Sir,

Angioid streaks with congenital hypertrophy of the retinal pigment epithelium: an association or a mere coincidence?

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Angioid streaks usually have a characteristic ophthalmoscopic appearance.[1] They are caused by histologically demonstrable ruptures in Bruch's membrane[2] and are most commonly associated with pseudoxanthoma elasticum (PXE).[3] We report this case, with angioid streaks and unilateral congenital hypertrophy of retinal pigment epithelium (CHRPE). This association has not been reported before.

Case report

A 45-year-old overweight Caucasian gentleman was referred to the eye clinic by the dermatology service with an unusual retinopathy. For 18 years he had recurrent painful folliculitis of the scalp. (Figure 1a, top row). This was treated with a variety of drugs including Dapsone, Doxycycline, Thalidomide, Roaccutane, Rifampicin, Clindamycin, Azathioprine, Cyclophosphamide, Cyclosporin, Methotrexate, Mycophenolate mofeitil, Colchicine, antibiotics and antihistamines. Despite extensive investigations including multiple skin biopsies no underlying cause had been found.

He underwent a colonoscopy for non-specific abdominal symptoms. No abnormalities were found and he was thought to have irritable bowel syndrome. He had diet-controlled Type II Diabetes mellitus. He had also been investigated for generalised morning stiffness and musculoskeletal aches, the only abnormalities discovered were degenerative changes in the lumbar spine (L3 downwards).

On ocular examination his visual acuities were 6/6 in either eye with normal pupillary reactions. The anterior segments and intraocular pressures were normal. Angioid streaks were present and the fundi had a mottled appearance (peau d'orange) most apparent temporal to the macula. There were multiple, variable sized, well-circumscribed, oval, grey lesions at the level of the retinal pigment epithelium in a 'bear-track' pattern in the superonasal quadrant of the left fundus. (Figure 1b, bottom row).

The patient was referred to a geneticist with a special interest in PXE families. There were no cutaneous changes of PXE; the neck, axillae, flexures and buccal mucosa were free of elastotic deposits. The radial pulses were weak; this can be a feature of PXE. He had a positive Beighton score of 4/9 with joint laxity of the hands and knees. His mother and son were also examined at the same visit. His mother had some degree of joint laxity with valgus positioning of

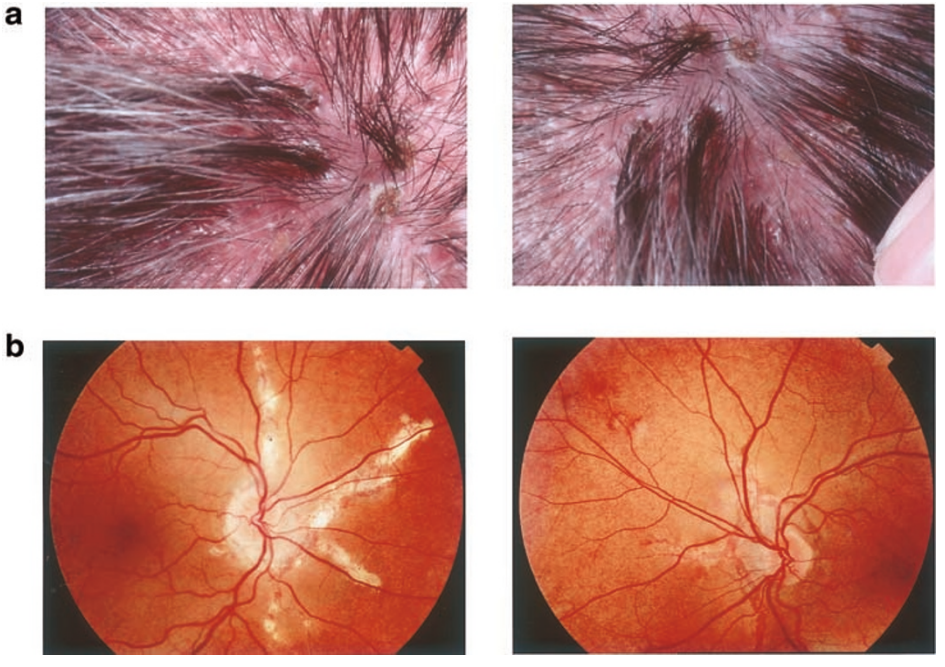

Figure 1 (a) Top row. Close up photographs of scalp folliculitis. (b) Bottom row. Fundus photographs showing bilateral angioid streaks with a mottled appearance (peau d'orange) of the fundi. The superonasal quadrant of the left fundus shows multiple, well-circumscribed, oval, grey lesions in a 'bear-track' pattern.

her knees and loose-jointedness. Her vision was 6/4 bilaterally, without any angioid streaks. His son had stiff joints except his knees, extensible skin over the neck and face and a normal ocular examination.

It was felt that our case was a PXE heterozygote and that angioid streaks were part of this clinical phenotype. In the absence of cutaneous changes homozygosity for the PXE gene is unlikely, although it cannot be totally excluded. DNA has been extracted and banked to test for the PXE gene in the future. A formal skin biopsy from the side of the neck showed minimal elastotic degeneration and elastic fibres were not Von Kossa positive. There were no particular features suggestive of PXE. The recent normal colonoscopy rules out Familial Adenomatous Polyposis (FAP).

Comment

Congenital hypertrophy of the retinal pigment epithelium (CHRPE) is a well-known association of Familial Adenomatous Polyposis (FAP). On reviewing literature there is one case report of a middle-aged Caucasian male with angioid streaks but no CHRPE who had extensive FAP.[4]

The relationship between CHRPE and angioid streaks is probably coincidental and has not been reported before. The previously reported case had extensive FAP necessitating a total colectomy, though his fundi did not show any features of CHRPE. Both these patients were Caucasian males, of similar age and with radiological evidence of degenerative changes in their lumbar vertebrae. The striking similarity between these cases and the fact that CHRPE and FAP often are part of the same syndrome suggests that their relationship with angioid streaks could be part of the same syndrome. In the present case the cutaneous manifestations of PXE were minimal, while in the previously reported case FAP was not associated with CHRPE, possibly indicating an incomplete expression.

References

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HS Ahluwalia, A Lukaris, CM Lane and C Blyth

Department of Opthalmology
University Hospital of Wales
Heath Park
Cardiff CF14 4XN, UK

Correspondence: CM Lane
Tel: 02920742083
Fax: 02920743222
E-mail: nicola.jones@uhw-tr.wales.nhs.uk

Sir,

Gonococcal keratoconjunctivitis in adults
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Gonococcal ocular infection can be divided into two distinct forms, one affecting neonates and the other affecting sexually active adults. Most cases occur in neonates or sexually active adults and are transmitted by contact with infected urine or genital secretions. Recently, the incidence of adult gonococcal conjunctivitis has shown a tendency toward increasing with time, especially penicillinase-producing *N. gonorrhoeae* (PPNG).[1,2]

As gonococcal eye infection in adults is relatively rare, the clinical diagnosis may be delayed. However, it is very important that a prompt confirmatory culture for isolation of gonococcal organisms and earlier parenteral antibiotic treatment is required, because the outcome of gonococcal conjunctivitis is related to the severity of disease at the start of adequate therapy.[3]

We present a rare case of bilateral gonococcal conjunctivitis with keratitis, gram-negative intracellular diplococci, penicillin-resistant *Neisseria gonorrhoeae*, associated with a sexual history of relations with a prostitute.

Case report

A 29-year old man presented redness and ocular discharge from both eyes for 13 days before admission to Pusan National University Hospital in Pusan city. At first, the left conjunctiva was markedly inflamed and there was intense dilatation of the conjunctival vessels without small petechial hemorrhages with purulent exudates. The right eye was completely normal. The other eye became involved on the 7th day after left conjunctivitis developed. Eventually, he failed to respond, so was referred to our clinic for proper management of mucopurulent conjunctivitis.

He had a distinctive sexual contact history with a prostitute 17 days previously, but there was no evidence of genitourinary symptoms. On the day of