Transcatheter Arterial Embolization of IgG4-Related Inflammatory Celiacomesenteric Trunk Aneurysm: A Case Report

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Abstract

A 56-year-old man presenting with abdominal pain had an elevated serum immunoglobulin 4 (IgG4) concentration. Computed tomography angiography revealed a celiacomesenteric trunk aneurysm with wall. After admission, the celiacomesenteric trunk aneurysm grew rapidly along with wall thinning. Emergency transcatheter arterial embolization was completed using detachable coils. After transcatheter arterial embolization, the patient’s abdominal pain disappeared completely. Steroid administration, which continues to the present day, was started 1 month after the transcatheter arterial embolization. No clinical symptoms associated with recurrent arteritis or other IgG4-related disease have been confirmed.

Key words: IgG4-related disease, celiacomesenteric trunk, transarterial embolization

INTRODUCTION

Immunoglobulin G4 (IgG4)-related disease is defined as a high serum IgG4 concentration with idiopathic inflammatory and sclerosing lesions infiltrated by numerous IgG4-positive plasma cells. Although IgG4-related disease occurs predominantly in glandular tissues and multiple lesions in different organs, it has been reported recently that the vascular system can also be affected. Visceral aneurysms of IgG4-related disease are uncommon, except for those in the aorta and coronary arteries [1-7].

A celiacomesenteric trunk, the common origin of the celiac trunk and the superior mesenteric artery (SMA) from the abdominal aorta, is not commonly encountered. As one might expect, IgG4-related aneurysms involving this arterial anomaly are also uncommon [8-10]. This report describes a case of IgG4-related celiacomesenteric trunk aneurysm treated using transcatheter arterial embolization.

CASE REPORT

Our institutional review board requires no approval for the publication of retrospective case reports. A 56-year-old man presented with progressive abdominal pain. He had neither a history of trauma nor evidence of arterial dysplasia or systemic disease. An aneurysm arising from the celiacomesenteric trunk with wall thickness and homogeneous wall enhancement at the late phase was found on contrast-enhanced computed tomography (CT) images. The aneurysm size, excluding the wall, was 18 mm x 6 mm at admission. Although no other IgG4-related lesion was found, his serum
Figure 1. A 56-year-old man with abdominal pain caused by IgG4-related celiacomesenteric trunk periarteritis. a, b, c) Contrast-enhanced computed tomography (CT) conducted 9 days after admission showed an aneurysm arising from the celiacomesenteric trunk, common hepatic artery of the distal portion (arrowhead), left gastric artery (arrow), and proximal splenic artery with wall thickness. d) Three-dimensional CT conducted immediately before transcatheter arterial embolization showed an aneurysm that arose from the trunk, common hepatic artery (arrow), and right hepatic artery. e) Lateral projection of celiacomesenteric arteriography revealed an aneurysm that arose from the trunk, common hepatic artery (white arrow), and splenic artery (white arrowhead), whereas the superior mesenteric artery (SMA) (black arrow) originated from the caudal side of the trunk. f) Anteroposterior projection of celiacomesenteric arteriography conducted immediately after transcatheter arterial embolization showed complete occlusion of the celiacomesenteric trunk aneurysm (arrow). It also provided visualization of the hepatic artery via the pancreatic arcade artery. g) A follow-up contrast-enhanced magnetic resonance image taken 2 years after transcatheter arterial embolization showed that the celiacomesenteric trunk aneurysm had disappeared and that SMA blood flow remained.
IgG4 concentration was high (161 mg/dl). A diagnosis of IgG4-related disease was made based on typical imaging features of periarteritis and a high serum IgG4 concentration. Although histopathological examination remains the gold standard for detecting organ involvement and diagnosis, in this case, it was difficult to obtain tissue by biopsy from the celiacomesenteric trunk. At 9 days after admission, the celiacomesenteric trunk aneurysm size had increased (16 mm × 10 mm excluding wall) with thinning of the wall (Fig. 1a-1d). Therefore, we decided to perform transcatheter arterial embolization for the celiacomesenteric trunk aneurysm because of the risk of rupture of the celiacomesenteric trunk aneurysm.

After a 4-Fr sheath was placed into the right femoral artery, a guiding catheter (FANSAC IV®, Terumo Clinical Supply Co., Ltd., Tokyo, Japan) was placed into the celiacomesenteric trunk. Selective celiacomesenteric arteriography revealed that the aneurysm rose from the trunk, that the common hepatic artery and splenic artery originated from the aneurysmal sac, and that the SMA originated from the caudal side of the trunk (Fig. 1e). The walls of the celiacomesenteric trunk aneurysm and the proximal portion of the common hepatic artery were irregular. A 2.1-Fr microcatheter (Sniper 2; Terumo Clinical Supply Co., Ltd.) was advanced into the splenic artery close to the celiacomesenteric trunk aneurysm via the celiacomesenteric trunk. After a microcatheter was inserted into the celiacomesenteric trunk, we inserted an additional 4-Fr guiding catheter (FANSAC IV®, Terumo Clinical Supply Co., Ltd.) in the SMA through the celiacomesenteric trunk to protect the SMA. This catheter made it possible to place the stent throughout if necessary. Detachable coils (two 6 mm × 15 cm coils, one 9 mm × 20 cm coil, one 10 mm × 30 cm coil, one 7 mm × 15 cm coil, and one 8 mm × 20 cm coil from GDC-18 360°; Stryker, Tokyo, Japan) were then delivered into the splenic artery. The common hepatic artery was embolized similarly (one 6 mm × 15 mm coil from GDC-18 360°; Stryker). The left gastric artery was visualized only to a slight degree using angiography. Therefore, transcatheter arterial embolization was not performed for the left gastric artery. Arteriographic observation through the celiacomesenteric trunk performed at this point revealed that blood flow from the aneurysmal sac to the common hepatic artery and splenic artery had ceased and revealed formation of the thrombus starting in the celiacomesenteric trunk aneurysm. Framing of the celiacomesenteric trunk aneurysm was attempted with detachable coils (one 18 mm × 40 cm coil and one 12mm × 30cm coil from GDC-18 360°; Stryker). Celiacomesenteric trunk arteriography applied immediately after transcatheter arterial embolization showed complete occlusion of the celiacomesenteric trunk aneurysm. It also provided visualization of the hepatic artery via the pancreatic arcade artery (Fig. 1f). At 3 weeks after transcatheter arterial embolization, fluorodeoxyglucose positron-emission tomography/CT was used for the patient for auxiliary diagnosis and future follow-up, revealing uptake of the celiacomesenteric trunk lesion. At 1 month after transcatheter arterial embolization, we started corticosteroid therapy (25 mg/day prednisolone) for IgG4-related disease. Although no liver infarction or bowel ischemia was observed during the clinical course, the patient had mild pancreatitis and partial splenic infarction, which were treated conservatively. The patient recovered. He was discharged 2 weeks after transcatheter arterial embolization. Follow-up contrast-enhanced magnetic resonance imaging conducted 2 years after transcatheter arterial embolization showed no celiacomesenteric trunk aneurysm, but the SMA blood flow remained (Fig. 1g). Although steroid therapy continues to the present day, the dosage has been reduced to 5 mg/day. No clinical symptoms associated with recurrent arteritis or other IgG4-related disease have been confirmed.

DISCUSSION

Recently, IgG4-related disease has been recognized as occurring in the cardiovascular system in the aorta and main branching arteries, such as celiac arteries, and in the superior mesenteric arteries. It often causes aneurysm and periarteritis [1-7]. Steroid therapy and surgical intervention have been reported, but no first-line treatment has been established for IgG4-related periarteritis [1, 5, 6]. Reportedly, the effects of steroid therapy on IgG4-related periarteritis are unsatisfactory for some patients [6, 7]. Additionally, Inoue et al. reported that steroid therapy might increase the risk of aneurysmal rupture by thinning of the arterial wall [1]. Rossi et al. reported transarterial liquid embolization for giant hepatic artery aneurysm associated with IgG4-related disease [2]. However, celiacomesenteric trunk periarteritis and aneurysm associated with IgG4-related disease are uncommon and/or they have been underreported.

Celiacomesenteric trunk is also uncommon, accounting for fewer than 1% of all splanchnic arterial anomalies. As one might expect, aneurysms involving this celiacomesenteric anomaly are not commonly encountered. Open surgery is the first-line treatment [8-10]. Nevertheless, few reports to date have described endovascular intervention.

For this case, we were hesitant about administering systemic steroid therapy before the procedure because we were unable to confirm that the condition resulted solely from IgG4-related disease and not from other diseases. Because of concern that steroid therapy might thin the aneurysmal wall, leading to the rupture of the celiacomesenteric trunk aneurysm, we chose a less invasive catheter treatment. The procedure was used to mitigate the risk of rupture and to facilitate treatment by steroid administration. In conclusion, our results show that transcatheter arterial embolization can be a useful therapeutic option not only to occlude the celiacomesenteric trunk aneurysm caused by IgG4-related periarteritis but also to safely support subsequent systemic steroid therapy.

CONFLICT OF INTEREST: The authors have no conflict of interest, financial or otherwise, related to this study.
**Acknowledgement:** This work was not supported by any grant.

**References**


