Paget’s disease and angioid streaks: one complication less?

One or two cherished pieces of ophthalmological folklore have been demolished recently. First there were the ‘cholesterol crystals’ of the Christmas tree cataract, shown by Hayes and Fisher to be no such thing; next there was the association between pre-existing chronic glaucoma and central retinal vein occlusion, and possibly now, in this month’s issue of the BJO, the widely taught dictum of Paget’s disease being associated with angioid streaks. One is worried about what may come next – perhaps some ophthalmological Illych to tell us that holes in the retina are not associated with detachment or, dare I say it, that raised intraocular pressure has nothing to do with glaucoma.

Traditions in medical literature, whereby one generation of clinicians takes it upon trust that certain associations of symptoms and signs make up genuine syndromes may be difficult to trace back to their origins. This is not a problem with eponymous syndromes, since the original paper or papers should be easily traceable. But even this exercise sometimes leads to surprises, and one finds that the syndrome bearing the eponymous name is hardly mentioned in the original paper, or, as in the case of Graves’ disease for example, the original communication was made at a meeting of a hospital society and not published at all (Graves’ paper can be found only in an anthology of clinical lectures). According to Duke-Elder the association of Paget’s disease with streaks was first pointed out by Batten in 1931 and by Verhoeff in the same year. Unfortunately both these references are seriously flawed. Batten does not report a case of his own but merely writes that he has seen a fundus painting in Hamblin’s collection of drawings of a case of Frank Juler’s which was said to have angioid streaks associated with Paget’s disease. In the same paper Batten quotes Verhoeff: ‘I have seen a typical case in a patient with Paget’s disease of the skull but I am not prepared to say that it is of any special significance.’ Unfortunately this is said to have been printed in a ‘presession book’ of the section of ophthalmology of the American Medical Association in 1928, but it does not seem to be available in the libraries I have access to. To make matters worse, the 1931 Verhoeff paper quoted by Duke-Elder is not about angioid streaks at all but about some entirely different fundus streaks resulting from choroidal detachment. Verhoeff even makes a point of emphasising that they are not angioid streaks. Thus, correctly as far as I can see, the most commonly quoted first description is usually that of Terry, as pointed out by Dabbs and Skjodt in their article. If Terry’s four examples of angioid streaks out of 50 cases of Paget’s were really definite, as he claims, it would be impossible to dismiss such an association of two such rare conditions as occurring by chance alone. But if one searches the literature, say for the years 1950 to 1980, one constantly meets allusions to the association, with few fresh cases being reported, rather like the Batten-Verhoeff fiasco. So the suspicion begins to creep in that perhaps there was something not quite right with Terry’s own observations and what we have been experiencing is the phenomenon of perpetuating a medical rumour.

It is of considerable interest that, in the paper by Shields and colleagues, of 56 patients with angioid streaks seen during a period of seven years not a single one was suffering from Paget’s disease, whereas no fewer than 30 had pseudoxanthoma elasticum. The literature abounds with case reports of angioid streaks most of which are entitled ‘Two cases of the Grönblad-Strandberg syndrome,’ but when the ocular associations with Paget’s disease are mentioned it is usually in the context of compressive lesions of the optic nerve due to bony changes in the skull. One begins to suspect therefore that Dabbs and Skjodt might be correct in their conclusion that Paget’s disease is only infrequently associated with angioid streaks, to put it mildly.

A paper which is not often cited is that of Mazalton and colleagues, which reports a case in which Paget’s disease and the Grönblad-Strandberg syndrome occurred in the same patient. The authors also reported that four other cases had been described previously. It was not entirely clear whether angioid streaks were seen in their particular case, but, given that they seem to occur in around 80% of cases of the Grönblad-Strandberg syndrome and that the case being described had a haemorrhagic retinopathy, it is likely that streaks were present. A boost to Terry’s theory is given in a paper from the Mayo Clinic in which precise percentages are given of patients with pseudoxanthoma elasticum (PXE) alone or PXE plus streaks and streaks alone or streaks combined with Paget’s disease. Whereas 63 of a total of 74 PXE cases had streaks, only two out of 32 cases of Paget’s disease had streaks – that is, around 6% – not far out of line with Terry’s 8%. Thus one’s original suspicion that the Paget’s-angioid streaks association was just another piece of ophthalmological folklore about to be disproved turns out not to be justified. Added confirmation that there is probably some truth in the story is given by Gass and Clarkson. They followed up a case to necropsy and describe not only the clinical features but the histopathological changes in Bruch’s membrane. There is only one defect in this paper and that is the published fundus photographs of the streaks, which are
not entirely sharp. However, this is not a serious criticism, since it is noticeable that fundus photographs in which streaks are alleged to show are often rather unconvincing. It appears that angioid streaks are not photogenic. The validity of a claim depends entirely on the opinion of the observer as to what is or is not an angioid streak on ophthalmoscopy; it is not possible to be certain one way or the other.

One can finally conclude that the coincidence of angioid streaks with Paget’s disease certainly does occur but perhaps not as often as some clinicians seem to think, and the streaks are far more likely to be found in the Grönlund-Strandberg syndrome (PXE) (not to mention one or two other conditions, not the subject of this editorial). Finally it has to be said that, if Paget’s disease can coexist with PXE, a case of Paget’s disease with streaks should obviously be very carefully examined to exclude PXE; and, who knows, the pure Paget’s-streaks association might yet turn out to be false.

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7 Verhoeff FH. The nature and origin of the pigmented streaks caused by the separation of the choroid. JAMA 1931; 97: 1873-6.