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30.1 Syphilitic Aneurysms

Syphilis was considered an important and frequent cause of arterial aneurysms. In developed countries, after the introduction of penicillin, its role steadily declined [1]. However, syphilis and its cardiovascular sequelae remain a relevant sanitary problem in several countries, especially where HIV infection is largely diffused [2]. Cases of syphilitic aneurysms of the aorta, both thoracic and abdominal, are still reported [3–5].

As for popliteal aneurysms, sporadic co-existence of syphilis is mentioned in some large series reported in the second half of the last century (Table 30.1), but, in general, the role of the infectious disease in the genesis of the aneurysm is poorly defined. As a matter of fact, in 1934, Barker [22], reviewing 13 cases of popliteal aneurysm reported as luetic, observed that only in one case the clinical diagnosis was supported by pathology findings and concluded that the presence of aneurysm in comparatively young men with strongly positive Wasserman reaction does not mean that the aneurysm is luetic. In 1952, Silver and Kahn [23], reporting a case of popliteal aneurysm probably luetic (the patient was a 60-year-old black man successfully treated by proximal and distal ligation and subtotal

Table 30.1 Incidence of syphilis in patients with popliteal aneurysm

Author, year	N patients	Study period	Syphilis
Gifford [6], 1953	69	1913–1951	6
Edmunds [7], 1965 ^a	82	1948–1963	1
Baird [8], 1966	46	1938–1964	1
Wychulis [9], 1970	152	1960–1968	1
Gaylis [10], 1974	43	1957–1972	0
Bouhoutsos [11], 1974	84		2
Hardy [12], 1975	23	18 years	0
Towne [13], 1976	80	1950–1975	1
Chitwood [14], 1978	26		1
Inahara [15], 1978 ^b	30	1963–1977	0
Szilagyí [16], 1981	62	1964–1979	0
Vermilion [17], 1981	87	1960–1980	1
Whitehouse [18], 1983	61	1943–1982	0
Reilly [19], 1983	159	25 years	0
Mangiante [20], 1984	34	1950–1980	0
Farina [21], 1989	36	15 years	0

^aCases of primary amputation excluded

^bOnly patients treated surgically

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resection of the aneurysm), assessed that it was almost impossible to find in the literature a well-documented case of syphilitic aneurysm of the popliteal artery.

In 1958, Rajam and Rangiah [24] reported a case of luetic popliteal aneurysm spontaneously cured having undergone thrombosis during specific anti-luetic treatment; they assessed that in tropical areas a great number of arterial aneurysms are luetic, because of a large reservoir of undiscovered or untreated syphilis; accordingly, they diagnosed as luetic five of the seven popliteal aneurysms observed at the Government Hospital of Madras from 1947 through 1956.

In 1974, Spay et al. [25] diagnosed as syphilitic the popliteal aneurysm of a 33-year-old man living in Dakar (treated by subtotal resection and vein grafting) warning about the difficult interpretation of serologic data in patients living in tropical and subtropical regions due to the multiple treponemic etiologies (venereal syphilis, endemic syphilis, pian, or framboesia).

30.2 Idiopathic Popliteal Aneurysms in Children

Popliteal aneurysms in the pediatric age are rare, being usually due to complex genetic disorders when not deriving from trauma or arthritis or infection. “Idiopathic” aneurysms are extremely rare and may be isolated [26] or associated with multiple aneurysms in the context of the so-called syndrome of idiopathic childhood aneurysms [27]. The latter is suspected to be the expression of a congenital disease apparently different from Ehlers–Danlos syndrome [28], representing a potentially lethal condition owing to the risk of rupture. According to Short [29], the noxious agent causing the outbreak of multiple and successive aneurysms could lose activity in the adolescence, but existing aneurysms would continue to enlarge eventually undergoing complications. The finding of an aneurysm in a child should prompt a complete imaging investigation of the entire vascular system [30].

Sterpetti et al. [31] proposed to classify congenital aneurysms into two large groups:

Table 30.2 Idiopathic popliteal artery aneurysm in childhood

Author, year	Patient	Clinical picture
Short [29], 1978	F, 7 years	Multiple aneurysms: abdominal aortic, renal, upper and lower limbs ^a
Schiller [33], 1983	M, 8 years	Multiple aneurysms: abdominal aortic, iliac, renal, upper and lower limbs
Bordeaux [34], 1990	F, 7 years	Multiple aneurysms: abdominal aortic, iliac, renal, upper and lower limbs ^b
Hurley [26], 1994	F, 14 years	Isolated popliteal aneurysm
Sheppard [27], 2000	M, 5 years	Multiple aneurysms: abdominal aortic, renal, sup. mesenteric, iliac, upper and lower limbs ^c
Lopez-Gutierrez [35], 2012	<1 year	Lower limb hypoplasia, multiple aneurysms: iliac, femoral, popliteal ^d

^aBilateral popliteal aneurysm

^bReported an associated “inflammatory syndrome”

^cAneurysms at knee level, non-specified if popliteal or branches

^dTruncal arterial malformation, persisting sciatic artery

- Aneurysms (usually multiple) due to generalized disorders of the arterial tissue
- Aneurysms (usually isolated) deriving from a localized defect of the arterial wall

Sarkar et al. [32] proposed a detailed classification, according to the different etiologies (relating to a genetic or acquired underlying disease) into nine classes, of which one consisting on idiopathic aneurysms.

In Table 30.2, the relevant data of some reported cases are illustrated.

30.3 Obscure Cases

In 2011, Akamatsu et al. [36] reported a case of popliteal aneurysm in which CT scan put into evidence characteristics quite similar to those commonly observed in inflammatory aneurysms of the abdominal aorta. The patient, a 67-year-old man, was affected with unresectable pancreatic cancer; he complained of fever and laboratory

tests showed mild leukocytosis (89% neutrophils) and high levels of C-reactive protein. The patient was treated with partial resection of the aneurysm and autologous vein grafting; post-operatively, laboratory findings returned to normal (the patient died after 8 months from pancreatic cancer). Tissue cultures and microscopy findings were negative for bacteria, fungi, and viruses. Microscopy showed marked transmural inflammation with thickening of the media and adventitia.

The differences with inflammatory aortic aneurysms were:

- Macroscopically: absence of the typical white glistening aspect of the inner surface.
- Microscopically: presence of a marked neutrophil infiltration.

Mellièrè et al. [37] observed five cases of popliteal aneurysm defined as idiopathic and presumed congenital in non-pediatric patients. Pathology study is available only in two patients; in the other three the aneurysm was left untouched and bypassed or surgery was not performed. A 20-year-old man was submitted to aneurysm resection and autologous vein grafting (ok after 2 years): histologically, the arterial wall was diffusely fibrotic, with few remnants of the internal elastic lamina and extensive lymphocytic infiltration of the adventitia. A woman aged 32 (the case was the object of an earlier report [38]) was submitted to aneurysm resection and autologous vein grafting with excellent result at 15-year follow-up: microscopy showed diffuse sclerosis of the arterial wall with perianeurysmal infiltration of lymphocytes and histiocytes frequently multinucleated.

In lack of any etiological definition, the authors propose to consider these aneurysms as congenital and suggest that the cause may be identified in some defect during the complex embryologic arrangement of the popliteal artery.

Of course, in these cases (as in many other fields of medicine) one should always consider that the term idiopathic means substantially a lack either of knowledge or understanding about the etiology. This applies also to the rare cases

reported as idiopathic popliteal pseudoaneurysms [39].

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