The "Problem" of Arteriomegaly

9

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In 1942–1943, Leriche [1, 2] reported on two patients with extraordinary elongation and dilatation of the pelvic and common femoral arteries, which, before operation, were misdiagnosed as aneurysmatic. He textually assessed:

> "La possibilité pour les artères de s'allonger et se dilater globalement de façon regulière, sur un point de leur trajet, sans que cette dilatation ait la moindre rassemblance avec un anévrysme fusiforme et sans que l'exploration artérielle révèle la moindre cause tangible à cet état anatomique singulier."

He called this condition *arteria mega et dolicho*; in one case, also the accompanying veins were involved. No microscopy findings were reported.

In 1966, Staple et al. [3] were apparently the first to report a short series of patients (nine) whose pelvic and limb arteries presented alterations similar to those described by Leriche. Relying on microscopy findings, they stated that arteriosclerosis was the underlying disease.

Some years later, Lea Thomas [4], from St. Thomas' Hospital in London, confirmed this etiopathogenetic theory but observed that similar cases had been reported in pediatric patients [5, 6]. He reported the first consistent series (30 patients, all male, aged 46–75 years) and coined the widely accepted term *arteriomegaly*.

The arteriosclerotic origin was furtherly confirmed by Callum et al. [7] and by Carlson et al. [8]. However, Randall et al. [9] observed that the basic pathological lesion was represented by a marked alteration of the media consisting of the decrease and fragmentation of the elastic fibers, completely lacking in some areas; no inflammation or mucoid deposition was observed; the intima showed reactive fibrosis, and telangiectasia of the vasa vasorum was evident in the adventitia. The authors concluded that arteriosclerotic changes were superimposed and that the fundamental lesion was not arteriosclerotic.

Successively, the group from St. Thomas' [10] recognized that the term arteriomegaly is purely descriptive and that a precise definition of the disease was difficult, asserting however that arteriomegaly is a real entity (likely occurring in 6% of the population over 50 years) which represents a pathological variant of the normal dilating process due to ageing, possibly not related to arteriosclerosis. But, later on, the same group [11] suggested that arteriomegaly could represent a variant of arteriosclerosis in which the elastic layer of the arterial wall is preferentially destroyed.

More recently, D'Andrea et al. [12], on the basis of ultrastructural studies focused on the comparison between arteriomegaly and arteriosclerosis, stressed the specificity of the alterations of the elastic component in the former, suggesting an inheritability of this vascular

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disorder. This would agree with the hypothesis of Lawrence et al. [13], who found a 36% incidence of familiarity studying the first degree relatives aged >50 years of 14 patients with arteriomegaly; this incidence was 1.5 times greater than that observed for 86 patients with abdominal aortic aneurysm.

A clear definition about the nature of arteriomegaly is difficult. However, while some authors [14] are still oriented to consider it as a variant of arteriosclerosis, the prevalent trend [15, 16] is to define it as a specific disease.

The circulatory alterations caused by arteriomegaly are mainly represented by a slowing of blood flow, sometimes so marked as to render difficult, in earlier studies, the angiographic visualization of distal arteries. The slow flow could be responsible for local thrombosis and/or embolism—even in the absence of aneurysms [8]—observed in angiograms as an abrupt arterial cut-off or emboli located also in arteries apparently spared by the dilating process. Lea Thomas [4] argued that these patients are prone to embolism and the clinical corresponding behavior would consist of the fact that symptoms, if any, have sudden onset and short duration.

One of the more intriguing aspects of arteriomegaly is the association with aneurysms. Hollier et al. [17] assert:

> "this entity, especially when associated with diffuse aneurysms at multiple levels, represents a particular process distinct from simple arteriosclerotic aneurysms occurring at multiple sites. It does not necessarily imply specific aneurysm formation, however aneurysms do occur and often are quite extensive. This diffuse aneurysmal disease that occurs in patients with arteriomegaly should be differentiated from multiple isolated arteriosclerotic aneurysms since in general more extensive, entails a higher morbidity and may require different surgical management."

These authors considered aneurysmal disease to be diffuse when involving at least three of the arterial segments examined; they found 91 male patients (aged 36–87 years, mean, 67.5 years) with arteriomegaly and diffuse aneurysmal disease out of 5771 consecutive patients submitted to angiography of the infrarenal abdominal aorta downward the popliteal arteries during the period 1968–1982 and proposed the following classifications:

- Type I (10/91 = 11%): aneurysms present in the aorta and iliac and common femoral arteries, with arteriomegaly of the superficial femoral and popliteal arteries
- Type II (7/91 = 8%): aneurysms in the common femoral, superficial femoral, and popliteal arteries, with arteriomegaly of the aorta and iliac arteries
- Type III (74/91 = 81%): aneurysms in the aortoiliac, femoral, and popliteal arteries, with arteriomegaly of intervening arteries that are not specifically aneurysmal

The authors stressed that arteriomegaly was observed in 300 of the 5771 angiograms (5.2%), but only 91/300 (30.3%) had also diffuse aneurysmal disease; however, as arteriomegaly appears to be a progressive arteriopathy, a number of patients with simple arteriomegaly would probably develop aneurysms. This was confirmed by the study of Barandiaran et al. [18], who, following up for 6–146 months (mean, 76 months) 67 patients with arteriomegaly, observed that only 13 of the 31 patients initially without aneurysms remained in this condition: the number of patients with associated aneurysms passed from 36 (54%) to 54 (81%). The classification proposed by Hollier et al. [17] appears to be clinically relevant, as sudden arterial occlusion was observed in 35% of group III patients and only in 10% and 7%, respectively, of patients in group I and in group II.

A classification of arteriomegaly per se, unrelated with the presence of aneurysm, is offered by Callum et al. [10], relying on 33 postmortem findings and 106 aortograms (all related to male subjects older than 50 years):

- Generalized arteriomegaly, with dilatation and tortuosity of all vessels
- Localized arteriomegaly: some dilated and tortuous segments and also some normal or stenotic or occluded vessels
- Tortuosity without dilatation
- Local dilatation without tortuosity

Tortuosity was experimentally related to failure of elastin by Dobrin et al. [19].

Partially adhering to the latter classification, Bartolo et al. [20], reviewing 1221 peripheral

arteriographies performed during 8 years, found 235 (19.2%) cases of arteriomegaly with the following distribution:

- Dilatation and elongation, 132 (56%)
- Elongation, 41 (17.5%)
- Dilatation, 26 (11.1%)
- Simple tortuosity, 36 (15.4%)

In the first three groups, 66 patients (33.2%) presented also aneurysms.

In patients with arteriomegaly, the occurrence of popliteal artery aneurysm (PAA) (Fig. 9.1) is frequent, as documented in Table 9.1.

A situation theoretically different from the arteriomegaly/aneurysm association is the one called *aneurysmosis* [21], i.e., extensive aneurysmal change involving all or many major arteries. A certain confusion exists about this term that should indicate a particular and extreme situation of multiple arteriosclerotic aneurysms. This was emphasized in the report by Dent et al. [22], who, in a large group of 1488 patients with aneurysm affecting the abdominal aorta or its peripheral branches, identified 57 (3.9%) patients with multiple aneurysms (45 aortoiliac and peripheral, five aortoiliac and visceral, seven multiple peripheral): on a total of 271 aneurysms, 46 (17%) were popliteal. Both in the report and in the extensive discussion on it, arteriomegaly is not mentioned but for the assertion, by Dr. Dent, about the difficulty encountered in separating diffuse ectasia (meaning arteriomegaly?) from true aneurysm formation.

The term aneurysmosis, as pointed out by Lawrence [23], is not included in the suggested standards for reporting on arterial aneurysms [24]; however it is used on several occasions, being considered expressive of diffuse aneurysmal disease. Belardi and Lucertini [25] proposed the following nomenclature:

- Aneurysm, localized arterial dilation
- Arteriomegaly, generalized enlargement of the entire arterial system without aneurysm

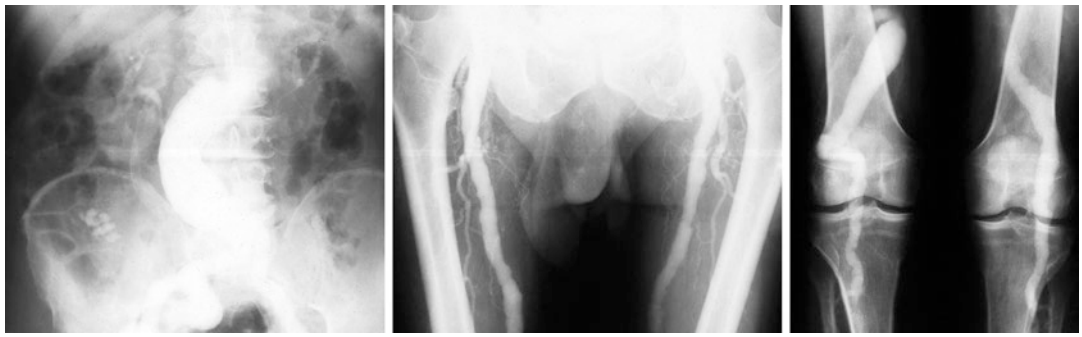

Fig. 9.1 PAA in a case of arteriomegaly

Table 9.1 Occurrence of PAA in patients with arteriomegaly

Author, year	Pts with arteriomegaly	Pts with aneurysm	Number of aneurysms	Pts with PAA	Number of PAAs
Staple [3], 1966	9	6	13	2	2
[a]Lea Thomas [4], 1971	30	20	42		19
Carlson [8], 1975	7	4	15	3	6
Hollier [17], 1983	300	91	410		80
[a]Chan [11], 1990	65			36	59
D'Andrea [15], 2010	18			10	
Barandiaran [18], 2012	67	36 (54)[b]		14 (20)[b]	

[a]From St. Thomas' Hospital, London
[b]Numbers in brackets are related to the end of follow-up (see text)

Table 9.2 Incidence of arteriomegaly or diffuse aneurysmal disease in series of atherosclerotic PAAs

Author, year	Number of patients	Patients with arteriomegaly	Patients with diffuse aneurysmal disease
Crichlow [26], 1966	42		4 (9%)
Bouhoutsos [27], 1974	102[a]	48[a] (47%)	
Evans [28], 1976	52		8 (15%)[b]
Vermilion [29], 1981	87		12 (14%)
Laskar [30], 1982	27	12 (44%)	
Mellière [31], 1986	50	1 (2%)	10 (20%)
Schellack [32], 1987	60		29 (48%)
Lilly [33], 1988	35	19[c] (54%)	
Lowell [34], 1994	106		45 (42%)
Taurino [35], 1998	23		1 (4%)
D'Andrea [15], 2010	40	10 (25%)	
Personal series	58	2 (3%)	

Reports at refs. 29, 32, 33, and 35: not stated if all aneurysms are atherosclerotic
[a]Numbers refer to aneurysms not to patients. The 48 aneurysms were observed in the group of 54 patients with dilating arteriopathy
[b]All eight patients are part of the group with bilateral popliteal aneurysms (34 patients) in which the incidence of diffuse aneurysmal disease is therefore 24%
[c]17/24 patients with bilateral PAA (70%); 2/11 patients with monolateral PAA (18%)

- Aneurysmosis, diffuse aneurysmal disease in patients with arteriomegaly. According to these authors, aneurysmosis would include the three types of arteriomegaly/aneurysm association described by Hollier et al. [17]; however, the occurrence of diffuse aneurysmal disease outside arteriomegaly remains excluded and would be considered as a particular subgroup of the first category.

Keeping into account the probable confusion derived from the variant use of terms, we attempted to tabulate the incidence of arteriomegaly and diffuse aneurysmal disease in some series of PAAs (Table 9.2).

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