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Case Report

Lumbar artery aneurysm: A rare manifestation of vasculopathy in a patient with neurofibromatosis type 1

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Abstract

Lumbar artery aneurysms are rare but important to diagnose, since rupture can have serious consequences due to life-threatening hemorrhage. A 49-year-old male with Neurofibromatosis type 1 (NF-1) was admitted to the emergency room with abrupt onset of severe abdominal pain. Ultrasound examination was normal, noncontrast CT revealed a tapered retroperitoneal mass adjacent to the right psoas muscle. A multiphasic contrast-enhanced CT scan raised suspicion of an arterial lumbar aneurysm and was confirmed by selective catheter based angiography. The patient underwent successful treatment with endovascular coil and the patient was discharged within a few days after an uneventful course.

Keywords: Lumbar artery aneurysm, Endovascular, Embolization, Neurofibromatosis type 1

Introduction

Vasculopathy is a well-recognized feature in neurofibromatosis type 1 (NF-1) and includes aneurysm formation, stenosis, and arteriovenous malformations. Multiple vascular lesions may be present and can occur in any anatomic region of the body [1]. Very few previous cases have reported lumbar artery aneurysms in patients with NF-1 [2–4]. Lumbar artery aneurysms are rare entities and often present as false aneurysms commonly associated with surgical procedures or trauma [2,5–7]. We present a rare case of a lumbar artery aneurysm in a patient with NF-1. Lumbar artery aneurysms may be incidental findings and diagnostic delay may occur due to ambiguous symptoms. However, some patients have a very acute clinical course due to rupture with retroperitoneal hemorrhage [5]. No well-defined correlation between aneurysm size and risk of rupture exists. Thus, management with repair and coil embolization of lumbar artery aneurysms is essential in order to prevent life-threatening hemorrhage. Attention to etiology, early identification, and prompt treatment is essential. Catheter-based treatment with coil-embolization is a minimally invasive, excellent treatment modality for medium-sized arteries [2].

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Fig. 1 – Lumbar artery aneurysm on MRI and PET-scan. a. Non-contrast MRI T2 fat saturated sequence revealing an inhomogenous tapered mass (arrow) adjacent to the right psoas muscle. b. PET-scan with FDG analogue showing peripheral uptake in the same retroperitoneal mass (arrow) found on MRI.

Case report

A 49-year-old man with NF-1, Crohn’s disease, and glucocorticoid-induced osteoporosis was admitted to the emergency department due to abrupt intensifying right upper quadrant abdominal pain radiating to the right shoulder blade and back. One week earlier, the patient had experienced intermittent light pain in the same anatomic region with no previous trauma. Crohn’s disease was well controlled for years, and the patient did not receive oral anticoagulant therapy. Clinical examination with deep palpation of the abdomen revealed distinct pain in the right upper quadrant and paravertebral region with no palpable mass and no abdominal

Fig. 2 – Contrast enhanced CT scan. a. Portal venous phase showing a lumbar artery aneurysm with mural thrombus (arrow). b. arterial phase showing the aneurysm (arrow) with an afferent (*) and efferent limb (**). c. portal venous phase, aneurysm (arrow) with clear view of the efferent limb (**). d. 3D reconstruction of the aneurysm (arrow).
Fig. 3 – Selective catheter-directed angiography. a. and b. Lumbar artery aneurysm (arrow) with an afferent (*) and efferent limb (**). c. Aneurysm (arrow) with coils in efferent limb (**). d. Angiography post embolization showing a completely thrombosed lumbar artery aneurysm with coils in the afferent (*) and efferent limb (**).

Guarding. Blood pressure was 143/93 mm Hg, pulse 105 bpm, and temperature 36.5°C. C-reactive protein was 19 mg/L and total leukocyte count was normal (7.69 10E9/L). Hemoglobin level was subnormal at 7.5 mmol/L. Laboratory tests showed normal liver and renal function. Urine analysis showed traces of blood.

Clinically, cholelithiasis and urolithiasis were suspected and an abdominal ultrasound examination revealed a morphologic normal liver and gall bladder and bile ducts without concrements. A noncontrast CT-scan of the abdomen excluded any urinary concrements. A 4 cm tapered retroperitoneal mass was found anterior and adjacent to the right psoas muscle. Providing high quality imaging of soft tissue, a noncontrast MRI T2 fat saturated sequence of the abdomen revealed a rounded retroperitoneal, inhomogeneous mass. (Fig. 1a) A subsequent PET-scan with an injected FDG-analogue revealed some peripheral FDG-uptake. (Fig. 1b) Tentative diagnoses proposed from MRI and PET were malignant neoplasia, necrotic lymph node, and abscess formation.

In the end, a CT-scan in arterial and venous phase was performed. (Fig. 2) The lesion displayed partial contrast enhancement with signs of an afferent and efferent limb. Thus, a vascular etiology was suspected and selective catheter-based angiography subsequently confirmed a fusiform lumbar artery aneurysm. (Fig. 3)

Angiography was performed via access in the left femoral artery. A 5F, 45 cm sheath (Terumo Medical Corp) was introduced and placed distal to the origin of the second right lumbar artery with the aneurysm. Selective catheterization of the artery was conducted with a 4F, SHK 65 cm guiding catheter, (Cordis). A 2,7F micro catheter, (Progreat, Terumo Medical Corp) was passed through the guiding catheter crossing the aneurysmal sac to the efferent branch of the aneurysm and several pushable microcoils 3-, 5- and 7 mm (Trufill, Cordis) were placed there. Subsequently pulling back the catheter, the
afferent part of the lumbar artery was embolized with more 7-, 5- and 3 mm microcoils. Finally, 2 detachable coils, 4 mm, 8 cm (Concerto, Medtronic) were placed in the artery close to the origin of the aorta. This “sandwich coiling” excluded the aneurysm. Thus, a subsequent aortogram demonstrated no filling of the aneurysm.

The patient received a CT scan after 6 months, which revealed shrinkage of the aneurysm from 4 cm to 1.8 cm, with no signs of flow.

Discussion

This case illustrates a rare entity of a lumbar artery aneurysm in a patient diagnosed with NF-1. Lumbar artery aneurysms may cause a diagnostic pitfall and when associated with ambiguous symptoms and hence diagnostic delay. Lumbar artery aneurysms are often modest in size and located deep in the abdomen. Physical examination has obvious limitations. In comparison, abdominal palpation by a physician has a sensitivity of 76% detecting an aortic abdominal aneurysm ≥ 5 cm [8]. Ultrasound is operator-dependent, obesity and overlying bowel gas can easily obscure visualization of retroperitoneal vessels and reduce sensitivity. Thus, ultrasound plays a minor role in detecting and characterizing deep abdominal vascular lesions [9]. CT is readily available and provides detailed visualization of the aorta and its branches. A CT scan can be performed quickly and patients with suspected disease in the aorta and its branches should undergo CT angiography for diagnosis confirmation [10]. In the case of aneurysmal disease, it provides multiplanar measurements of aneurysm size and form. On noncontrast CT, a lumbar artery aneurysm presents as a soft tissue density mass as in this case located on the anterior psoas muscle [5]. Noncontrast CT is helpful in detecting calcifications, inflammation and fat-stranding, and is able to some extent to differentiate between fat and other components of an unknown mass. Single arterial phase CT is the most important investigation and provides excellent overview of an aneurysm with its feeding vessels and thrombosis information. Importantly, CT can detect or rule out multiple vascular lesions in patients with NF-1. Occasionally, arterial aneurysms are detected as incidental findings on portal venous phase CT scans and this is often adequate to verify the diagnosis. Further, CT is useful in an acute setting and planning treatment [5].

Direct catheter-based angiography allows for assessment of dynamic filling of the aneurysm thereby providing excellent visualization of feeding vessels and allows for precise placement of endovascular coils in order to achieve successful treatment. However, it carries the risk of arterial puncture, muscle or peripheral nerve infarction and serious infarction of the spinal cord and should be limited to patients where treatment is indicated, or when CT is inconclusive [11]. Even if intravenous contrast is not tolerated in patients and becomes a risk factor for developing postcontrast acute kidney injury, this case is still informative [12].

Most lumbar artery aneurysms are false aneurysms. They are rare entities and pseudoaneurysms carry a high risk of rupture with significant mortality. In this case, it is unclear whether this patient suffers from a true or false aneurysm. However, this patient was suspected of having a false aneurysm irrespective of previous history and treated accordingly. Correlation between aneurysms size and risk of rupture is uncertain. A threshold of 2 cm has been proposed for visceral arterial aneurysms [13]. Symptomatic aneurysms should prompt treatment in order to decrease morbidity and mortality. Patient history and comorbidity should also be considered [14,15].

Endovascular coil-embolization has been performed successfully in a patient with NF-1 suffering from a ruptured lumbar artery aneurysm [3]. This approach is minimally invasive, effective and safe in the management of lumbar artery aneurysms with obvious advantages in accessing a surgically challenging anatomic region. It can be done in local anesthesia and eliminates problems encountered using general anesthesia in patients with NF-1 [16].

Open or conventional surgery has been performed, but with a poor outcome – this has mainly been explained as related to the location of lumbar artery aneurysms in a hardly accessible anatomic region and a difficult, narrow operating field and blood loss [2,6]. Procedures involving the use of general anesthesia exposes patients with NF-1 to a higher risk compared to otherwise healthy patients. Neurofibromas may also be found in the oropharynx and larynx and potentially produce difficulties with laryngoscopy and tracheal intubation. Patients with NF-1 may as well suffer from pulmonary fibrosis as well as other cardiovascular manifestations increasing the risk of general anesthesia [16].

Percutaneous embolization of a lumbar pseudo-aneurysm has been performed with success in a patient with type IV Ehlers-Danlos syndrome and in a patient with an iatrogenic lumbar pseudo-aneurysm following renal biopsy [17,18]. Ultrasonic guided compression has been performed successfully in 1 case [19]. However, the aneurysm was already largely thrombosed at the time of treatment.

In conclusion, endovascular coil-embolization of lumbar artery aneurysms seems to provide an effective and safe treatment option in patients with von Recklinghausen disease.

Compliance with ethical standards

Ethical approval

No IRB existed at the time the study was initiated. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent

Informed consent was obtained from all individual participants included in the study.

Consent for publication

Consent for publication was obtained for every individual person’s data included in the study.
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