Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

An inflammatory myofibroblastic tumor of the ampulla of Vater, an exceptional location: a case report

Aliou Zabeirou, MD[a,*], Boubacar Efared, MD[b], Tarek Souiki, MD[c], Lassey James Didier, MD[b], Karim I. Majdoub, MD[c], Imane Toughrai, MD[c], Laila Chbani, MD[c], Khalid Mazaz, MD[c]

Introduction and importance: Inflammatory myofibroblastic tumors constitute a group of mesenchymal tumors associated with inflammatory infiltration. They occur mainly in young patients. It is classified by the World Health Organization as a borderline neoplasm. They are observed in many organs, particularly the lungs. Digestive localization is rare, and localization into the ampulla of Vater has been reported once.

Case presentation: We report the case of a 39-year-old patient who was admitted for cholestatic jaundice with right hypochondrium pain. Computed tomography and magnetic resonance imaging revealed a tumor at the biliopancreatic junction. A cephalic duodenopancreatectomy was performed, and a histological examination of the surgical specimen revealed an inflammatory myofibroblastic tumor of the ampulla of Vater. The postoperative evolution was without any complications.

Clinical discussion: This is the second case of localization of an inflammatory myofibroblastic tumor in Vater's ampulla. The therapeutic approach is the complete excision of these inflammatory tumors, thus reducing the risk of local recurrence. In the literature, all cases of incomplete excision have resulted in recurrences.

Conclusion: Inflammatory myofibroblastic tumors are rare. The diagnosis was based on histopathological findings and confirmed using immunohistochemical techniques.

Keywords: ampulla of Vater, cephalic duodenopancreatectomy, inflammatory myofibroblastic tumor, surgical resection

Introduction

Inflammatory myofibroblastic tumors were described by Brunn for the first time in 1939 in the lung, which is the site of predilection. These tumors develop from mesenchymal tissues, and their etiopathogenesis is still not elucidated[1]. They have several synonyms because of the variability of their cellular composition, such as inflammatory pseudotumors, plasma cell granulomas, postinflammatory tumors, xanthomatous pseudotumors, and fibrous histiocytomas[2]. They can invade adjacent organs, recur after excision, or exceptionally cause distant metastases. The debate persists regarding their inflammatory, tumoral, benign, or malignant nature. They are classified into the intermediate category by the WHO. The diagnosis is almost always based on a histopathological examination and is confirmed by immunohistochemical techniques[3]. Treatment is poorly codified, but management is usually surgical[4].

We report the case of a 39-year-old patient who was admitted with an inflammatory myofibroblastic tumor of the ampulla of Vater. Cephalic duodenopancreatectomy was performed to resect the tumor. This article has been reported in line with the SCARE 2020 criteria[5].

HIGHLIGHTS

- Inflammatory myofibroblastic tumors constitute various mesenchymal tumors whose etiopathogenesis is not yet elucidated.
- Digestive localization is rare, and localization into the ampulla of Vater was reported once.
- This is the second case of localization of inflammatory myofibroblastic tumors into the ampulla of Vater, as reported in the medical literature.
- The main effective management procedure is a complete surgical resection.
- Generally, the final diagnosis is made after a histological examination of the surgical specimen, completed by immunohistochemical techniques.

[a]Department of General and Visceral Surgery, Hôpital Général de Référence, [b]Faculty of Health Sciences, Abdou Moumouni University of Niamey, Niamey, Niger and [c]Faculty of Medicine and Pharmacy, Sidi Mohamed Ben Abdellah University of Fez, Fez, Morocco

Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

*Corresponding author. Address: Department of General and Visceral Surgery, Hôpital Général de Référence, Quartier Koirategui, Niamey, Niger. Tel.: +227 842 15518; fax: +227 964 48244. E-mail: ali.zabeirou@gmail.com (A. Zabeirou).

Annals of Medicine & Surgery (2023) 85:6237–6242

Received 23 August 2023; Accepted 9 October 2023

Published online 17 October 2023

http://dx.doi.org/10.1097/MS9.0000000000001416

Case presentation

A 39-year-old male patient with a history of smoking for 15 years was admitted to the surgical department with complaints of mucocutaneous cholestatic jaundice, pruritus, and right hypochondrium pain for 3 months. He also reported weight loss. The clinical examination found a patient with an altered general condition classified as grade 2 on the performance status scale of the ECOG (Eastern Cooperative Oncology Group)/WHO (World Health Organization) system. General examination revealed mucocutaneous jaundice accompanied by scratching lesions. The rest of the clinical examination was unremarkable.

Blood examination found hemoglobin decreased to 8.7 g/dl, a normal platelet rate of 234 000 U/mm^3, hyperleukocytosis at 12 000 U/mm^3, a correct prothrombin level of 72%, total bilirubin at 180 µmol/l (9 times normal), direct bilirubin at 162 µmol/l (40 times normal), gamma-glutamyl transpeptidase at 450 UI/l (10 times normal), alkaline phosphatase at 1250 UI/l (8 times normal), AST (aspartate aminotransferase) at 97 U/l (2 times normal), ALT (alanine aminotransferase) at 110 U/l (2 times normal), and CRP (C-reactive protein) increased to 78 mg/l. The renal function and ionogram were correct.

Abdominal ultrasound showed distension of the gallbladder with dilation of the intrahepatic bile ducts and common bile duct to 15 mm (Fig. 1). Abdominal computed tomography (CT) revealed dilatation of the intrahepatic and common bile ducts with a mass at the biliopancreatic intersection (Fig. 2). Magnetic resonance imaging (MRI) showed stenosis of the lower part of the common bile duct associated with a suspected malignant process at the level of the ampulla of Vater (Fig. 3).

Duodenoscopy revealed an ulcer-budding process in the duodenal papilla. Biopsy revealed nonspecific subacute interstitial duodenitis without malignant proliferation.

The patient was taken into the operating room and underwent laparotomy. Surgical exploration revealed a tumor measuring ~3 cm in size located in Vater's ampulla. The surgical procedure comprised cephalic duodenopancreatectomy according to the Whipple procedure. We then performed a digestive reconstruction according to the Child's procedure (pancreatojejunostomy, hepato-jejunal anastomosis, and gastrojejunostomy). The surgical procedure proceeded without any major complications. The operating time was ~5 h and 30 min. The blood loss was ~200 ml. The postoperative course was unremarkable, and the patient was discharged 7 days postoperatively.

Macroscopic (Fig. 4), histological (Fig. 5), and immunohistochemical (Fig. 6) examination of the surgical specimen revealed a myofibroblastic and inflammatory tumor of Vater's ampulla. The resection margins were healthy. Analysis of peripancreatic lymph node dissection found 8 healthy nodes out of 8.

The patient has been under clinical and CT monitoring for 2 years, with no tumor recurrence or metastases on various surveillance CT scans.

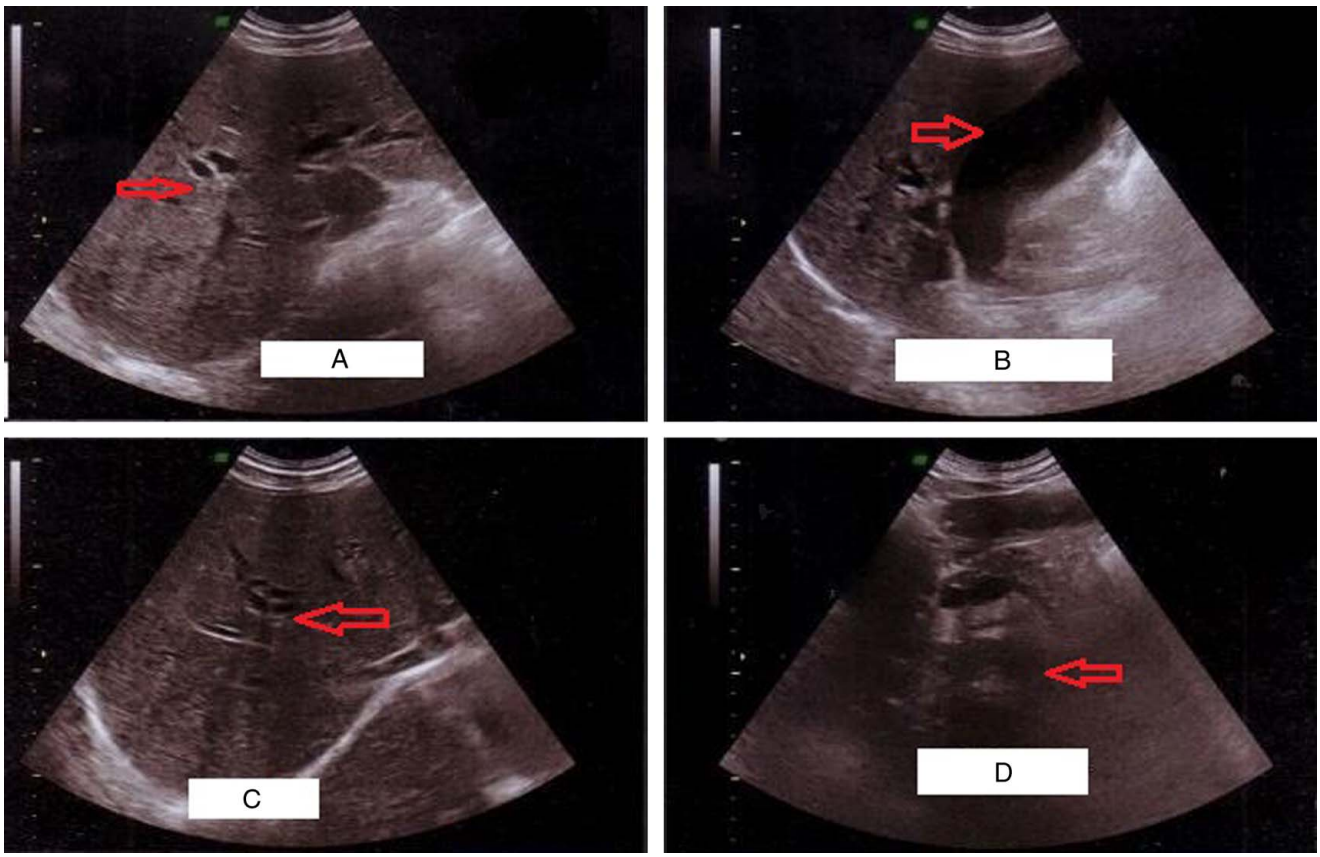

Figure 1. Abdominal ultrasound image showing: (A) intrahepatic bile duct dilatation (red arrow); (B) dilatation of the gallbladder (red arrow); (C) common bile duct dilatation (red arrow); (D) pancreas (red arrow).

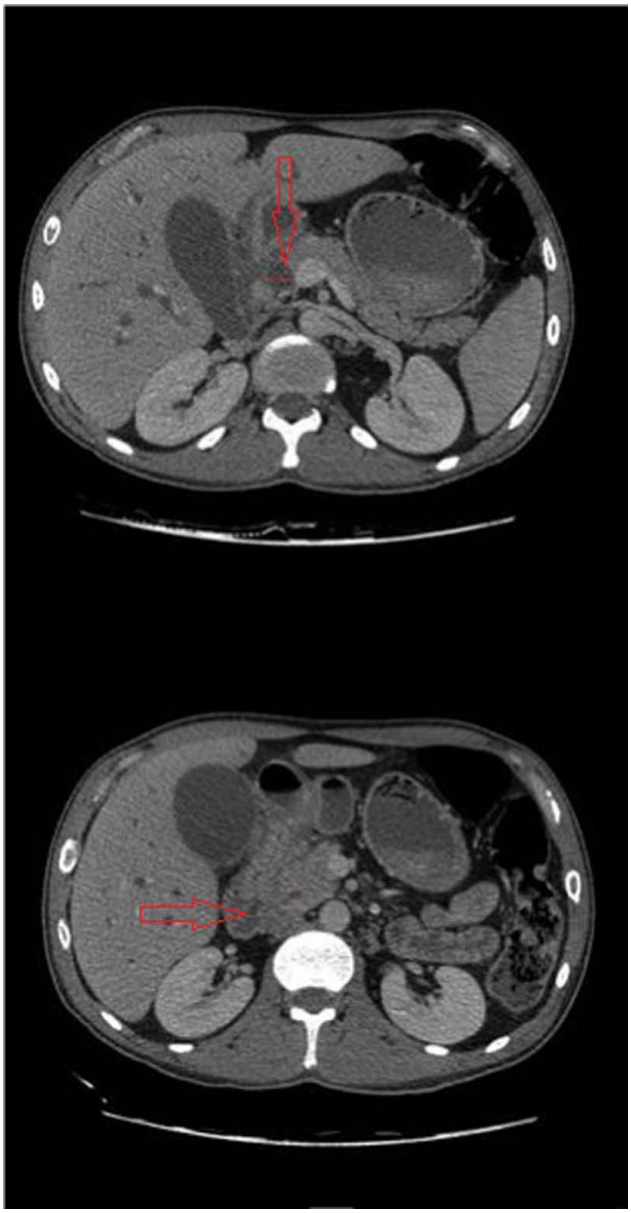

Figure 2. Abdominal computed tomography showing dilatation of the common bile duct (red arrow) upstream of a tissue obstacle.

Discussion

Inflammatory myofibroblastic tumors affect patients of all ages, with a predilection for patients aged 30 years[6]. This tumor affects women and men equally. It is a rare lesion characterized by benign myofibroblastic proliferation with a vague storiform pattern and varying degrees of inflammatory infiltration. It may be characterized by rapid growth and local invasiveness and occurs in all soft tissues of the body. The lung is the main location[7]. The abdominal locations are the omentum and mesentery (43%). Other sites, such as the appendix, Meckel's diverticulum, duodenum, and stomach, remain extremely rare[8]. Only one case of localization of an inflammatory myofibroblastic tumor in the ampulla of Vater was reported. To date, the origin of inflammatory myofibroblastic tumors remains unknown. Several hypotheses have been proposed but not demonstrated[9]. The neoplastic versus reactional nature of inflammatory myofibroblastic tumors remains controversial. Chronic smoking or alcoholism is not clearly associated with the occurrence of inflammatory myofibroblastic tumors. However, viral infections with Epstein–Barr virus and human herpesvirus-8, inflammation, trauma, and surgery may contribute to the development of inflammatory myofibroblastic tumors[10]. Histological examination of an inflammatory myofibroblastic tumor revealed proliferating spindle cells with infiltrating plasma cells, lymphocytes, and eosinophils. Other patterns include compact fascicular spindle cells with myxoid and collagenized regions with an inflammatory cell background and a denser collagen pattern. Immunochemical examination of an inflammatory myofibroblastic tumor showed reactivity to vimentin, smooth muscle actin, muscle-specific actin, and desmin but was negative for myoglobin and S100[11].

The clinical expression of abdominal inflammatory myofibroblastic tumors is nonspecific and depends on the location. Patients generally describe night sweats or various digestive signs, depending on the site, not primarily evoking this type of tumor: nausea, heartburn, vomiting, a feeling of heaviness, and abdominal pain[12]. A clinical examination rarely evokes the diagnosis of an inflammatory myofibroblastic tumor, but it follows the functional complaints of the patient to guide the explorations that will make the diagnosis a certainty[13]. The idea is to perform a general examination, looking for pallor, jaundice, fever, or an alteration in the general condition, as well as looking for an abdominal mass and characterizing it[14]. Biological abnormalities, such as anemia, thrombocytosis, and hypocalcemia, can be associated with these tumors[15]. However, these biological disturbances are nonspecific to inflammatory myofibroblastic tumors. Localization at the biliopancreatic junction can manifest as clinical and biological cholestasis syndrome. Radiological explorations (US, CT scan, MRI) represent an essential time for the diagnosis of these tumors and highlight the extension to neighboring elements, presence of voluminous lymph nodes, or metastasis[16]. The hollow organs allow the use of echo-endoscopy for better analysis and biopsies for histological diagnosis[17]. Treatment of inflammatory myofibroblastic tumors achieves a cure and reduces the risk of recurrence. The treatment is essentially surgical. If disease control is not possible or difficult to achieve through optimal surgery, other therapeutic measures are discussed[18]. Therefore, the treatment in this second scenario is administered to control the size of the tumor, alleviate symptoms, and improve the comfort and quality of life of the affected person. The proposed adjuvant treatments have proven their effectiveness in various ways, including nonsteroidal anti-inflammatory drugs, steroids, and chemotherapy[19]. The prognosis of inflammatory myofibroblastic tumors appears excellent, especially because they have a very low metastatic potential. Radical surgery is sufficient in most cases. The following factors are correlated with a poor prognosis: the large size of the tumor, its site, and its relationship with adjacent vital structures; the presence of metastases; and recurrence despite resection[20].

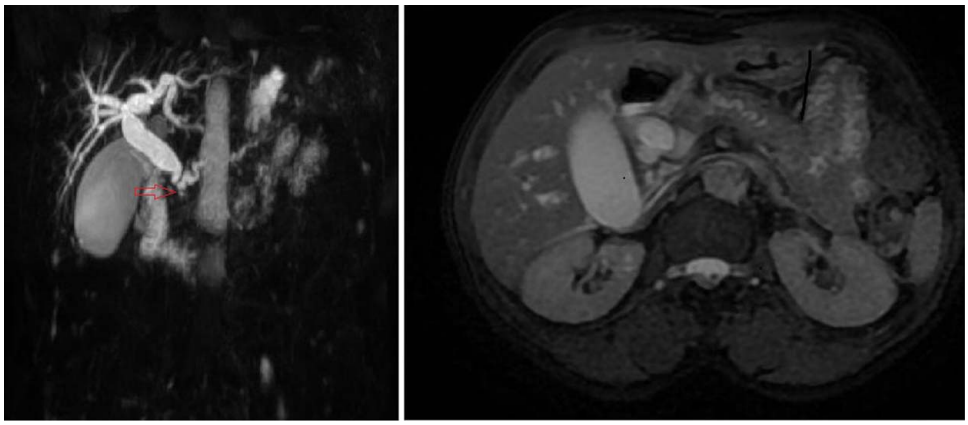

Figure 3. Abdominal magnetic resonance imaging (MRI): axial section and cholangio MRI, we note the dilation of the common bile duct upstream of a stenosis of the distal part of the bile duct (arrow).

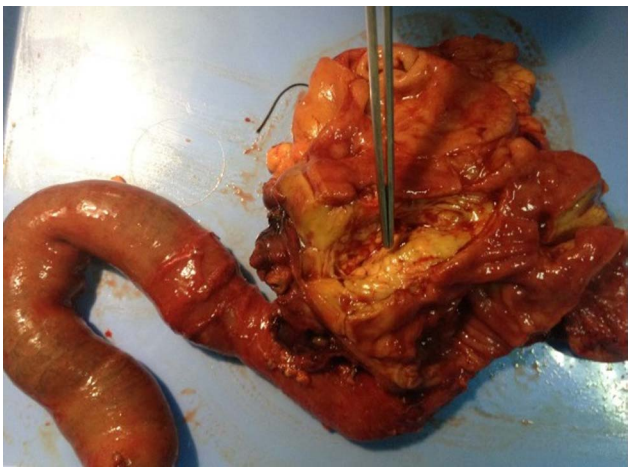

Figure 4. Specimen of cephalic duodenopancreatectomy showing ampulla of Vater tumor process.

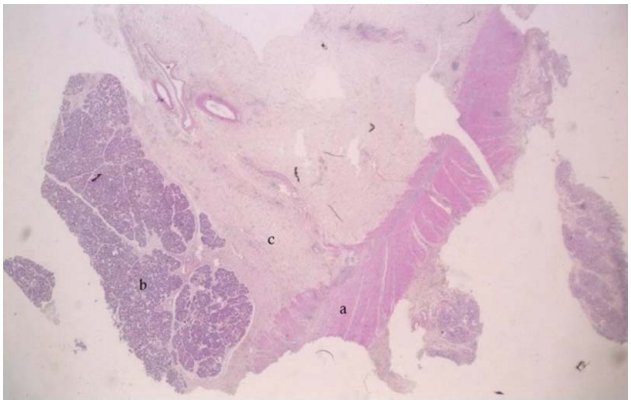

Figure 5. Photomicrograph of fragment showing normal duodenal mucosa (A), pancreatic tissue (B), and inflammatory myofibroblastic tumor (C).

Conclusion

The diagnosis of inflammatory myofibroblastic tumors generally remains a surprise on histological examination. This difficulty lies first in the fact that endoscopic and transparietal biopsy samples are not contributory to the diagnosis and that some patients may undergo iterative samples where the result will be nonspecific inflammation, as in our patient, where the biopsy revealed non-specific duodenitis. It is a diagnosis of exclusion whose certainty can only be conceived with histopathological examination of the surgical specimen completed using immunohistochemical techniques.

Ethics approval

Ethical approval for this study (Ethical Committee No. NAC 52) was provided by the Ethical Committee NAC of our Hospital on 20 August 2023.

Consent for publication

Written informed consent was obtained from the patient for the publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Sources of funding

No sources of funding.

Author contribution

A.Z.: operated on the patient, wrote the article, and made substantial contributions to the conception and design of the article; B.E.: made the histopathological diagnosis; T.S., J.D.L., I.M.K., I.T., L.C., and K.M.: have been involved in drafting the manuscript and revising it critically for important intellectual content. All authors read and approved the final version of the manuscript.

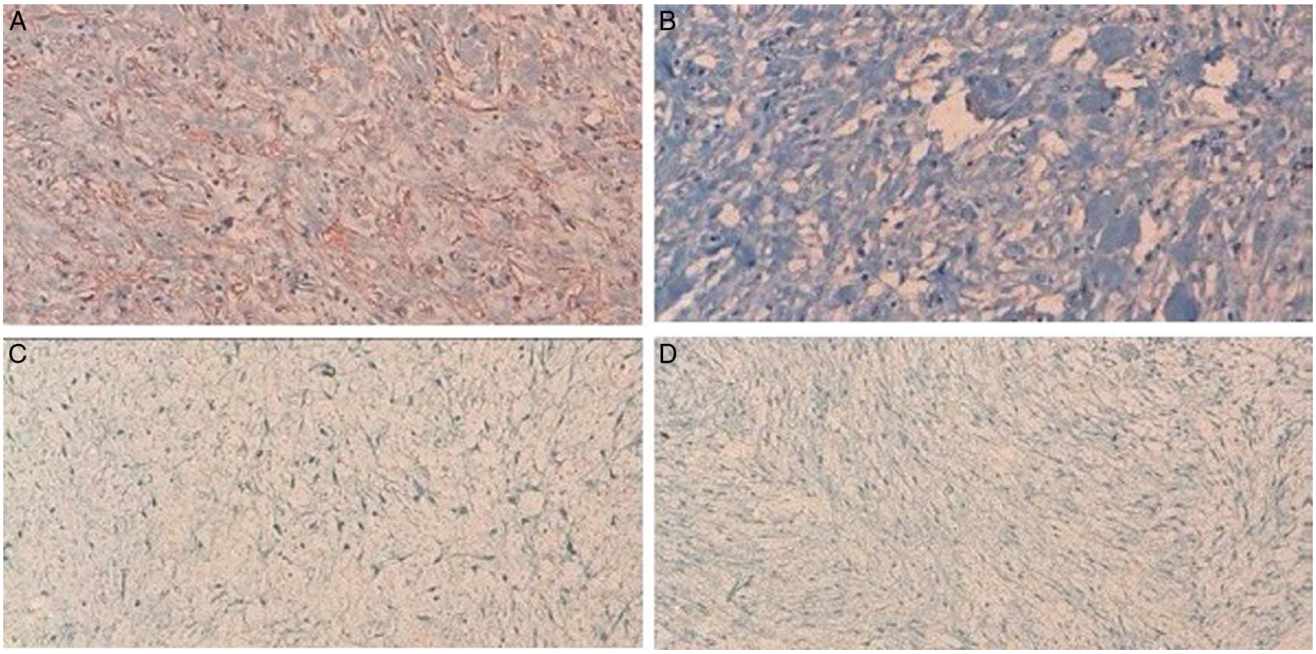

Figure 6. Immunohistochemical study: (A) tumor cells express α-smooth muscle actin, but negative for the rest of the markers, (B) creatine kinase antibodies, (C) anti-desmin antibodies, and (D) myogenin monoclonal antibody.

Conflicts of interest disclosure

All authors declare that they have no conflicts of interest.

Research registration unique identifying number (UIN)

Not applicable.

Guarantor

Zabeirou Aliou.

Provenance and peer-review

Not commissioned, externally and internally peer-reviewed.

Data availability statement

Data sharing is not applicable to this article, as no data sets were generated or analyzed during the current study.

Acknowledgements

The authors thank the patient for providing her written consent for the publication of this case report.

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