Egyptian Journal of Radiology and Nuclear Medicine

CASE REPORT Open Access

Hepatic adrenal rest tumor in a patient with multifactorial liver cirrhosis: a case report with CT and MRI findings and pathologic correlation

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Abstract

Background: Adrenal rest tumor is an ectopic collection of adrenocortical cells in an extra-adrenal site, more frequently located around the kidney, retroperitoneum, spermatic cord, para-testicular region and broad ligament, but very rarely occurring also in the liver. Hepatic adrenal rest tumor poses a diagnostic challenge in differentiating it from hepatocellular carcinoma, particularly in a cirrhotic liver.

Case presentation: An 83-years-old male was referred to our hospital by his family doctor for hepatological evaluation due to multifactorial liver cirrhosis. Ultrasound revealed a centimetric hypoechoic nodule in the VI hepatic segment in the context of a liver with signs of cirrhosis and steatosis. The patient first underwent MRI and then CT, which showed a fat containing focal liver lesion in the subcapsular location of the right lobe, strictly adjacent to the homolateral adrenal gland. The nodule was hypervascular in the arterial phase, washed out in the portal-venous and transitional phases, resulting hypointense in the hepato-biliary phase at MR imaging. In the suspicion of a hepatocellular carcinoma, the nodule was surgically removed, and the patient's postoperative course was unremarkable. The final histopathological diagnosis was of adrenal rest tumor of the liver.

Conclusions: Hepatic adrenal rest tumor is an extremely rare hepatic tumor, often without any clinical manifestation, that can also occur in the cirrhotic liver as in our case. Although there are not specific imaging findings, the possible diagnosis of HART should be considered when we observe a well-defined lesion in the subcapsular location of the right lobe, with fat containing, hypervascularity after contrast medium injection and vascular supply from the right hepatic artery.

Keywords: Hepatic adrenal rest tumor, Liver, Liver neoplasms, CT, MR imaging

Background

Adrenal rest tumor is an ectopic collection of adrenocortical cells in an extra-adrenal site [1], more frequently located around the kidney, retroperitoneum, spermatic cord, para-testicular region and broad ligament [2, 3], but very rarely occurring also in the liver. These tumors may be nonfunctional or hormonally active with manifesting as an endocrine syndrome [4].

Histologically, hepatic adrenal rest tumor (HART) is composed by sheets or confluent cords of large polygonal cells with abundant eosinophilic and clear cytoplasm, similar to adrenocortical tissue.

At imaging, HART generally appears as round, well-defined mass, with subcapsular localization in the right

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hepatic lobe, showing macroscopic fat-containing and hypervascularity [1, 5–7]. These radiological findings can also be present in other focal liver lesions, which range from hepatocellular adenoma and angiomyolipoma (AML) to hepatocellular carcinoma (HCC) with fatty change [1, 5–7]. Moreover, patients with chronic liver disease, especially with cirrhosis, who present with a solid focal liver lesion are at a very high risk for having HCC and in this setting the correct diagnosis is particularly relevant and critical for patient's management.

Case presentation

An 83-years-old male was referred to our hospital by his family doctor for hepatological evaluation due to multifactorial liver cirrhosis.

His medical history was positive for metabolic diseases (in particular insulin resistance and dyslipidemia), prostatic hypertrophy, past hepatitis B virus (HBV) and hepatitis C (HCV) infections, the latter eradicated with antiviral therapy in 2018.

The laboratory investigation showed a normal level of alpha-fetoprotein (AFP, 6.0 ng/mL), a normal hepatic function (albumin level 4.3 gr/dL, total bilirubin level 1.6 mg/dL, prothrombin activity 79%) and hepatic enzymes within normal levels (aspartate aminotransferase 23 U/L, alanine aminotransferase 18 U/L, gamma-glutamyl transferase 19 U/L, alkaline phosphatase 94 U/L).

At ultrasound follow-up, a centimetric hypoechoic nodule was identified in the VI hepatic segment in the context of a liver with signs of cirrhosis and steatosis.

Considering the medical history of the patient, the hepatologist requested a MR study of the liver to better characterize the focal liver lesion.

MRI was performed on 1.5 T system (Signa Excite HDxt; GE Healthcare) with a phased-array multi-channel coil; examination was carried out using spoiled gradient-echo (SPGR) T1-weighted in- and out-phase sequences, T2-weighted images, and diffusion-weighted imaging. A 3D fat-suppressed breath-hold T1-weighted LAVA (Liver Acquisition with Volume Acceleration) sequence was performed before and after intravenous administration of Gd-EOB-DTPA (Primovist®, Bayer HealthCare) in a dose of 0.1 mmol/kg of body weight as a bolus injection at a flow-rate of 2 ml/s, followed by injection of isotonic saline (25 ml). Contrast-enhanced dynamic images were acquired in the arterial dominant phase, in the portal-venous phase, in the transitional phase (at 3 min) and in the hepato-biliary phase (about 20 min after administration).

MR confirmed the presence of an 11-mm-in size nodule in the subcapsular VI hepatic segment, near the inferior vena cava, with signal intensity decay in the in/out-of-phase T1-weighted sequences (Fig. 1), whereas the hepatic nodule was not clearly recognizable on diffusion-weighted imaging. The lesion was strictly adjacent to the right adrenal gland from which it was not possible to recognize a clear cleavage plan. After contrast

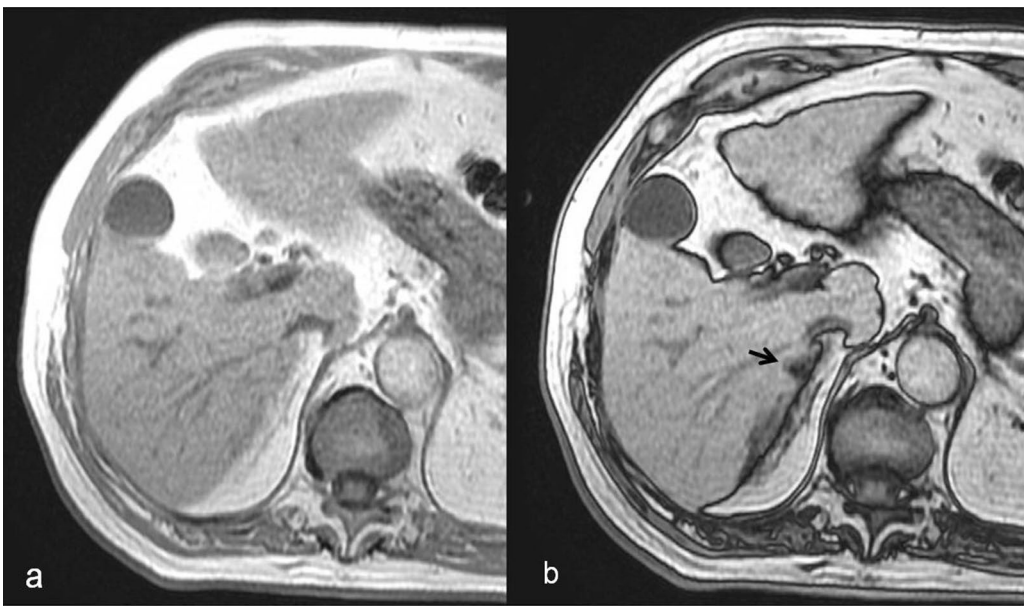

Fig. 1 **a**, **b**—Axial in-phase (**a**) and out-of-phase (**b**) MR images exhibit signal drop of the subcapsular lesion in the VI segment (black arrow)

injection, the nodule appeared hyperintense in the arterial phase, showed a wash-out in the portal-venous and transitional phases and resulted hypointense in the hepatobiliary phase (Fig. 2).

On the basis of a possible radiological diagnosis of HCC and to better define the anatomical details and relationship of the nodule with the right adrenal gland, the patient underwent CT of the abdomen that was performed using a 64-slice CT scanner (LightSpeed VCT; GE Healthcare). Patient received plain and triphasic contrast-enhanced CT (arterial, portal-venous and delayed phase) after intravenous injection of non-ionic iodinated

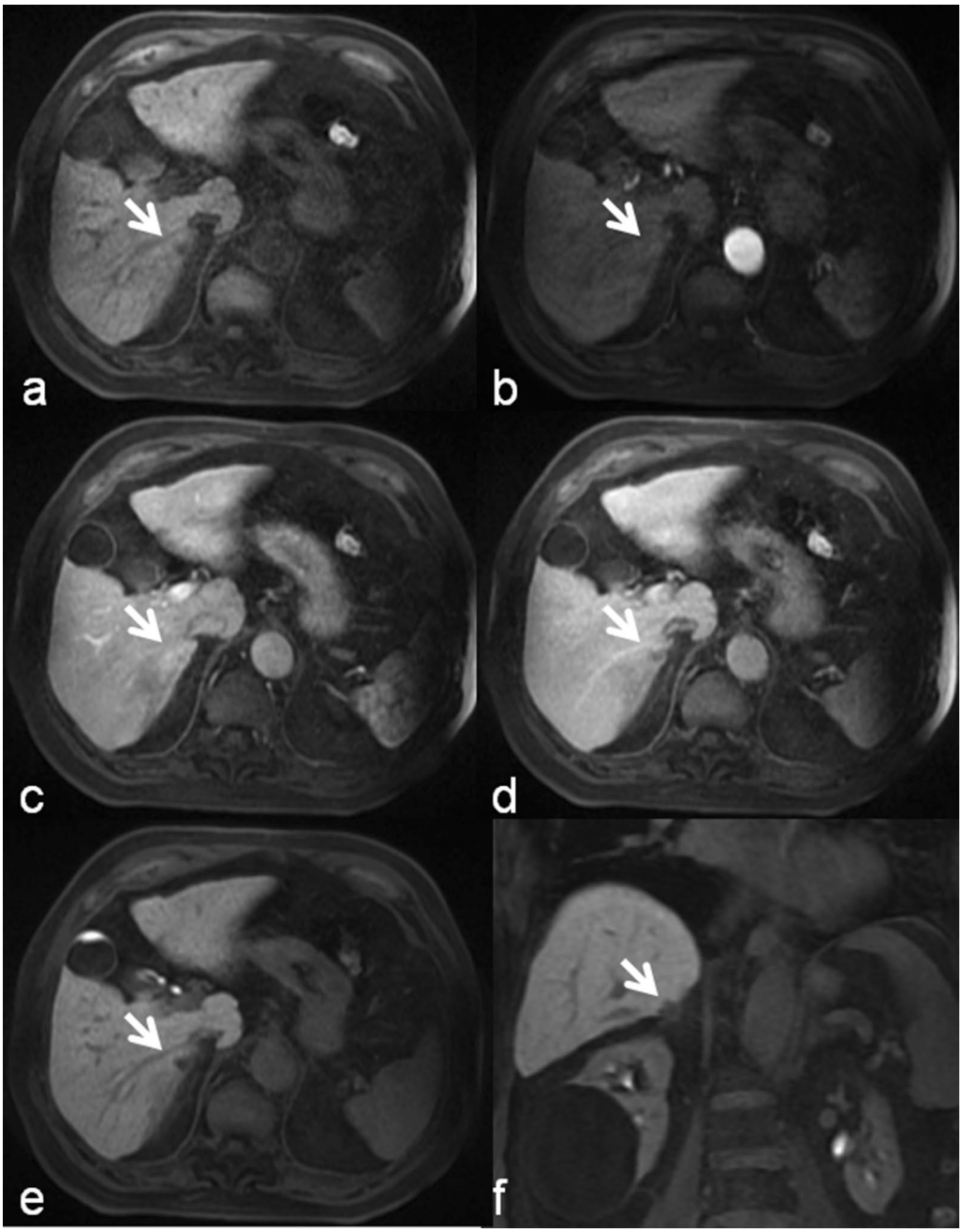

Fig. 2 **a**–**f**—Unenhanced fat-suppressed MR image (**a**) shows a hypointense nodule in the VI segment (white arrow). After Gd-EOB-DTPA injection, the lesion is slightly hyperintense in the arterial phase (**b**) and hypointense in the portal-venous (**c**) and transitional (**d**) phases (white arrows). The lesion is clearly hypointense in the hepatobiliary phase both in the axial (**e**) and in the coronal (**f**) images (white arrows)

contrast medium (iomeprolo 400 mg/mL, Iomeron 400, Bracco), covering the upper abdomen. On CT scans, the nodule was hyperdense in the arterial phase, appeared hypodense in the delayed phase (Fig. 3) and was vascularized by a thin collateral branch of the right hepatic artery (Fig. 4). Right adrenal gland was partially visualized and very close to the nodule (Fig. 3).

The radiological orientation was that of a neoplastic nature of the nodule, in particular for the diagnosis of hepatocellular carcinoma in a cirrhotic liver. This was supported by the vascular pattern of the nodule both on MRI and CT, by the hypointensity in the hepato-biliary phase and by the clinical context. However, it was not possible to exclude with certainty the possible adrenal relevance also for the signal intensity of the nodule in the in/out-of-phase sequences.

After a long multidisciplinary discussion (hepatologist, radiologist, abdominal surgeon and oncologist), the team decided to propose the surgical treatment to the patient, who accepted. The opportunity of performing a liver biopsy of the nodule was considered, but it was judged technically not feasible by the interventional radiologist.

The patient underwent surgical atypical resection of the caudate process and of a portion of the VI segment near the inferior vena cava, partial resection of the right adrenal gland and cholecystectomy. The patient's postoperative course was unremarkable.

At macroscopic examination of the surgical specimen, the pathologist observed a yellowish-orange lesion measuring $2 \times 1.5 \times 1$ cm that was located in the subcapsular portion of the liver. Histologically, the mass was composed of large polygonal cells with eosinophilic or pale and lipid-rich cytoplasm, arranged in nest or trabecular pattern with abundant capillary vessels similar to adrenal cortex (Fig. 5). Immunohistochemically, the lesion expressed Melan-A/MART1 and did not stain for liver marker HepPar1 and renal marker PAX8.

The morphology along with the immunohistochemical profile supported the final diagnosis of adrenal rest tumor of the liver.

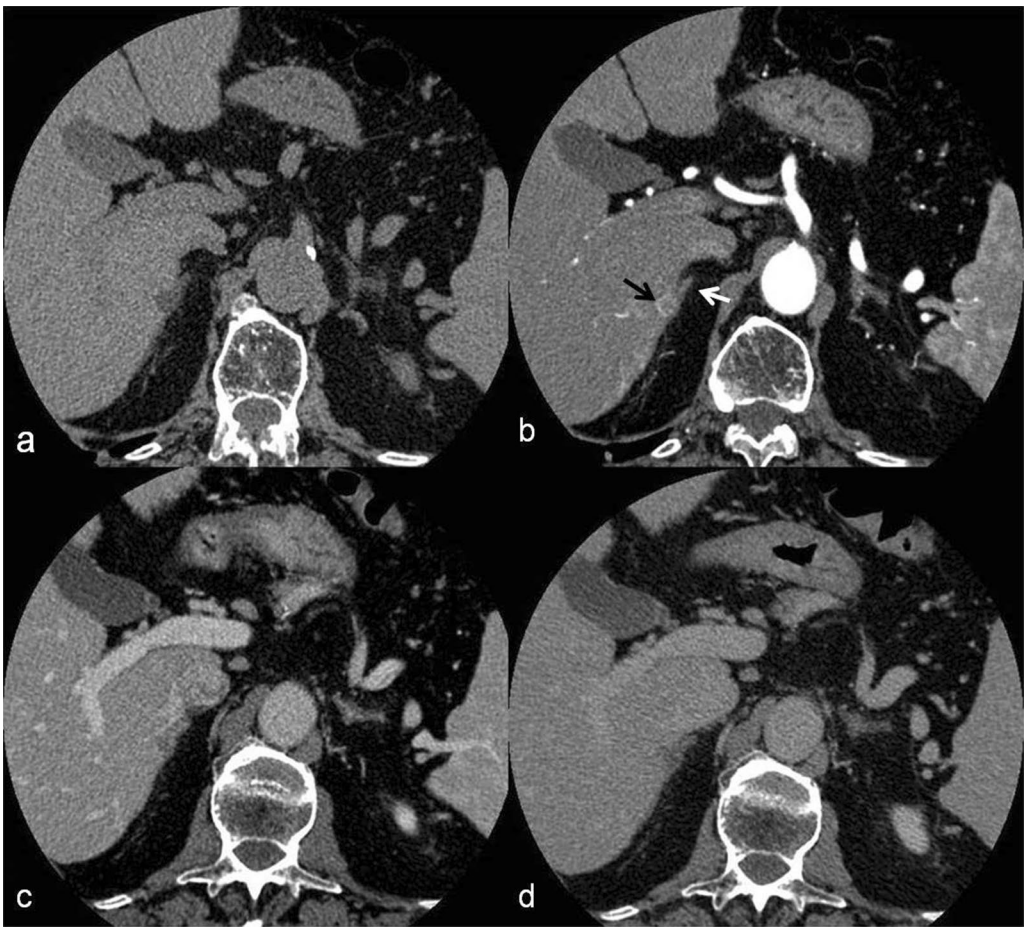

Fig. 3 **a**–**d**—Plain CT scan (**a**) depicts a low-density nodule in the VI segment. The nodule (black arrow) is hypervascular in the arterial phase (**b**) and is not distinctly cleavable from the right adrenal gland (white arrow). The lesion washed out in the portal-venous (**c**) and delayed (**d**) phases

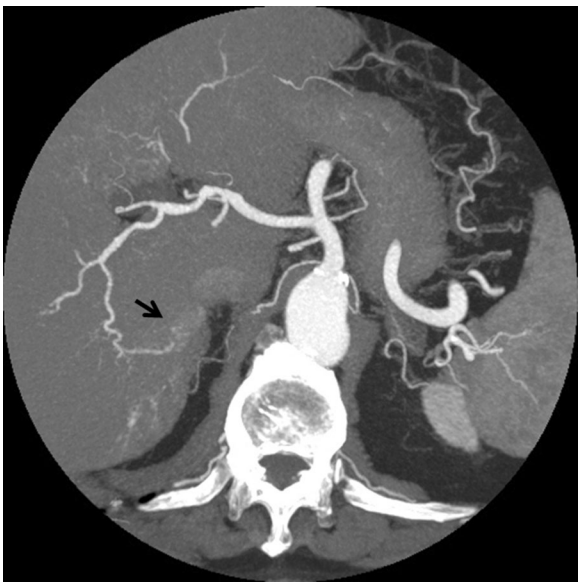

Fig. 4 MIP reconstruction in the axial oblique plane from arterial CT scans shows that the vascular supply of the lesion (black arrow) arises from the posterior branch of the right hepatic artery

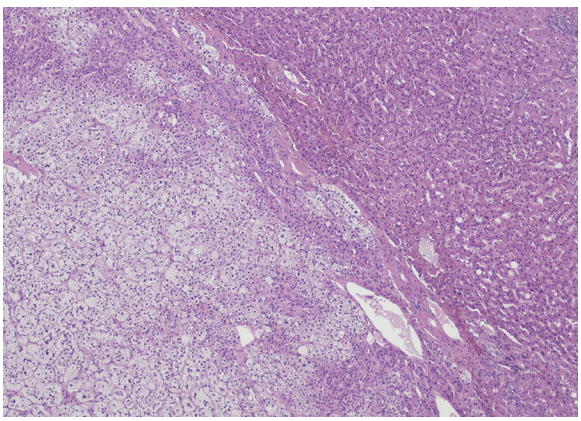

Fig. 5 Microscopically, the lesion was composed of epithelioid cells with a pale cytoplasm that formed nests or cords accompanied by abundant capillary vessels, similar manner with the features of the adrenocortical tissue. The cell nucleus was round or oval and presented slight atypia (HE × 10HPF)

Discussion

As already reported above, the adrenal rest tumor is considered a collection of adrenocortical cells outside the adrenal gland. The adrenal cells have different embryological origin: the cortex is derived from the mesoderm and the medulla from the ectoderm of the neural crest. It is not clear what happens during the fetal development when an ectopic collection of adrenocortical cells is formed. Its occurrence is postulated for the localization near the genital structures [2, 3], where the adrenal rests within the genital structures are due to mechanical separation and displacement of portions of cortical tissue associated with gonadal tissue during its descent. Bozic et al. suggested that the most common locations where this ectopic tissue can be found might be related to misplaced mesothelial cells or autonomous differentiation of mesodermal elements [8].

The ectopic adrenocortical tissue is rarely detected in the liver, and the adrenal rest tumor is quite rare. These tumors may be nonfunctional or functional with manifesting as an endocrine syndrome due to hormonal product [4, 9]; howsoever, they are generally nonfunctional and clinically asymptomatic, and a malignant transformation is anecdotic [4].

In our patient with HBV/HCV-related and metabolic cirrhosis, the HART appeared as a round, well defined nodule, with subcapsular localization in the right hepatic lobe. The preoperative diagnosis was very difficult due to the clinical context and to the absence of specific characteristic at CT and MR imaging. Furthermore, the lesion was difficult to cleave from the right adrenal gland on imaging.

However, there are radiological findings that can address toward the correct diagnosis of HART. The first one is the identification of the fat component [7] inside the nodule that reflects the presence of intracytoplasmic lipid droplets in the tumor cells; for the identification of this feature, MRI represents the most specific diagnostic technique thanks to the typical behavior on chemical shift imaging. In fact, axial in- and out-phase images showed lower signal intensity of the lesion on the out-of-phase image compared with the in-phase image.

The second one is the hypervascularity of the nodule [1]; although this is not a specific radiological finding, it was identified at the dynamic study both on MRI and on CT scans. Besides, CT well highlighted that vascular supply of HART was mainly represented by the posterior branch of the right hepatic artery.

Few case reports of HARTs are currently published in the literature; anyway, most of them appear as hypervascular and fat-containing lesion as in our case [1, 5–7]. Interestingly, in other published cases [1, 5–7] hepatic adrenal rest tumors had similar location to the current case, in the posterior part of the right hepatic lobe or subcapsular. This localization of HART might be explained by the prevailing migration of aberrant adrenal tissue in this portion of the hepatic lobe. Besides, our case concurs with a previous case report by Tajima et al. [1], suggesting that the vascular supply of HART is mainly provided by hepatic arteries.

Nevertheless, imaging features of HART can be nonspecific, and differential diagnosis with other hepatic

lesions should always be considered preoperatively [5, 7]. The radiological differential diagnosis should first include hepatic lesions containing fat components and then the hypervascular focal liver lesions [7–10]. In the present case, hepatic AML and HCC with massive fatty metamorphosis are the most probable differential diagnoses due to the fat internal component, the fibrous capsule and distinctive neovascularity associated with the tumor. Furthermore, in our case the fact that the patient had a liver cirrhosis made the differential diagnosis even more complex and directed the therapeutic choice of the multidisciplinary team.

Macroscopically the HART appeared as a well-circumscribed, yellowish, nodular lesion occurring beneath the capsule of the right hepatic lobe. In these lesions, close morphological examination including immunohistochemistry [11] may be warranted to allow an adequate diagnosis and differential diagnosis especially for perivascular epithelioid cell tumor, also known as PEComa, renal cell carcinomas metastasis and hepatocellular carcinomas.

Conclusions

HART is a very rare hepatic tumor, often without any clinical manifestation, that can also occur in the cirrhotic liver as in our case. Although there are not specific imaging findings, the possible diagnosis of HART should be considered when we observe a well-defined lesion in the subcapsular location of the right lobe, with fat containing, hypervascularity after contrast medium injection and vascular supply from right hepatic artery. MRI has a potential role in improving the diagnosis of HART.

Abbreviations

HART: Hepatic adrenal rest tumor; HCC: Hepatocellular carcinoma; AML: Angiomyolipoma; HBV: Hepatitis B virus; HCV: Hepatitis C virus; AFP: Alpha-fetoprotein; SPGR: Spoiled gradient-echo; LAVA: Liver Acquisition with Volume Acceleration; PEComa: Perivascular epithelioid cell tumor.

Acknowledgements

None.

Authors' contributions

PB, FT, FD, FP and PC collected the data, provided the images, reviewed the literature and supported in writing the manuscript. AB performed the histological examination of the liver and supported in writing the manuscript. NR and LU performed surgery and supported in writing the manuscript. PB, PC and LU revised the manuscript for intellectual content. All the authors read and approved the final version of the manuscript.

Funding

None.

Availability of data and material

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Ethical approval was waived by the local Ethics Committee, since it is a retrospective case report study. Patient provided written informed consent to the processing of personal data even for study purposes.

Consent for publication

Written consent was obtained.

Competing interests

The authors declare that they have no competing interests.

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Received: 1 March 2021 Accepted: 11 November 2021
Published online: 17 November 2021

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