Aortic coarctation in the elderly: how many errors lie behind an unexpected diagnosis?

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PHYSICAL EXAMINATION

Aortic coarctation in the elderly: how many errors lie behind an unexpected diagnosis?

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A 65-year-old woman was admitted to our department because of chest pain and untreated hypertension. She had been well until 6 years previously when a diagnosis of hypertension was made. She reported poor control of blood pressure associated with leg weakness in coincidence with tentatively aggressive anti-hypertensive therapy.

Physical examination showed blood pressure of 200/80 mmHg and a heart rate of 80 beats/min. Strong arterial pulsations were present in the neck while femoral pulsations were weak and delayed compared to the brachial pulses. An abnormal difference in blood pressure between upper and lower extremities was present; in fact, blood pressure in the lower extremities was 110/75 mmHg, yielding an ankle/brachial index of 0.55. No clinical evidence of heart failure was observed.

Chest radiography revealed clear lung fields with the cardiac silhouette at the upper limits of normal. The echocardiogram disclosed normal diameters and contractility of the left ventricle, and a mild dilatation of the left atrium (40 mm). Inter-ventricular septum was mildly thickened (12 mm). The aortic valve was sclerotic and displayed mild regurgitation. The evaluation of aortic arch was technically impossible because of the presence of thyroid goitre.

A clinical diagnosis of aortic coarctation was made [1], and a spiral TC study revealed a stenosis of the aorta 2 cm below the origin of the subclavia (Fig. 1, black arrow); a 2 cm aneurysm was present in the proximity of the stenosis (Fig. 1, white arrow) and epiaortic arteries were dilated.

Therefore, a mechanical intervention, by surgical correction or stent implantation, was warranted; however, the patient refused any invasive correction. Anti-hypertensive therapy with enalapril (20 mg/day) and amlodipine (10 mg/day) was started and the patient was sent to follow-up. After 18 months the patient was in good clinical condition, but the control of blood pressure was not adequate. During this period, the patient reported that she had undergone total thyroidectomy because of thyroid goitre without complications.

Aortic coarctation is an unusual cause of resistant hypertension in the adult. Long-term survival in patients with untreated aortic coarctation is rare; in fact the mean survival of these patients is 35 years, with 75% mortality

Fig. 1 Spiral TC revealing a stenosis of the aorta 2 cm below the origin of the subclavia (*black arrow*), and a 2 cm aneurism in proximity of the stenosis (*white arrow*)

by 50 years [2, 3]. In those who survive, the beneficial effect of surgical repair on systolic hypertension has been questioned. Highly effective compensatory mechanisms may eventually develop in some patients [3], accounting for the long-term survival, scant cardiac involvement, absence of heart failure and good outcome at surgery observed in our patient. In accordance with our finding, severe aortic coarctation has been recently reported in an 80-year-old man [4].

In conclusion, the diagnosis of aortic coarctation should always be excluded in an adult in the presence of resistant hypertension. The clinical hallmarks of differential pulse and blood pressures between the upper and lower limbs can be detected clinically. Hence, the importance of a detailed physical examination in all patients with hypertension, including palpation of all the pulses, should be emphasised.

Comment

P.A. Modesti

There are two main points of interest in the present case report. At first, the report of a classic form of congenital secondary hypertension first diagnosed at adult age confirms that it is possible to reach adult age without experiencing blood pressure measurement and physical examination. Secondly, the report discloses the importance of another aspect of clinical skill, that is the capability to communicate effectively with the patient. The clinical course of the patient can be altered by both these parts of clinical examination.

Different epidemiological surveys repeatedly show that about one third of hypertensive patients are unaware of their condition. This is well recognised in mature-adult age when the prevalence of hypertension is so high (50% over 60 years). However, in the paediatric population, probably due to the lower prevalence of high blood pressure, the importance of hypertension is even less appreciated than in adults. Notwithstanding the numerous prevention campaigns prepared by scientific societies and governments, the occasion to have blood pressure measured by a doctor is quite rare for an otherwise "healthy" young person. Military duty forces young adults to pass a clinical screening. A woman has a similar occasion during childbearing or at the time of delivery, but only in this case she gets pregnant. Otherwise, except for cases of preparticipation athletic examinations, an asymptomatic hypertensive young subject may remain *undiagnosed* until their first cardiovascular complication. In aortic coarctation the clinical manifestations depend on the degree of obstruction and the presence of associated anomalies. Severe outflow obstruction leads to early congestive heart failure in infancy. Conversely, more moderate degrees of stenosis allow for circulatory compensation with the development of arterial collaterals so that most children remain asymptomatic although they may occasionally complain of exercise-induced claudication. When adult age is reached, aortic coarctation, although covered by guidelines, cannot be specifically searched for as it is considered only typical of paediatric age. Indeed, peripheral pulse palpation was never previously performed in this middle-aged hypertensive woman although antihypertensive treatment was started. Thus a congenital condition such as aortic coarctation (although rare) can be missed twice, in youth because blood pressure is not measured, and in the adult because it is not specifically searched for. Iuliano did and the mystery of hypertension was solved.

In particular in aortic coarctation, the lack of diagnosis of the hypertensive status stays side by side with the lack of detection of physical signs of reduced perfusion at lower limbs. Indeed although peripheral artery disease is prevalent, physician awareness and detection of the disease is low [5]. Was this the first time this patient referred leg weakness? The symmetrical leg weakness might have been considered by previous doctors. Could this complaint have been attributed to the lack of physical exercise and training, an otherwise common condition in post-menopausal women? This is crucial because when the diagnosis and operation are performed in adult age, the postsurgical survival period is often shorter than that of patients diagnosed and operated on in paediatric age [6].

The next problem for doctors was "what can we do now?". Revascularisation appeared mandatory because blood pressure drop found at lower extremities clearly supported the haemodynamic role of the stenosis and uncorrected aortic coarctation has a high probability of cardiovascular death. Furthermore, according to the information provided, the patient was at low risk for surgery. In this condition, as well as in the stenosis of renal artery, the cardio-cerebral and renal districts have largely different ranges of autoregulation so that great care has to be taken in the perioperative period to limit the possible negative consequences of a rapid increase in renal blood flaw. However, the patient's refusal to undergo revascularisation made this problem academic. Although informed consent is an imperative aspect of clinical medicine, it is often influenced by communication problems. Often patients do not recognise written consent as primarily serving their interests, believing that the main function of signing the consent form is to protect the hospital from litigation, and to give doctors control over what happens [7]. To engage meaningfully in shared decision making and to provide truly informed consent, patients need to have a clear understanding of the benefits and harms of a treatment. To improve the comprehension of educational patient materials used to obtain consent, patient participation in developing informed consent information is sought. As regards

the second point, strong and consistent evidence shows that stated preferences for medical interventions may depend on how the treatment effects are described. The average number of patients in an intervention group who must be treated for a specific period to observe 1 fewer adverse outcomes by the end of this period compared with those in a control group (NNT) provides an easily understood way to describe the effort needed to prevent adverse outcomes. In a recent survey a total of 2754 attendees to a population-based health study were randomly assigned to hypothetical scenarios that presented the benefits of preventive drug therapies in terms of number needed to treat (NNT) or postponement of adverse events. Responders were more likely to consent to treatment when the outcome was described as the number needed to treat to prevent 1 myocardial infarction. They were less likely to consent when the intervention was described as not preventing but delaying a myocardial infarction by 2 months for all persons or by 8 months for 25% of persons. This position cannot be easily extended to surgical correction of aortic disease due to the lack of randomised studies. However the harm of not having revascularisation performed may be particularly relevant for this "two kidneys–two clips woman" where antihypertensive drugs alone can just impair renal function (more important than simple leg blood flow reduction) rather than significantly affect blood pressure in the cardio-cerebral district.

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