Inferior vena cava syndrome caused by retroperitoneal fibrosis after pelvic irradiation: A case report

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A 60-year-old-female who has history of pelvic irradiation developed severe leg edema. Radiological examination revealed obstruction of IVC with metallic stent placement, her symptom relieved.

1. Introduction

There are many causes of inferior vena cava (IVC) syndrome such as thrombosis, tumor thrombosis, iatrogenic occlusion caused by liver transplantation, congenital abnormality such as Budd-Chiari syndrome, compression by extravascular neoplasm, liver abscess, retroperitoneal fibrosis, or uterus during the third trimester of pregnancy (Harris, 1976). Retroperitoneal fibrosis (RPF) is categorized into an idiopathic type and secondary type. While idiopathic type accounts for majority of RPFs, secondary RPFs are caused by malignancy, medications, and prior history of irradiation (Tzou et al., 2014).

We describe here a case who received pelvic chemoradiation therapy for her locally advanced uterine cervical cancer and subsequently developed IVC syndrome one year after irradiation due to secondary RPF, and effectively treated with metallic stent placement.

2. Case presentation

A 60-year-old-female received definitive concurrent chemoradiotherapy for FIGO stage IB squamous cell uterine cervical cancer with only limited para-aortic lymph node metastasis being extra-pelvic lesion. Concurrent weekly cisplatin (40 mg/m²) and 45 Gy in 25 fractions of extended-field pelvic irradiation followed by 24 Gy in 4 fractions of image-guided adaptive brachytherapy and lymph node boost up to 56 Gy with external beam irradiation was performed. Two months after chemoradiation, follow-up image studies identified sacral bone fracture and single lung nodule. Four months after chemoradiation, PET-CT found only a single lung lesion and another pubic fracture, so the lung metastasis was treated by stereotactic body radiation therapy (SBRT) of 60 Gy in 10 fractions. Four months after lung SBRT, CT showed no progressive disease with left iliac bone fracture. Seven months after lung SBRT, CT found another pelvic bone fracture in right iliac bone and pubic bone. Two weeks after the CT, she developed severe bilateral lower leg edema which impacted her mobility necessitating a wheelchair (Fig. 1). One month after the development of severe bilateral lower leg edema, she developed abscess around pubic bone fracture with fever requiring hospitalization and antibiotics. Besides pelvic abscess, a radiologist pointed out suspicious IVC stenosis which may be the cause of the severe bilateral lower leg edema. Although no thickening around retroperitoneal great vessels was found on CT, with the presence of IVC and right ureter stenosis and past history of pelvic irradiation, it was clinically diagnosed that secondary RPF caused by pelvic irradiation was the cause of IVC and ureter stenosis. Subsequently, angiography was performed and IVC stenosis with collateral formation as well as right side moderate hydronephrosis was pointed out (Fig. 2a–c). Then, two spiral-Z stents (18/12 × 80 mm and 16 × 60 mm, Medico's Hirata inc., Osaka, Japan) were placed into the stenotic portion (Fig. 2d, e). Soon after stent placement, her symptom resolved quickly with no adverse event (Fig. 3). At the last visit, overall survival is 16 months after primary radiation therapy and no other recurrent disease was found other than solitary lung metastasis which was...
controlled by SBRT. No other radiation-related toxicity was reported other than pelvic bone fractures or RPF.

Written informed consent was obtained from the patient and this case report was approved by the Institutional Review Board of National Cancer Center Hospital (approved number is 2017-331) according to the ethical standards laid down in the Declaration of Helsinki.

3. Discussion and conclusions

There are many etiologies which can cause IVC obstruction. They are categorized into intravascular obstruction and extravascular compression. Thrombosis, tumor thrombosis, vascular catheters are included in the former group, while extravascular neoplasm, retroperitoneal fibrosis, liver disease with external pressure on the IVC, aortic aneurysm, or enlarged uterus during pregnancy are included in the latter group (Harris, 1976).

Unlike superior vena cava (SVC) obstruction, known as SVC syndrome which is one of well-known oncologic emergencies requiring prompt treatment, IVC obstruction generally does not recognized as oncologic emergency. The symptoms of IVC obstruction are depend upon the level of obstruction and they include intravascular hypovolemia, liver damage, kidney damage, edema of lower extremities, and ileus. In rare occasions, intravascular hypovolemia caused by IVC obstruction induces circulation collapse and result in shock, a life-threatening condition (Mohammed et al., 2018). The treatments of IVC obstruction are varied according to the cause of IVC obstruction. Obstruction of vena cava by extravascular compression from a malignant tumor can be treated by irradiation (Armstrong et al., 1987) or endovascular stent placement (Takeuchi et al., 2018).

RPF is originally reported by Ormond (Ormond, 1948) and can be grouped into two categories; idiopathic type and secondary type (Tzou et al., 2014). Idiopathic RPF accounts for the majority of cases and its pathogenesis remains unclear but some sort of inflammatory reaction has something to do with the fibrotic change and recently relation

Fig. 1. A photograph before IVC stent placement showing severe bilateral leg edema.

Fig. 2. a–c Infra vena cavograms before and after metallic stent placement. Obstruction of the infra vena cava at the lower part of infra vena cava was shown in Fig. 2a–c. Bilateral collateral blood flow was shown in Fig. 2a and b. When the tip of the guide wire penetrated the stenotic part, clear visualization of upper part of infra vena cava was obtained (Fig. 2e). Two spiral-Z stents were placed into the stenotic portion of IVC (Fig. 2d). Fig. 2e shows significant improvement of blood flow of IVC.
between idiopathic RPF and IgG4-related systemic disease have been reported (Stone, 2011). On the other hand, secondary RPF is caused by neoplasms, medications, and history of irradiation. Although evident soft tissue thickening around retroperitoneal great vessels was not found on CT in this case, because this patient had prior history of pelvic irradiation with extended field up to para-aortic lymph node area and this patient developed both IVC and ureter stenosis without disease recurrence in the retroperitoneum, it was clinically diagnosed that IVC obstruction was caused by secondary RPF induced by irradiation. The treatment for idiopathic RPF is systemic corticosteroids (van Bommel et al., 2007). However, because this case was not idiopathic case and no thickening of the retroperitoneal tissue was found, no steroid was applied and revascularization procedure using stent (Sato et al., 2012) was performed with quick symptom relief.

RPF caused by radiation is rarely reported (Majdoub et al., 2017), but IVC syndrome caused by radiation is not reported so far. This is, to the best of our knowledge, a first report of IVC syndrome caused by secondary retroperitoneal fibrosis after pelvic irradiation effectively salvaged by metallic stent placement.

Ethics approval and consent to participate and consent for publication

Written informed consent was obtained from the patient and this case report was approved by the Institutional Review Board of National Cancer Center Hospital (approved number is 2017-331) according to the ethical standards laid down in the Declaration of Helsinki.

Conflict of interests

Department of Radiation Oncology, National Cancer Center Hospital helps Pfizer with performing phase III clinical trial (Avelumab MSB0010718C) and receives fee according to the number of patients enrolled in the study.

Author contribution

NM wrote the main manuscript body. NM and YT are in charge of the presented patient and initially treated her cervical cancer with radiation therapy. YA performed stents insertion for infra vena cava syndrome. YT, KO, KT, KI, HI, YN, JI, and YA read the article and gave the first author suggestions to improve the manuscript.

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Fig. 3. A photograph one week after IVC stent placement shows almost complete resolving of leg edema.