https://doi.org/10.17816/DD623322

Multiple Biliary Microhamartomas Diagnosed in an Unsuspecting Elderly Patient

Marina Balbino[1], Manuela Montatore[1], Giacomo Fascia[1], Ruggiero Tupputi[2], Federica Masino[1], Gianmichele Muscatella[1], Domenico Mannatrizio[1], Giuseppe Guglielmi[1,2,3]

[1] University of Foggia, Foggia, Italy;
[2] Dimiccoli Hospital, Barletta, Italy;
[3] Casa Sollievo della Sofferenza Hospital, Foggia, Italy

ABSTRACT

Multiple biliary hamartomas are a benign incidental finding in the liver. They are not easily detected if one has never seen them, and if appropriate imaging tests are unavailable, and also can be challenging to differentiate from other liver lesions based on imaging alone. Thus, this study aimed to expand the radiologist's digital image library, enabling a quick and precise differential diagnosis. This paper also highlights the importance of thorough radiological assessment and need for a multidisciplinary approach, involving radiologists, hepatologists, and pathologists, to ensure a precise diagnosis.

The patient presented at the hospital for a computed tomography scan and an abdominal magnetic resonance imaging recommended by his general practitioner to assess the biliary tree (magnetic resonance cholangiopancreatography), owing to persistent abdominal pain. The patient had never undergone an abdominal magnetic resonance imaging previously; hence, the discovery of hepatic lesions was incidental and unexpected.

Magnetic resonance imaging revealed multiple benign lesions in both the hepatic lobes comparable to the Von Meyenburg complex. These lesions are multiple hamartomas and behave differently in all magnetic resonance imaging sequences.

Images acquired with different magnetic resonance imaging sequences were carefully examined. Multiple lesions were found scattered throughout the liver; however, the lesions were benign and consistent with the diagnosis of multiple biliary hamartomas.

Medical practitioners should examine the presence of multiple biliary hamartomas and consider them in the differential diagnosis when patients present with hepatic abnormalities. This can prevent unnecessary interventions and guide appropriate patient management.

Keywords: liver; hamartomas; Von Meyenburg complex; magnetic resonance imaging; diagnostic imaging; magnetic resonance cholangiography; bile duct neoplasms.

To cite this article:
Balbino M, Montatore M, Fascia G, Tupputi R, Masino F, Muscatella G, Mannatrizio D, Guglielmi G. Multiple biliary microhamartomas diagnosed in an unsuspecting elderly patient. *Digital Diagnostics.* 2024;5(2):334–341. DOI: https://doi.org/10.17816/DD623322

Submitted: 14.11.2023 **Accepted:** 19.12.2023 **Published online:** 02.07.2024

DOI: https://doi.org/10.17816/DD623322

Множественные билиарные микрогамартомы, случайно диагностированные у пожилого пациента

M. Balbino[1], M. Montatore[1], G. Fascia[1], R. Tupputi[2], F. Masino[1], G. Muscatella[1], D. Mannatrizio[1], G. Guglielmi[1,2,3]

[1] Университет Фоджи, Фоджа, Италия;
[2] Dimiccoli Hospital, Барлетта, Италия;
[3] Casa Sollievo della Sofferenza Hospital, Фоджа, Италия

АННОТАЦИЯ

Множественные билиарные гамартомы, также называемые комплексами фон Мейенбурга — это доброкачественные новообразования печени, которые могут быть выявлены случайно. Их нелегко обнаружить, если ранее не доводилось с ними встречаться, а также если соответствующие методы визуализации на момент обследования недоступны. К тому же их сложно отличить от других поражений печени на основании визуализационных данных. Таким образом, описание данного клинического случая направлено на пополнение библиотеки цифровых изображений, что позволит радиологу быстро и точно провести дифференциальную диагностику. Кроме того, подчёркивается важность тщательной рентгенологической оценки этого заболевания и необходимость мультидисциплинарного подхода с участием радиологов, гепатологов и патологов для постановки точного диагноза.

Пациент поступил в больницу для проведения компьютерной томографии и магнитно-резонансной томографии брюшной полости по рекомендации терапевта для оценки состояния билиарного дерева (магнитно-резонансная холангиопанкреатография) в связи с постоянными болями в животе. Пациенту никогда ранее не проводили инструментальное обследование брюшной полости, поэтому новообразования печени оказались случайной и неожиданной находкой.

Магнитно-резонансная томография выявила множественные доброкачественные образования в обеих печёночных долях, сопоставимые с комплексами фон Мейенбурга. Эти поражения представляют собой множественные гамартомы и ведут себя по-разному во всех последовательностях сканирования.

В ходе исследования были тщательно изучены изображения, полученные с помощью различных последовательностей магнитно-резонансной томографии. В печени обнаружены множественные очаги поражения с признаками доброкачественности, которые соответствовали диагнозу «множественные билиарные гамартомы».

Практикующие специалисты должны уметь распознать это заболевание и учитывать его при дифференциальной диагностике у пациентов с патологиями печени. Это позволит предотвратить ненужные вмешательства и выбрать правильную тактику ведения пациентов.

Ключевые слова: печень; гамартомы; комплекс фон Мейенбурга; магнитно-резонансная томография; диагностическая визуализация; магнитно-резонансная холангиография; новообразования желчных протоков.

Как цитировать:
Balbino M., Montatore M., Fascia G., Tupputi R., Masino F., Muscatella G., Mannatrizio D., Guglielmi G. Множественные билиарные микрогамартомы, случайно диагностированные у пожилого пациента // Digital Diagnostics. 2024. Т. 5, № 2. С. 334–341. DOI: https://doi.org/10.17816/DD623322

Рукопись получена: 14.11.2023 **Рукопись одобрена:** 19.12.2023 **Опубликована online:** 02.07.2024

DOI: https://doi.org/10.17816/DD623322

一名老年患者偶然诊断出的多发性胆管微小错构瘤

Marina Balbino[1], Manuela Montatore[1], Giacomo Fascia[1], Ruggiero Tupputi[2], Federica Masino[1], Gianmichele Muscatella[1], Domenico Mannatrizio[1], Giuseppe Guglielmi[1,2,3]

[1] University of Foggia, Foggia, Italy;
[2] Dimiccoli Hospital, Barletta, Italy;
[3] Casa Sollievo della Sofferenza Hospital, Foggia, Italy

摘要

多发性胆管错构瘤又称 von Meyenburg 复合体，是一种可能偶然发现的良性肝肿瘤。如果以前没有遇到过，而且在检查时没有适当的成像技术，就不容易被发现。根据影像学检查结果，它们也很难与其他肝脏病变区分开来。因此，本病例的描述旨在为数字图像库增添新的内容，使放射科医生能够做出快速准确的鉴别诊断。此外，该病例还强调了对该疾病进行全面放射学评估的重要性，以及放射科医生、肝病科医生和病理科医生多学科合作以做出准确诊断的必要性。

患者因持续腹痛，在全科医生的建议下入院进行腹腔计算机断层扫描和磁共振成像，以评估胆道树的状况（磁共振胰胆管造影）。患者以前从未接受过腹腔器械检查，因此肝脏肿瘤是一个偶然的意外发现。

磁共振成像显示了，两个肝叶都有多发性良性病变，与 von Meyenburg 复合体相似。这些病变代表多发性错构瘤，在所有扫描序列中表现不同。

研究仔细检查了不同磁共振成像序列获得的图像。在肝脏中发现了多个具有良性迹象的病灶，符合“多发性胆管错构瘤”的诊断。

医生应该能够识别这种疾病，并在肝脏病变患者的鉴别诊断中加以考虑。这将有助于避免不必要的干预，并能选择正确的治疗策略。

关键词：肝脏；错构瘤；von Meyenburg 复合体；磁共振成像；诊断成像；磁共振胰胆管造影；胆管肿瘤。

引用本文：
Balbino M, Montatore M, Fascia G, Tupputi R, Masino F, Muscatella G, Mannatrizio D, Guglielmi G. 一名老年患者偶然诊断出的多发性胆管微小错构瘤. *Digital Diagnostics.* 2024;5(2):334–341. DOI: https://doi.org/10.17816/DD623322

收到： 14.11.2023 **接受：** 19.12.2023 **发布日期：** 02.07.2024

INTRODUCTION

Multiple biliary hamartomas are relatively rare and often an incidental finding during imaging studies. Identifying and accurately diagnosing these lesions are crucial because they can mimic other hepatic lesions, such as cysts or tumors, leading to potentially unnecessary invasive procedures or treatments [1-4].

DESCRIPTION OF THE CASE

Medical History

An 82-year-old male presented to the hospital with a primary complaint of abdominal pain persisting for more than 6 weeks. The patient has no personal/family history significant for liver disease. A thorough medical evaluation, including physical examination and blood tests, was conducted. However, these initial assessments did not yield conclusive results nor provide a clear diagnosis.

Diagnostic Assessment

The patient underwent contrast-enhanced CT (computed tomography) and subsequently MRI [5–6].

Multiple millimetric disorganized hypodense lesions, both subcapsular and intraparenchymal, were detected incidentally in both hepatic lobes on CT.

After the administration of contrast medium, enhancement of lesions was not observed (Fig. 1).

Subsequent MRI examination identified the same lesions with different characteristics in various sequences. In T2-weighted MRI images, the areas appear homogeneously and intensely hyperintense (Fig. 2).

In T1-weighted images, all the lesions were homogeneously hypointense.

In diffusion-weighted imaging (DWI), the lesions persist as hyperintense at a low B-value (50 sec/mm^2) but disappear at a high B-value (800) (Fig. 3).

In T2-weighted MRI–cholangiography sequences, the liver presents a "starry sky" appearance due to the presence of multiple small hyperintense lesions; however, the communication with the bile duct are usually not recognized [7] (Fig. 4).

The dynamic study after contrast administration did not show enhancement in the arterial and venous phases (Fig. 5).

Differential Diagnosis

Radiographic findings may be nonspecific and may not differentiate biliary hamartomas from other lesions.

The differential diagnosis is made with polycystic liver disease, multiple simple hepatic cysts, metastases, micro-abscesses, and Caroli's disease [8]. A more heterogeneous appearance of lesions and communication with the bile duct is observed in the latter.

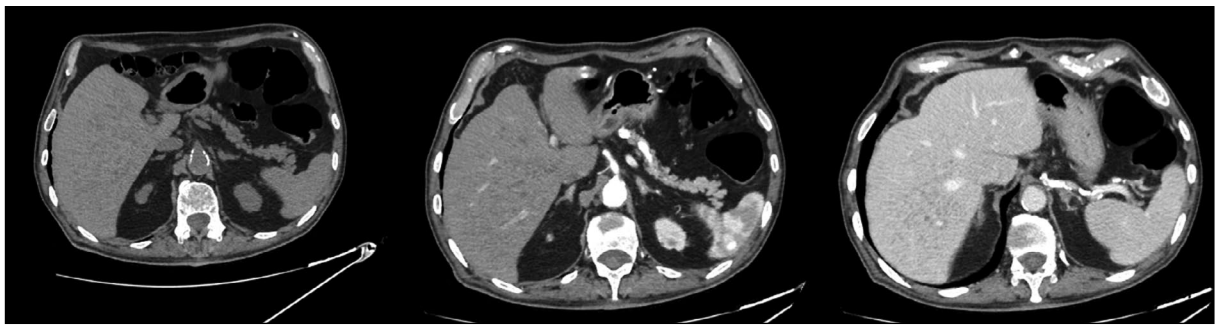

Fig. 1. Axial CT image: hypodense lesions without significant contrast enhancement in the arterial and venous phases.

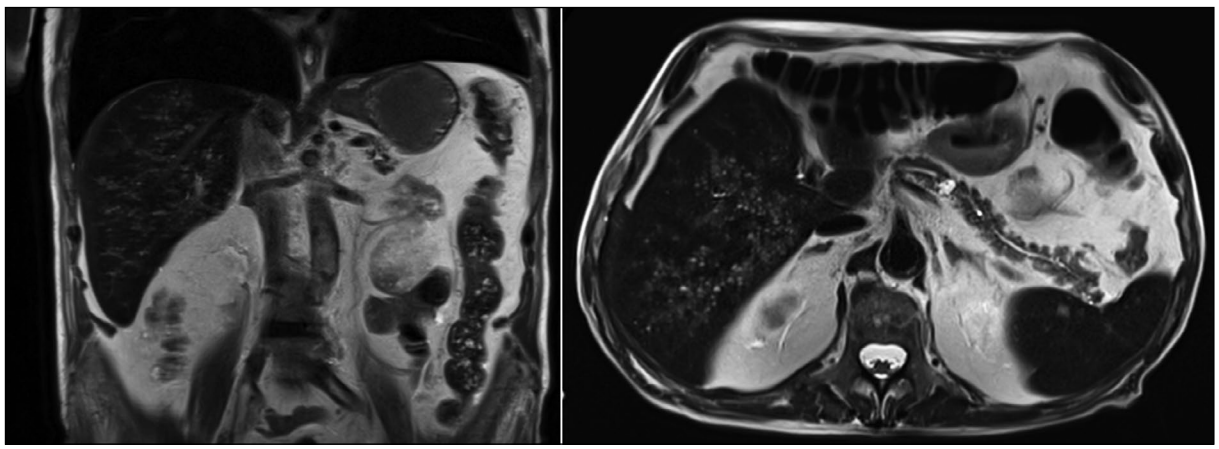

Fig. 2. Coronal and axial MRI images with HASTE T2 sequence: multiple small hyperintense lesions throughout the hepatic area.

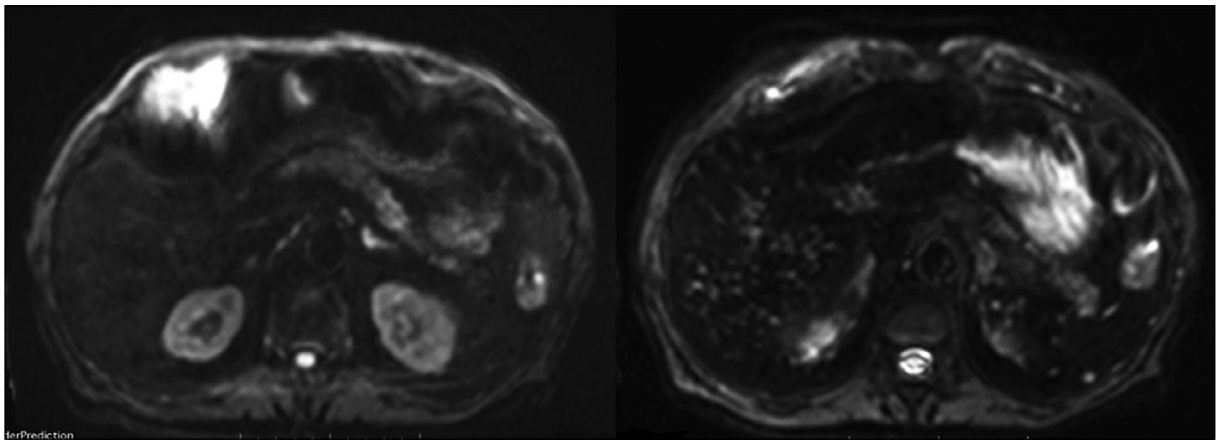

Fig. 3. Axial MRI image with DWI sequence: hyperintense lesions at low B-value (50 sec/mm^2) on the right, disappearing at high B-values on the left (800).

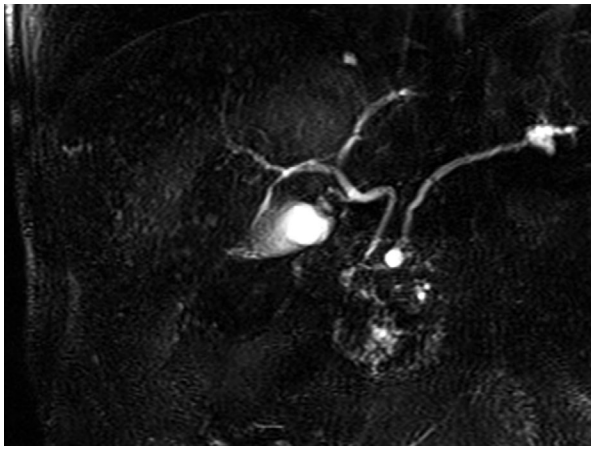

Fig. 4. MRI image with T2 MRI–cholangiography sequence: multiple small hyperintense lesions; the liver with a "starry sky" appearance.

Interventions

In the present case, the lesions were not directly related to symptoms or complications therefore, no surgical or pharmacological treatment was required.

If Von Meyenburg complexes are causing symptoms or complications, treatment may include surgical removal of the affected liver tissue or drainage of large cysts [9–11].

It is critical for individuals diagnosed with these complexes to consult a hepatologist or liver specialist to determine the appropriate management approach based on their specific case.

Follow-up and Outcomes

The management of the patient's condition was based on observation and scheduled monitoring, allowing for the timely assessment of any potential alterations. However, no significant changes led to alterations in the diagnostic choices made.

DISCUSSION

Multiple biliary hamartomas, also called Von Meyenburg complexes, are rare benign liver lesions characterized by small disorganized cystic structures affecting both hepatic lobes with some predilection for subcapsular regions [12–13]. They are multiple small round, or irregular lesions and are usually 5–30 mm in size on imaging.

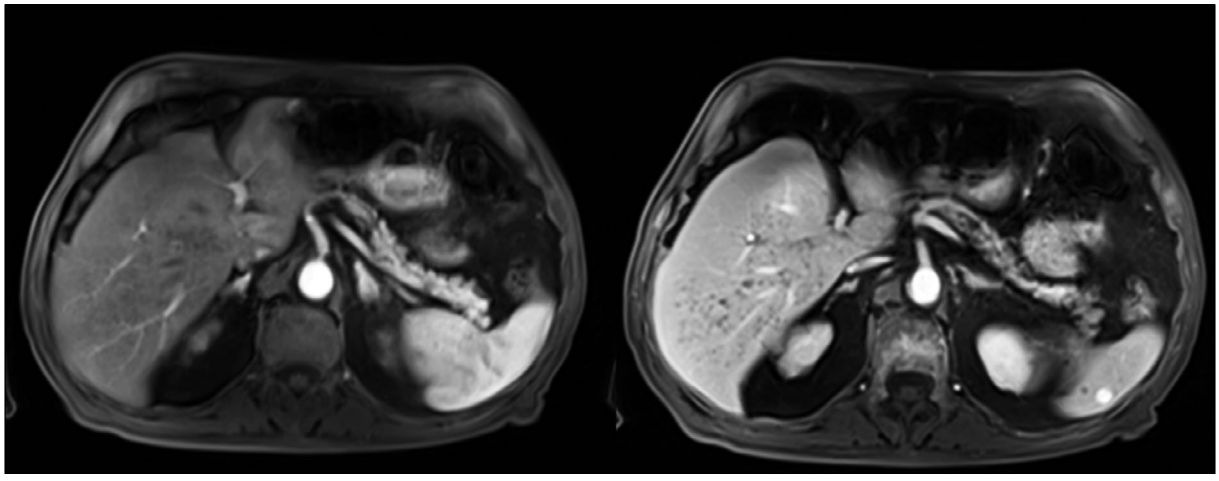

Fig. 5. Axial MRI image with T1 sequence showing hypointense lesions without significant contrast enhancement in the arterial and venous phases.

These structures are composed of abnormal bile ducts and are typically discovered incidentally during medical imaging studies, such as ultrasound, CT, and MRI, which are often performed for other reasons. Although hamartomas can communicate with the biliary tree, but usually not. They are believed to arise from embryonic bile duct remnants that have failed to involute.

Von Meyenburg complexes are considered noncancerous and are usually asymptomatic. They are generally not associated with liver dysfunction or clinical symptoms. Laboratory data are generally nonspecific and within normal limits. Most individuals with these lesions do not require treatment. In some cases, Von Meyenburg complexes can be associated with various liver conditions, including polycystic liver disease, Caroli's disease, and congenital hepatic fibrosis [14].

When these conditions are present, they can lead to more significant liver-related problems and may require medical management.

CONCLUSION

Von Meyenburg complexes a rare medical condition that is often detected incidentally. This study highlights the importance of accurate diagnosis and challenges in distinguishing these lesions from other hepatic lesions. Management based on observation and periodic monitoring is a feasible approach when the lesions are asymptomatic. However, clinical awareness on multiple biliary hamartomas is crucial to avoid misdiagnosis and unnecessary invasive interventions. Future studies may further our understanding of this condition and its clinical implications.

ADDITIONAL INFORMATION

Funding source. This study was not supported by any external sources of funding.

Competing interests. The authors declare that they have no competing interests.

Authors' contribution. All authors made a substantial contribution to the conception of the work, acquisition, analysis, interpretation of data for the work, drafting and revising the work, final approval of the version to be published and agree to be accountable for all aspects of the work.

Consent for publication. Written consent was obtained from the patient for publication of relevant medical information and all of accompanying images within the manuscript in Digital Diagnostics Journal.

REFERENCES

1. Zheng RQ, Zhang B, Kudo M, Onda H, Inoue T. Imaging findings of biliary hamartomas. *World J Gastroenterol.* 2005;11(40):6354–6359. doi: 10.3748/wjg.v11.i40.6354

2. Gil-Bello D, Ballesteros E, Sanfeliu E, Andreu FJ. Calcification in biliary hamartomatosis. *Br J Radiol.* 2012;85(1012):e099–e101. doi: 10.1259/bjr/95019559

3. Thommesen N. Biliary hamartomas (von Meyenburg complexes) in liver needle biopsies. *Acta Pathol Microbiol Scand A.* 1978;86(2):93–99. doi: 10.1111/j.1699-0463.1978.tb02019.x

4. Aguado IC, Álvarez MH, Hernández JS, La Orden Izquierdo E. Hamartomatosis biliar en una lactante con colitis alérgica: revisión a propósito de un caso. *Rev Pediatr Aten Primaria.* 2013;15(59):e111–e114. doi: 10.4321/S1139-76322013000400014

5. Horton KM, Bluemke DA, Hruban RH, Soyer P, Fishman EK. CT and MR imaging of benign hepatic and biliary tumors. *Radiographics.* 1999;19(2):431–451. doi: 10.1148/radiographics.19.2.g99mr0443

6. Brancatelli G, Federle MP, Vilgrain V, et al. Fibropolycystic liver disease: CT and MR imaging findings. *RadioGraphics.* 2005;25(3):659–670. doi: 10.1148/rg.253045114

7. Bravo-Acosta M, Rosendo-Namías J, Martínez-Méndez D. Hamartomatosis biliar múltiple: "imagen en cielo estrellado". *Rev Gastroenterol MEX.* 2020;86(2). doi: 10.1016/j.rgmx.2020.08.002

8. Choi BI, Yeon KM, Kim SH, et al. Caroli disease: central dot sign in CT. *Radiology.* 1990;174(1):161–163. doi: 10.1148/radiology.174.1.2294544

9. Kin HK, Jin SY. Cholangiocarcinoma arising in von Meyenburg complexes. *Korean J Hepatol.* 2011;17(2):161–164. doi: 10.3350/kjhep.2011.17.2.161

10. Song JS, Lee YJ, Kim KW, et al. Cholangiocarcinoma arising in von Meyenburg complexes: report of four cases. *Pathol Int.* 2008;58(8):503–512. doi: 10.1111/j.1440-1827.2008.02264.x

11. Xu AM, Xian ZH, Zhang SH, Chen XF. Intrahepatic cholangiocarcinoma arising in multiple bile duct hamartomas: report of two cases and review of the literature. *Eur J Gastroenterol Hepatol.* 2009;21(5):580–584. doi: 10.1097/MEG.0b013e3282fc73b1

12. Venkatanarasimha N, Thomas R, Armstrong EM, et al. Imaging features of ductal plate malformations in adults. *Clin Radiol.* 2011;66(11):1086–1093. doi: 10.1016/j.crad.2011.05.008

13. Desmet VJ. Pathogenesis of ductal plate malformation. *J Gastroenterol Hepatol.* 2004;19(S7):S356–S360. doi: 10.1111/j.1440-1746.2004.03702.x

14. Soreide K, Korner H, Havnen J, et al. Bile duct cysts in adults. *Br J Surg.* 2004;91(12):1538–1548. doi: 10.1002/bjs.4815

СПИСОК ЛИТЕРАТУРЫ

1. Zheng R.Q., Zhang B., Kudo M., Onda H., Inoue T. Imaging findings of biliary hamartomas // World J Gastroenterol. 2005. Vol. 11, N 40. P. 6354–6359. doi: 10.3748/wjg.v11.i40.6354

2. Gil-Bello D., Ballesteros E., Sanfeliu E., Andreu F.J. Calcification in biliary hamartomatosis // Br J Radiol. 2012. Vol. 85, N 1012. P. e099–e101. doi: 10.1259/bjr/95019559

3. Thommesen N. Biliary hamartomas (von Meyenburg complexes) in liver needle biopsies // Acta Pathol Microbiol Scand A. 1978. Vol. 86, N 2. P. 93–99. doi: 10.1111/j.1699-0463.1978.tb02019.x
4. Aguado I.C., Álvarez M.H., Hernández J.S., La Orden Izquierdo E. Hamartomatosis biliar en una lactante con colitis alérgica: revisión a propósito de un caso // Rev Pediatr Aten Primaria. 2013. Vol. 15, N 59. P. e111–e114. doi: 10.4321/S1139-76322013000400014
5. Horton K.M., Bluemke D.A., Hruban R.H., Soyer P., Fishman E.K. CT and MR imaging of benign hepatic and biliary tumors // Radiographics. 1999. Vol. 19, N 2. P. 431–451. doi: 10.1148/radiographics.19.2.g99mr0443
6. Brancatelli G., Federle M.P., Vilgrain V., et al. Fibropolycystic liver disease: CT and MR imaging findings // RadioGraphics. 2005. Vol. 25, N 3. P. 659–670. doi: 10.1148/rg.253045114
7. Bravo-Acosta M., Rosendo-Namías J., Martínez-Méndez D. Hamartomatosis biliar múltiple: "imagen en cielo estrellado" // Rev Gastroenterol MEX. 2020. Vol. 86, N 2. doi: 10.1016/j.rgmx.2020.08.002
8. Choi B.I., Yeon K.M., Kim S.H., et al. Caroli disease: central dot sign in CT // Radiology. 1990. Vol. 174, N 1. P. 161–163. doi: 10.1148/radiology.174.1.2294544
9. Kin H.K., Jin S.Y. Cholangiocarcinoma arising in von Meyenburg complexes // Korean J Hepatol. 2011. Vol. 17, N 2. P. 161–164. doi: 10.3350/kjhep.2011.17.2.161
10. Song J.S., Lee Y.J., Kim K.W., et al. Cholangiocarcinoma arising in von Meyenburg complexes: report of four cases // Pathol Int. 2008. Vol. 58, N 8. P. 503–512. doi: 10.1111/j.1440-1827.2008.02264.x
11. Xu A.M., Xian Z.H., Zhang S.H., Chen X.F. Intrahepatic cholangiocarcinoma arising in multiple bile duct hamartomas: report of two cases and review of the literature // Eur J Gastroenterol Hepatol. 2009. Vol. 21, N 5. P. 580–584. doi: 10.1097/MEG.0b013e3282fc73b1
12. Venkatanarasimha N., Thomas R., Armstrong E.M., et al. Imaging features of ductal plate malformations in adults // Clin Radiol. 2011. Vol. 66, N 11. P. 1086–1093. doi: 10.1016/j.crad.2011.05.008
13. Desmet V.J. Pathogenesis of ductal plate malformation // J Gastroenterol Hepatol. 2004. Vol. 19, N S7. P. S356–S360. doi: 10.1111/j.1440-1746.2004.03702.x
14. Soreide K., Korner H., Havnen J., et al. Bile duct cysts in adults // Br J Surg. 2004. Vol. 91, N 12. P. 1538–1548. doi: 10.1002/bjs.4815

AUTHORS' INFO

*** Giuseppe Guglielmi,** MD, Professor;
ORCID: 0000-0002-4325-8330;
e-mail: giuseppe.guglielmi@unifg.it

Marina Balbino, MD;
ORCID: 0009-0009-2808-5708;
e-mail: marinabalbino93@gmail.com

Manuela Montatore, MD;
ORCID: 0009-0002-1526-5047;
e-mail: manuela.montatore@unifg.it

Giacomo Fascia, MD;
ORCID: 0000-0001-5244-5093;
e-mail: giacomo.fascia@unifg.it

Ruggiero Tupputi, MD;
e-mail: rutudott@gmail.com

Federica Masino, MD;
ORCID: 0009-0004-4289-3289;
e-mail: federicamasino@gmail.com

Gianmichele Muscatella, MD;
ORCID: 0009-0004-3535-5802;
e-mail: muscatella94@gmail.com

Domenico Mannatrizio, MD;
ORCID: 0000-0003-3365-7132;
e-mail: dr.mannatrizio@gmail.com

ОБ АВТОРАХ

*** Giuseppe Guglielmi,** MD, Professor;
ORCID: 0000-0002-4325-8330;
e-mail: giuseppe.guglielmi@unifg.it

Marina Balbino, MD;
ORCID: 0009-0009-2808-5708;
e-mail: marinabalbino93@gmail.com

Manuela Montatore, MD;
ORCID: 0009-0002-1526-5047;
e-mail: manuela.montatore@unifg.it

Giacomo Fascia, MD;
ORCID: 0000-0001-5244-5093;
e-mail: giacomo.fascia@unifg.it

Ruggiero Tupputi, MD;
e-mail: rutudott@gmail.com

Federica Masino, MD;
ORCID: 0009-0004-4289-3289;
e-mail: federicamasino@gmail.com

Gianmichele Muscatella, MD;
ORCID: 0009-0004-3535-5802;
e-mail: muscatella94@gmail.com

Domenico Mannatrizio, MD;
ORCID: 0000-0003-3365-7132;
e-mail: dr.mannatrizio@gmail.com

* Corresponding author / Автор, ответственный за переписку