Spontaneous rupture of the femoral arteries

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Keywords: Ehlers-Danlos; aneurysm; connective tissue disorder

The Ehlers-Danlos (ED) syndrome comprises nine distinct inherited connective tissue defects[1]. In type IV, the abnormality affects type III collagen, which predominates in arteries, skin, gut and mesothelium[2]. Clinically this results in arterial aneurysms, prematurely aged skin and visceral perforations. Aneurysms occurring in young people, especially in unusual sites, should arouse suspicion, and ED type IV kept in mind as surgery in these patients is hazardous. The condition may be diagnosed on clinical grounds alone, provided that the distinctive phenotype is recognized.

Case report

A 37-year-old man developed a tender right inguinal swelling, initially diagnosed as a hernia, but subsequently, after developing extensive bruising, as a 'ruptured vessel'. One year later the same occurred on the left, but on this occasion was complicated by marked calf claudication.

Five months later he was seen in the Bloomsbury Vascular Unit, by which time he was almost asymptomatic. A past history of emergency laparotomy for a 'burst colon' was noted. Other than an absent left posterior tibial pulse his vasculature was normal, and despite looking for manifestations of connective tissue disorder none was found. His specific hypermobility index[3] was normal, as were routine blood tests. His urinary hydroxyproline was marginally elevated. Arteriography showed normal carotids, subclavians and aorta, but aneurysmal segments in the ileofemoral regions (Figure 1) with broadened femoropopliteal segments.

To prevent further rupture, exclusion of the aneurysmal areas by aorto-femoral bypass was attempted. From the onset there were technical difficulties. A ligature around a side branch of the left femoral artery cut through and so friable was the vessel wall that all attempts at control failed. Fortunately the aorta was never fully exposed, but 3 h were spent trying to obtain haemostasis in the left groin. This was finally achieved by nylon tape ligation of the external iliac artery, with a synthetic patch plus sleeve around the common femoral.

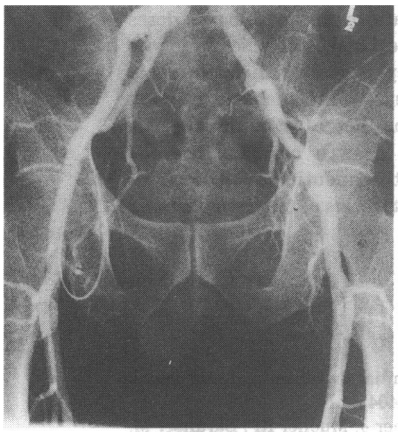

Figure 1. Transfemoral arteriogram - Ileo-femoral view. Note the aneurysmal segments in both common iliac arteries, and in the left common femoral

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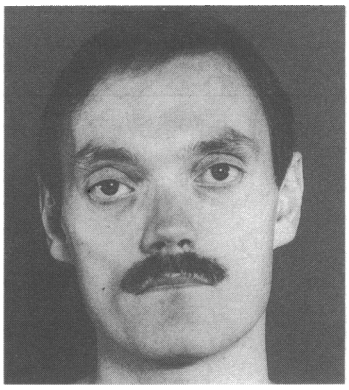

Figure 2. The characteristic facies, in our patient, of the acrogeric form of Ehlers-Danlos syndrome (published with the permission of the patient)

The patient made a slow recovery, complicated by an acute gastric dilatation. He was subsequently seen in consultation by one of us (FMP), who identified the typical facies of ED type IV. A skin biopsy was taken for fibroblast culture. These were shown to produce abnormal type III collagen.

One year later, he remained well without claudication, and now had a left popliteal pulse despite the ligated external iliac artery.

Several members of the family have been reviewed, and a pedigree established. His sister and nephew had typical facies, but except for minor bruising were asymptomatic. All members (sister, nephew, two daughters) had biochemical evidence of ED type IV, either producing abnormal type III collagen, or having abnormal dermal collagen fibrils on electron microscopy. His mother had died at 49 years of 'acute pancreatitis', she too had the typical facies.

Discussion

The association between inherited connective tissue disorders and peripheral aneurysms has long been recognized[4]. In ED type IV the cutaneous and facial manifestations along with the arterial wall defects should render it recognizable. Most families, like the one described here, have prematurely aged skin, lobeless ears, pinched noses and owl-like eyes (Figure 2), whilst others are more difficult to recognize. Hypermobility, in contrast to other types of Ehlers-Danlos syndrome, is not a constant feature. The fragility of the arterial wall renders elective reconstruction almost impossible, and we consider ourselves and our patient very fortunate to have escaped so lightly. Rupture is often fatal, and the diagnosis is sometimes made only at autopsy[5].

Given more precise methods of characterizing the structural proteins of the arterial wall, it is possible that we will have to revise our concepts of the causation of aneurysms, hitherto rather simplistically dismissed as a manifestation of atherosclerotic degeneration.

References

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(Accepted 1 October 1991)