

Steroid-induced reconstitution of the biliary tree ravaged by IgG4-related disease

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Abstract

The steroid-induced, rapid healing of the biliary tree ravaged by IgG4-related disease shows that the point of irreversibility remains to be defined.

KEYWORDS

autoimmune pancreatitis, biliary stricture, IgG4, perihilar cholangiocarcinoma, primary sclerosing cholangitis

A 52-year-old man was referred for perihilar cholangiocarcinoma (Figure 1A). The available PET scan showed high activity at the perihilar mass and a discrete 1.7-cm-area in the right lobe (Figure 1B).

The biochemistry results were as follows: AST:88 U/L, ALT:114U/L, ALP: 418 U/L (<105), GGT:383 U/L (5-85);

total bilirubin, CEA, and CA19-9 were within normal limits. The MRCP taken at our institution showed a biliary tree ravaged by sclerosing cholangitis (Figure 2A). The IgG4 level was 1360 mg/dL (<201). The tests for ANA, ALKMA, AMA, ASMA, and p-ANCA were negative. Oral prednisolone treatment (40 mg/day) resulted in a biochemical response on

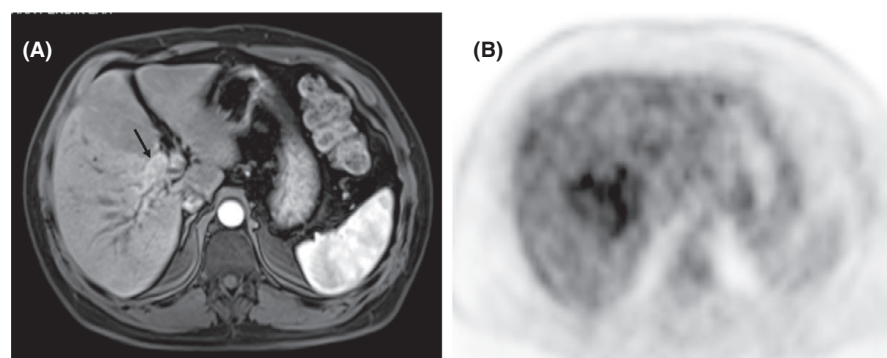
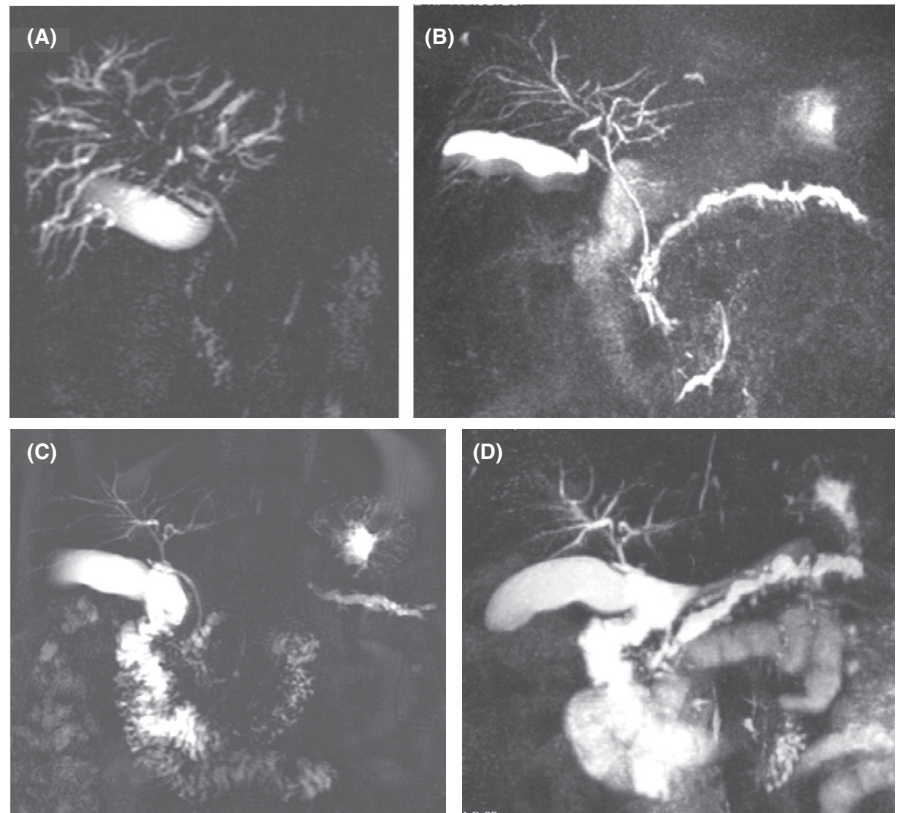


FIGURE 1 A, MRI showed a hypervascular perihilar mass obstructing the biliary tree (arrow), with more prominent dilation in the right lobe. B, High FDG uptake (SUVmax: 10.5) at the hilar mass and a discrete 1.7-cm-area in the right lobe (SUVmax: 8.6) (not visualized on the MRI)

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FIGURE 2 A, The MRCP image before treatment. B, The MRCP image after one month of steroid treatment. C, The MRCP image at 9 mo after the initial MRCP. D, The MRCP image at 21 mo after the initial MRCP



the 6th day and marked improvement of the biliary tree on MRCP at 1 month; however, Wirsung duct dilation persisted. (Figure 2B).

Liver enzyme and IgG4 levels returned to normal limits, and the steroid dose was tapered to 5 mg/day. The MRI at 9 (Figure 2C) and 21 months (Figure 2D) showed a normal biliary tree but a markedly dilated, tortuous Wirsung duct; the hypervascular mass had disappeared. The patient is receiving azathiopurine only (75 mg/day) at 41 months with normal biochemistry.

A steroid trial entails the risk of administering immunosuppression to a malignancy patient.¹ Because our patient was not a candidate for surgery, a trial would not radically worsen his prognosis. Prevention of irreversible injury is vital.² The rapid healing of the seemingly hopeless radiologic picture shows that the point of irreversibility remains to be defined. The progression of the Wirsung duct dilation is unexplained.

ACKNOWLEDGMENTS

Published with written consent of the patient.

CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

İÖ and SK: served as the attending physicians of the patient, wrote and proof-read the article; AP: involved in radiologic assessment, writing and proof-reading; YS: involved in PET assessment, writing and proof-reading.

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REFERENCES

1. Oseini AM, Chaiteerakij R, Shire AM, et al. Utility of serum immunoglobulin G4 in distinguishing immunoglobulin G4-associated cholangitis from cholangiocarcinoma. *Hepatology*. 2011;54:940-948.
2. Khosroshahi A, Wallace ZS, Crowe JL, et al. International Consensus Guidance Statement on the Management and Treatment of IgG4-Related Disease. *Arthritis Rheumatol*. 2015;67:1688-1699.

How to cite this article: Özden İ, Poyanlı A, Sanlı Y, Kaymakoğlu S. Steroid-induced reconstitution of the biliary tree ravaged by IgG4-related disease. *Clin Case Rep*. 2020;8:3552–3553. <https://doi.org/10.1002/ccr3.3109>