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Transcatheter Embolization of a Ruptured Internal Pudendal Artery Pseudoaneurysm in a Patient with Neurofibromatosis Type 1

Neurofibromatosis type 1 (NF-1) is an autosomal dominant disorder. Pseudoaneurysms formation and rupture is an unusual complication of neurofibromatosis. To date, pseudoaneurysm of the internal pudendal artery associated with NF-1 has not been reported. In this article, we present a 62-yr-old man with NF-1 suffering from spontaneous hematoma of the perinea and scrotum. A digital substraction angiography disclosed a ruptured pseudoaneurysm of the right internal pudendal artery, which was successfully managed with transcatheter embolization.

Key Words : *Neurofibromatoses; Aneurysm, False; Embolization, Therapeutic*

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INTRODUCTION

NF-1, also called von Recklinghausen's disease, is an autosomal dominant disorder characterized by three major features: multiple neural tumors, café-au-lait spots, and pigmented iris hamartomas. Pseudoaneurysms formation and rupture is a rare complication of neurofibromatosis. To the best of our knowledge, the pseudoaneurysm of internal pudendal artery associated with NF-1 has never been reported in the literature. Here, we present a case of a spontaneous ruptured pseudoaneurysm of the right internal pudendal artery associated with NF-1, which was successfully managed with transcatheter embolization.

CASE REPORT

A 62-yr-old man was admitted to the emergency department because of a sudden pain and rapid enlarging mass in the perinea and scrotum regions. He had a history of NF-1 since childhood but denied any family history or traumatic event. His vital signs were normal. Physical examination revealed multiple neurofibromas all over the body, a large hematoma measuring about $8 \times 8 \times 6$ cm in the perinea and scrotum regions, and the extremely swollen penis (Fig. 1). The hematoma was soft, non-pulsating and had an undefined margin. Computed tomography (CT) of the abdomen and pelvis disclosed a large slightly high-density mass in the ischiorectal cavity, which extended to the perinea and scrotum regions. Routine blood samples on admission showed a red blood cell count of 4.45×10^{12}/L, hemoglobin concentration of 122 g/L, platelet of 236×10^{9}/L. He suffered from a recurrent and intolerable pain in the perineal region the next day. An emergent blood routine test showed a drop of red blood cell count to 3.08×10^{12}/L, hemoglobin to 84 g/L. However, his vital signs were stable, the blood pressure was 140/68 mmHg, and the heart rate was 84 beats/min.

Transfemoral bilateral external and internal iliac artery angiography revealed a ruptured pseudoaneurysm and active bleeding of the right internal pudendal artery. Then, a 3-french microcatheter (COOK, Bloomington, MN, USA) was navigated coaxially through a 5-french Cobra catheter (TERUMO, Tokyo, Japan) into the right internal pudendal artery. We tried to advance the microcatheter beyond the pseudoaneurysm so as to occlude both afferent and efferent vessel but failed. So, only afferent vessel just proximal to the pseudoaneurysm was embolized with two spiral coils (Cook) in dimension of 2×20 mm. Post-embolization bilateral internal iliac arteriogram showed obliteration of the pseudoaneurysm. After endovascular management, the patient was given conservative therapy because the clinical symptoms mitigated obviously with the hematoma getting softer and smaller the next day. He was transferred to a community hospital 23 days later for further therapies. There was no complication or recurrent hemorrhage during the next 6-months follow-up.

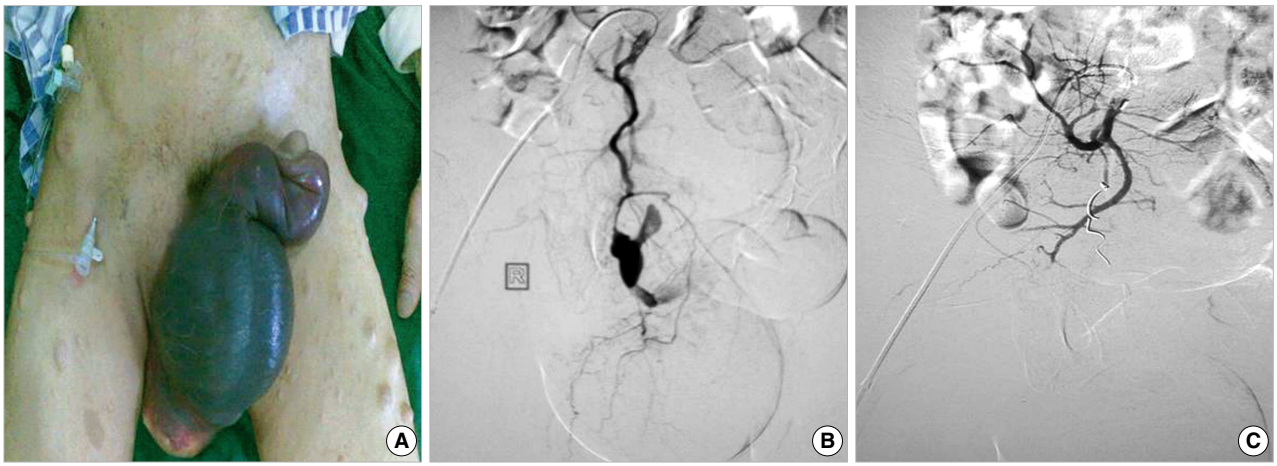

Fig. 1. A 62-yr-old man with a history of neurofibromatosis type 1. (A) Physical examination revealed multiple neurofibromas all over the body, a large hematoma on the perinea and scrotum regions, the extremely swelled penis and scrotum. (B) Right internal iliac arteriography revealed a ruptured pseudoaneurysm of the right internal pudendal artery. (C) Post-embolization right internal iliac arteriography showed complete occlusion of the pseudoaneurysm with coils.

DISCUSSION

NF-1, also called von Recklinghausen's disease, is an autosomal dominant disorder linked to chromosome 17q11.2 (1), characterized by growth impairment of the neural crest cells manifested by multiple neural tumors, cutaneous pigmentations, and Lisch nodules. The birth incidence of NF-1 lies between 1 in 2,500-3,300 and its prevalence in the population is 1 in 5,000 (2). However, 30-50% patients suffered from NF-1 has no family history. Sporadic case, as shown in our report, may arise from germ cell mutations (3).

It is well known that NF-1 can also involve the vascular tree, which may result in stenosis or occlusion of the vessel as well as arteriovenous malformation, arteriovenous fistula and aneurysm formation. Fibrous dysplasia or fibrosis of the vessel wall complicated with myocyte atrophy or sacculated aneurysm are its common pathological features. However, NF-1 is remarkable for its association with occlusive or aneurismal arterial disease affecting predominantly the renal arteries and less often the abdominal aorta, and mesenteric and peripheral arteries (4).

Aneurysm or pseudoaneurysm formation in NF-1 has been thought to be a result of friable vasculature secondary to arterial dysplasia or vascular invasion by the neurofibroma. The neurofibromatous tissue itself also has an abnormal vascular structure with thin-walled ecstatic blood vessels lying in loose neural stroma that replaces the normal adipose tissue. Many cases of aneurysm or pseudoaneurysm with NF-1 have been reported in the literature. The involved arteries include intrathoracic, gastrointestinal, extracranial arteries and so on (5-7). However, to the best of our knowledge, this is the first case of NF-1 with spontaneous ruptured pseudoaneurysm of the internal pudendal artery. As for aneurysms or pseudoaneurysms of NF-1, surgery as well as transcatheter embolization can be applied (4-7). But transcatheter embolization is preferred because surgical repair is aggressive and complex, and vessel reconstruction is limited by arterial fragility (8).

Obviously, excluding the pseudoaneurysm from the native circulation and preserving the parent artery is ideal in endovascular interventional procedures. However, it is uncommon and limited in these patients which associated with arteriole, just as shown in our case. For aneurysm, it is a general rule to occlude both proximal and distal ends of the parent vessel. We tried to occlude the distal end but failed. It is lucky the bleeding ceased. Embolization of the feeding arteriole is an efficient means to halt active bleeding, which can serve as a preoperative procedure or a radical cure management.

Embolization of the internal pudendal artery can cause sexual functional disturbance such as erectile dysfunction (9), because the penile arterial supply is derived from the internal pudendal arteries. However, sexual function kept intact in this case 6 months later, which may be explained by the affluent communication of the bilateral internal iliac artery.

In conclusion, we report here on the first documented case of a spontaneous ruptured pseudoaneurysm of the right internal pudendal artery associated with NF-1, which was successfully managed with embolization of the feeding internal pudendal artery. Endovascular techniques proved to be effective in the management of vascular complication of the NF-1.

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