Case Report

Bilateral Radial Artery Aneurysms in the Anatomical Snuff Box Seen in Marfan Syndrome Patient: Case Report and Literature Review

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We describe the first Marfan syndrome case of non-traumatic bilateral radial artery aneurysms in the anatomical snuff box. A 74-year-old woman with Marfan syndrome had a pulsatile mass in her bilateral anatomical snuff box. The color Doppler ultrasonography showed an aneurysm of radial artery located in the bilateral anatomical snuff box. Resection of the right radial artery aneurysm was completed without complications. Histopathological analysis showed a true aneurysm with atherosclerotic changes in the arterial wall. We review the literature on non-traumatic or bilateral radial artery aneurysm in the anatomical snuff box, and discuss the clinical presentation and surgical management.

Key words: radial artery aneurysm, anatomical snuff box, Marfan syndrome

INTRODUCTION

Radial artery aneurysms are rare, usually being caused by accidental or iatrogenic penetrating trauma. Most of the reported cases occurred after trauma at the level of the wrist.1, 2) The incidence of radial artery aneurysm in the anatomical snuff box is very small and very few cases have been reported.3-9) Furthermore, bilateral radial artery aneurysms were also extremely rare.10-12) On the other hand, as far as we know, radial artery aneurysm in the patient with Marfan syndrome has not been reported. We describe the first case of non-traumatic bilateral radial artery aneurysms seen in Marfan syndrome patient.

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CASE REPORT

A 74-year-old woman was referred to our department for a pulsatile mass with slight pain in the right anatomical snuff box (Fig. 1). She underwent Bentall’s operation for annulo-aortic ectasia (AAE) and aortic regurgitation (AR) when she was 53 years old. At the time of operation, she was diagnosed as Marfan syndrome. She was a housewife and there was no history of trauma or previous infections. On her medical interview, the mass in the right anatomical snuff box had enlarged and became increasingly tender with accompanying pain. On physical examination, a pulsatile mass in the left snuff box was also revealed. Bilateral pulsatile masses induced no physical signs or clinical symptoms, such as bruit, thrill, or numbness, except slight pain in the right anatomical snuff box. The color Doppler ultrasonography showed an aneurysm of radial artery located in the bilateral anatomical snuff box (Fig. 2). The right radial artery aneurysm was a saccular shape with a wide neck. Aneurysm of the left radial artery was a fusiform aneurysm. No laminar clot was detected in either aneurysm. The maximum diameter of the right and left radial artery aneurysm was 9 mm and 5 mm, respectively.

Considering the likelihood of rupture or thrombo-em-
the distal vessel. Then, the distal and proximal vessels of the aneurysm were ligated and the aneurysm was excised (Fig. 3). After surgery, the motility and sensibility of the thumb and all other four digits remained normal. The patient was discharged on the day after surgery without any complications. Regarding the left radial artery aneurysm, careful observation was recommended because of less possibility of rupture and lack of clinical symptom. Histological section confirmed the presence of a true

Fig. 1 Arrow indicates the pulsatile mass in the right anatomical snuff box.

Fig. 2 Ultrasonography of the right (A, C) and left (B, D) radial artery aneurysm in the anatomical snuff box.
atherosclerotic aneurysm with thrombus (Fig. 4). The obvious findings that were characteristic of Marfan syndrome such as cystic medionecrosis were not recognized histologically.

**DISCUSSION**

Aneurysmal formation in the radial artery is rare. Most of the reported cases occurred after trauma or arterial catheterization at the level of the wrist. Therefore, most radial artery aneurysms are found in the unilateral wrist. Only 3 cases of the bilateral radial artery aneurysms were reported previously. And only 10 cases of the radial artery aneurysm in the anatomical snuff box have been described in 7 publications. Causes of aneurysmal formation in the anatomical snuff box were trauma in 3, mycotic in 2, and idiopathic in 5 cases. Trauma including accidental, malicious, or iatrogenic is a common cause of the radial artery aneurysm. However, a careful medical interview excluded a traumatic or iatrogenic injury as an etiologic mechanism in this case. The first case of a non-traumatic radial artery aneurysm was reported by Thorren et al. in 1966, and since then 13 publications on the subject have appeared. In those cases of non-traumatic aneurysm, some concomitant systemic or local disorders were reported as follows, one case of Behcet’s dis-
We described the first case of non-traumatic bilateral radial artery aneurysms in the anatomical snuff box seen in Marfan syndrome patient.

REFERENCES


Conclusion

We described the first case of non-traumatic bilateral radial artery aneurysms in the anatomical snuff box seen in Marfan syndrome patient.

Our patient was diagnosed as Marfan syndrome 21 years ago at the time of Bentall's operation for AAE and AR. We could also diagnose retrospectively her as Marfan syndrome according to the Ghent criteria. Peripheral artery aneurysm, however, is an unusual manifestation of Marfan syndrome. Especially, aneurysm of distal to the subclavian artery is extremely rare and reported only in 3 cases, as 2 of the axillary-subclavian artery aneurysm and the accompanying pain. Regarding the left radial artery aneurysm compounding atherosclerotic arterial disease. The surgical procedure, excision alone7) or reconstruction,8) is dependent on the patency of collateral circle. In our case, we confirmed the safety of ligation by intra-operative direct clamp test of the radial artery using pulse oximeter in addition to the preoperative Allen's test.

The obvious findings that were characteristic of Marfan syndrome such as cystic medionecrosis were not recognized histologically. In the report of the axillary-subclavian artery aneurysm seen in Marfan syndrome, cystic medionecrosis was confirmed by histopathological examination. But it was not confirmed in the ulnar artery aneurysm case. Because the medial smooth muscle cell layer of the peripheral artery, such as the radial or ulnar artery, is thinner than that of the aorta which is usual manifestations of the Marfan syndrome. It is difficult to prove it clearly as an aneurysm related to the Marfan syndrome.

Regarding the treatment of the aneurysms in literature, observation alone,3) compression with bandage,25) ultrasound-guided embolization26) and surgical intervention were reported. According to the previous reports, operative indication depends on the possibility of traumatic rupture and distal ischemia due to a chronic damage of the vascular supply, embolism from a laminar clot, distal extension of a thrombus, or ischemic effects of the aneurysm compounding atherosclerotic arterial disease. We decided to make a surgical intervention to the right radial artery aneurysm because of the shape of aneurysm and the accompanying pain. Regarding the left radial artery aneurysm, we recommended careful observation because of less possibility of rupture and lack of symptom. The surgical procedure, excision alone25) or reconstruction,27) is dependent on the patency of collateral circle. In our case, we confirmed the safety of ligation by intra-operative direct clamp test of the radial artery using pulse oximeter in addition to the preoperative Allen's test.