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Aneurysm of the Transverse Cervical Artery Occurring in Association with a Cavernous Hemangioma as a Complication of Klippel-Trénaunay Syndrome: Report of a Case

KUNIHIDE NAKAMURA,[1] TOSHIO ONITSUKA,[1] YASUNORI KOGA,[1] KOICHIRO SHIBATA,[1] YUICHI TSUCHIDA,[1] KAZUKI NABESHIMA,[2] and AKINOBU SUMIYOSHI[3]

[1] Second Department of Surgery, [2] Second Department of Pathology, and [3] First Department of Pathology, Miyazaki Medical College, Kiyotake, Miyazaki, 889-16 Japan

Abstract: We report herein the case of a 14-year-old girl with Klippel-Trénaunay syndrome who developed an aneurysm of the transverse cervical artery. Because it was continuing to increase in size, with an associated risk of rupture, an aneurysmectomy was performed. Pathological examination of the resected specimen revealed a cavernous hemangioma located near the aneurysm. To our knowledge no other case of an aneurysm occurring in association with a cavernous hemangioma as a complication of Klippel-Trénaunay syndrome has ever been reported.

Key Words: aneurysm, cavernous hemangioma, Klippel-Trénaunay syndrome

Introduction

In 1900, Klippel and Trénaunay[1] first described the classic triad of cutaneous hemangiomata, hemihypertrophy, and venous varicosities. In 1907, Weber[2] reported a similar triad and additionally described the signs of arteriovenous (A-V) fistulae. In a subsequent paper published in 1918,[3] Weber reported arteriovenous fistulae as an additional feature of the syndrome, and various names have since been used to describe this syndrome. In fact, attempts have been made to relegate the name "Parkes-Weber syndrome" to those patients with findings of arteriovenous fistulae, and to designate the remainder as having "Klippel-Trénaunay syndrome."[4] The etiology of the Klippel-Trénaunay syndrome remains obscure, but a mesodermal defect during fetal development could explain all of its features.[4] It appears that venous angiodysplasia is the central defect despite the potential polyvalent angiologic picture.[5] Atresia, hypoplasia, or obstruction of the deep venous system have been noted in almost all cases of Klippel-Trénaunay syndrome. Other vascular abnormalities, particularly cavernous hemangioma, have been reported in a variety of organs including the colon,[6] kidney,[7,8] and urinary tract.[9] However, arterial abnormalities are not commonly found in patients with Klippel-Trénaunay syndrome. We describe herein the case of a young woman with Klippel-Trénaunay syndrome who presented with a pulsatile supraclavicular tumor found to be a transverse cervical artery aneurysm associated with a cavernous hemangioma.

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Case Report

A 14-year-old girl was referred to our clinic on March 13, 1989, for investigation of a pulsatile tumor in the right supraclavicular fossa that had been present since September, 1988. Although the tumor had been increasing in size it was not painful. Her past medical and family histories were unremarkable and there was no history of trauma. On examination, her right arm was purple and slightly longer than the left arm, and the skin of her right arm was atrophic. She also had large persistent embryonic veins and superficial varicosities on the right shoulder which had been present since the age of 8. A computed tomography (CT) scan revealed the tumor to be 32 mm in diameter on cross-section. Angiography subsequently confirmed that the tumor was an aneurysm of the transverse cervical artery (Fig. 1a), while venous pooling around the aneurysm on late phase angiography demonstrated an adjacent cavernous hemangioma (Fig. 1b). There was no predominant feeding artery, draining vein, or arteriovenous shunt, nor was there any evidence of a functioning arteriovenous fistula on physical examination, arteriography, or venous blood oxygen saturation measurements.

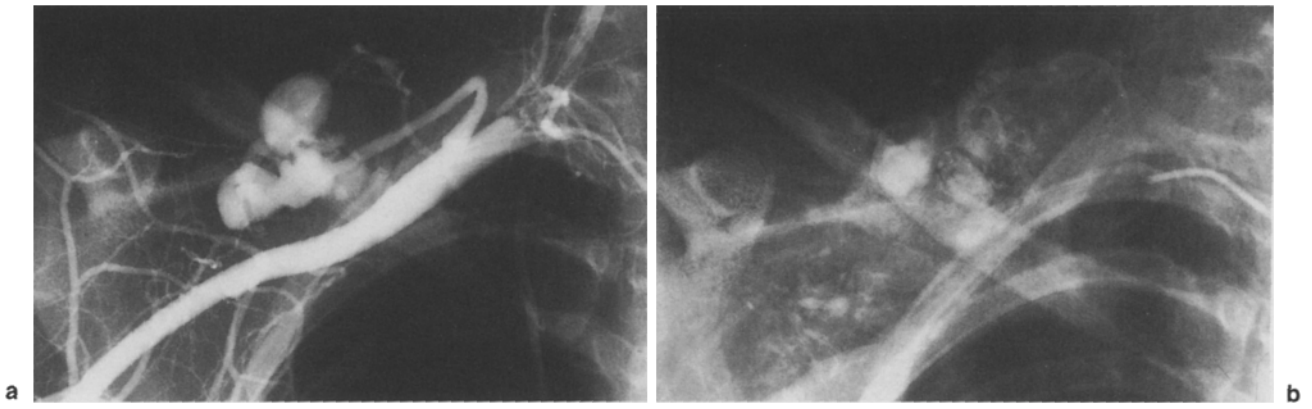

Fig. 1. a Arteriography revealing an aneurysm of the transcervical artery measuring 32 mm in diameter. **b** Late phase of the arteriography demonstrating a cavernous hemangioma around the transcervical artery aneurysm

Because the aneurysm was increasing in size, with the associated risk of rupture, resection was performed on May 24, 1989. There were no intraoperative complications and her postoperative course was uneventful. The patient was discharged from hospital shortly thereafter and has not shown any evidence of recurrence in the subsequent 4 years.

The aneurysm had an irregular macroscopic appearance (Fig. 2). Microscopically, the aneurysm wall lacked an intima, internal elastic lamina, and the smooth muscle layers of the media. It consisted predominantly of thin collageneous connective tissue containing large cavernous vessels (Fig. 3A). The wall was thin enough in some areas to expose the cavernous vessels to fibrin thrombi (Fig. 3B). The nonaneurysmal portion of the resected artery also contained these angiomatous vessels which interrupted the medial smooth muscle at irregular intervals (Fig. 3C). The walls of these vessels were irregularly thickened by adventitial fibrosis. The angiomatous vessels were seen both outside (Fig. 3B) and inside (Fig. 3D) the aneurysm wall. These findings suggested that the cavernous hemangioma, which was found in the soft tissue adjacent to the arterial wall, extended into the vessels, destroying their normal architecture and resulting in aneurysmal formation.

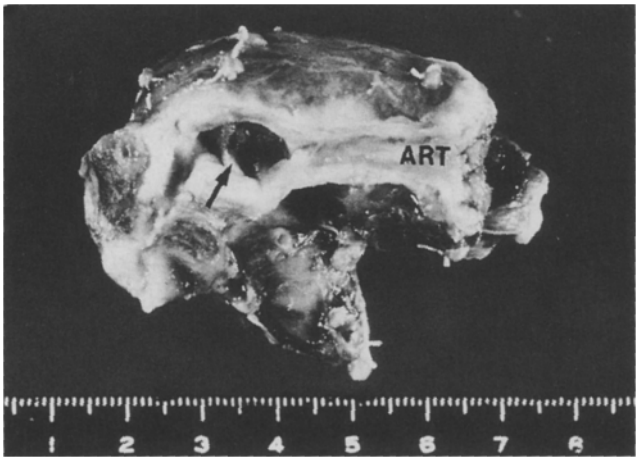

Fig. 2. Photograph of the aneurysm after excision. The transcervical artery (*ART*) was opened longitudinally to expose the aneurysm (*arrow*)

Discussion

The classic triad of (1) a cutaneous vascular nevus, or port-wine stain hemangioma, (2) varicose veins, and (3) hemihypertrophy was first described in 1900 by Klippel and Trénaunay.[1] Since 1907 and 1918, when Weber independently reported a similar syndrome which also included the presence of arteriovenous fistulae,[2,3] the Klippel-Trénaunay syndrome has been used to describe patients without arteriovenous fistulae.

The most common form of hemangioma is the capillary type, being the port-wine nevus which is pink to purple in color and represents diffuse telangiectasias of the superficial vessels of the dermis.[10] Cavernous hemangiomata or lymphangiomata may also be found and in rare cases, hemangiomatous involvement of the internal organs can lead to severe complications.[6] In fact, rectal bleeding and hematuria have been reported as a result of pelvic, urinary tract, and abdominal hemangiomata.[7,9] Campistol et al.[8] reported a case of renal hemangioma and renal artery aneurysm occurring in association with Klippel-Trénaunay syndrome. In that case, the renal artery showed a saccular aneurysm with alteration of the artery wall. In our patient, a hemangioma in the soft tissue of the supraclavicular region invaded the transverse cervical artery, resulting in subsequent aneurysm formation.

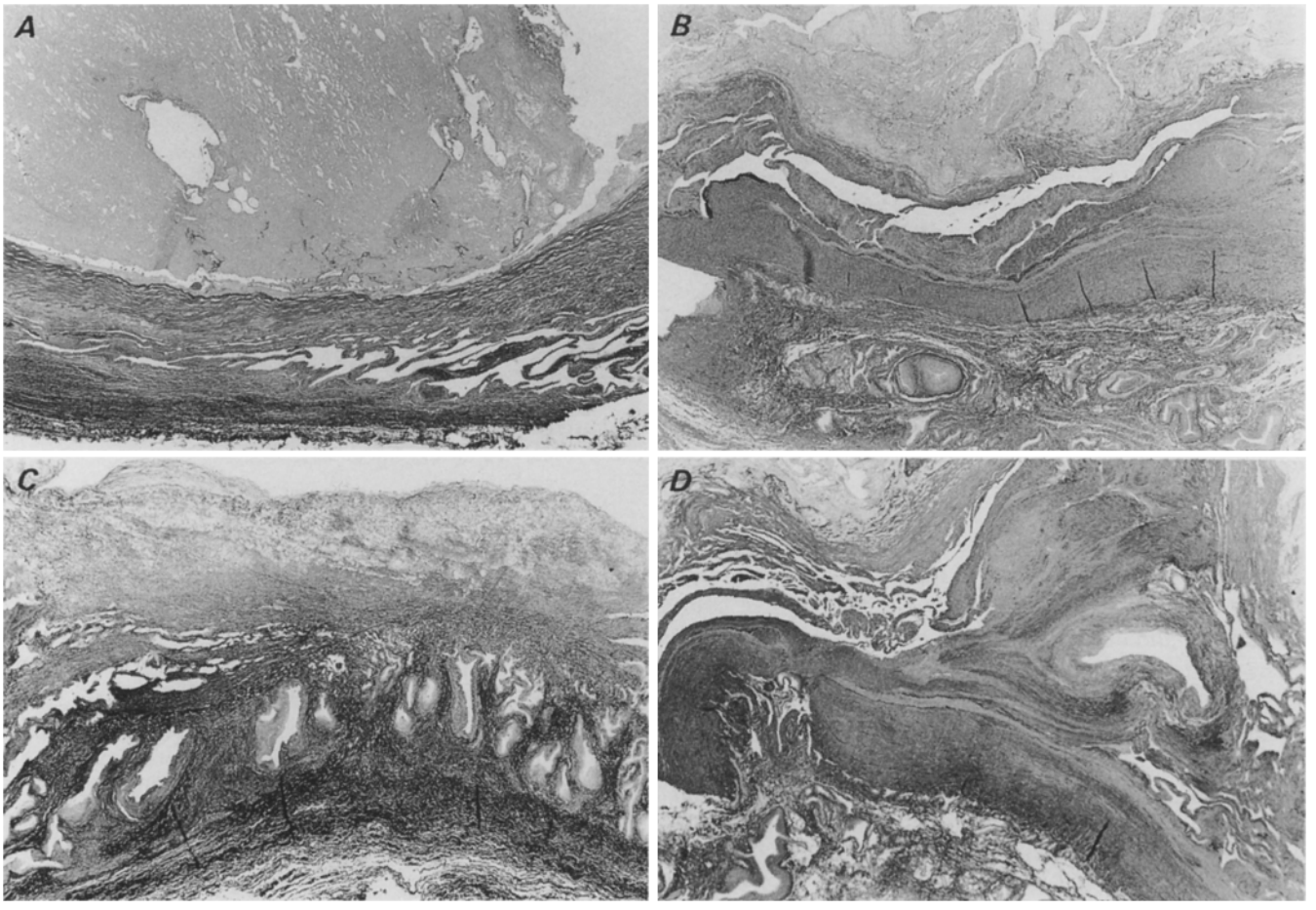

Fig. 3A–D. Microscopic examination of the aneurysm wall (Elastica van Gieson, original magnification ×15). **A** Aneurysm wall with large angiomatous vessels. The wall is thin without any smooth muscle layers of the media. A fibrin thrombus can be seen in the lumen. **B** Aneurysm wall with nearly ruptured angiomatous vessels. Angiomatous vessels can also be seen outside the aneurysm wall. **C** Nonaneurysmal portion of the arterial wall with angiomatous vessels. The angiomatous vessels show adventitial fibrosis and irregularly spaced medial smooth muscle bundles. **D** Aneurysm wall with penetrating angiomatous vessels. The adventitia is completely interrupted by the angiomatous vessels, which seem to be connected to those outside the aneurysm wall

Shobinger et al.[5] reported that Klippel-Trénaunay syndrome is a silent dysplasia, eminently venous in character and behavior. He divided the Klippel-Trénaunay syndrome into four types according to the vascular polyvalence:

1. That without A-V shunts or anomalies of the deep venous system
2. That with anomalies of the deep veins
3. That with inactive A-V shunts
4. That with lymphatic involvement

Lie[11] described that the arterial changes consisted of nonspecific, bland, fibrous intimal proliferation, with medial calcification. The most common form of venous dysplasia had similar characteristics.

The absence of an A-V fistula in our patient makes the hemihypertrophy of the arm difficult to explain. However, Servelle[12] reported that venous stasis is responsible for the edema and elongation of the limb. Baskerville et al.[13] conducted a study of blood flow in the calves of patients with Klippel-Trénaunay syndrome using venous occlusion plethysmography and demonstrated normal flows. They[13] concluded that Klippel-Trénaunay sundrome is caused by a mesodermal abnormality during fetal development, leading to the maintenance of microscopic arteriovenous communications in the limb bud, which may result in hemihypertrophy. Lie[11] supported this interpretation with anatomic studies.

In this article, we described an arterial aneurysm which occurred in a patient with Klippel-Trénaunay syndrome. According to the literature, the majority of peripheral arterial aneurysms resulted from arterial trauma. Less frequently, the etiology is mycotic or necrotizing arteritis, or arteriosclerosis.[14] In Japan, arteriosclerotic peripheral aneurysms occur less fre-

quently than in Europe and the USA.[15] Furthermore, congenital arterial aneurysms of the upper extremity are rare and have seldom been reported in infancy and childhood.[16] In this patient, there was no evidence of true vasculitis or atherosclerosis, and her history excluded the possibility of a posttraumatic or mycotic aneurysm. This, in combination with the microscopic findings, suggests that the aneurysm was caused by direct extension of the cavernous hemangioma. To our knowledge, there is no other report of an aneurysm occurring in association with a hemangiomata in a patient with Klippel-Trénaunay syndrome. It is therefore unlikely that this aneurysm developed as a result of a mesodermal defect during fetal development.

In conclusion, surgery may be indicated for lesions that show: (a) an atypically rapid growth rate, for example, tripling in size within a few weeks; (b) Kasabach-Merritt syndrome; or (c) encroachment of the lesion into vital tissues.[17] The third indication was applicable to our patient. On the other hand, patients with minimal varicosities, in whom the hemangioma is a port-wine superficial nevus with hemihypertrophy under 1 cm, usually do well without surgical treatment.[10]

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