Section case reports

Huge bladder diverticula associated with Ehlers-Danlos syndrome

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Ehlers-Danlos syndrome may cause diverticula in unobstructed male bladders. Experience suggests a conservative approach, unless excretory function or symptoms warrant surgery. We present a case and review the literature on the subject.

Case report

A four-year-old boy with no previous urinary infection was referred following a week of dysuria and ballooning of a tight foreskin. At 7 weeks he had had bilateral inguinal and at 2 years recurrent right inguinal and umbilical hernia repairs. Investigation for muscular hypotonia led to an initial diagnosis of Ehlers-Danlos syndrome (EDS) type VII. Neither parent had EDS although his mother had hypermobile joints, with spontaneous hip dislocation during pregnancy, and his father had recurrent inguinal hernias.

Examination revealed the characteristic facies, lax skin and hypermobile joints of EDS. He had a protuberant lower abdomen with a 4 cm palpable bladder and a tight, inflamed prepuce. His biochemistry and haematology were normal and, although urine cultures were negative and he was apyrexial, he was treated for urinary infection and his foreskin was relieved by preputial plasty. His voiding symptoms improved, although the bladder remained palpable.

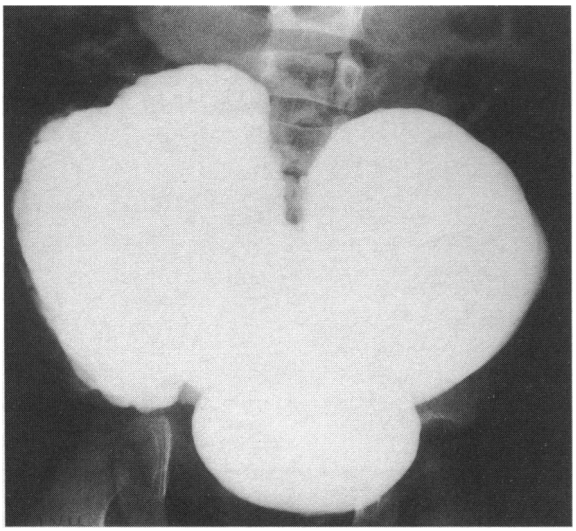

Figure 1. Cystogram showing the bladder displaced to the right by two large left-sided diverticula

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Ultrasound suggested an enormous obstructed bladder with multiple diverticula and a hydronephrotic left kidney with ureteric dilatation to the vesico-ureteric junction. Urethrocystoscopy showed no urethral obstruction and a cavernous, trabeculated bladder, so he was catheterized and put on prophylactic antibiotics. Cystogram (Figure 1) and intravenous urogram after 10 days revealed a small bladder with two enormous left sided diverticula. The left kidney appeared obstructed and mild vesico-ureteric reflux was demonstrated on the right. A MAG3 renogram showed delayed excretion and 37% of function on the left.

At operation two huge diverticula were seen; one anterior and another postero-lateral to the bladder displacing it to the right and compressing the left lower ureter. Both were excised, leaving a small residual bladder into which the left ureter was reimplanted with difficulty, due to very friable tissue. Histology of the diverticula revealed transmural inflammation with attenuated muscle fibres and abnormal elastin deposition. Four weeks later repeat urogram showed improvement of the left upper tract and 9 months later he is asymptomatic with good bladder capacity.

Case presented to Section of Urology, 25 March 1993

Discussion

The Ehlers-Danlos syndrome is a congenital abnormality of collagen metabolism with clinical and genetic heterogeneity and ten recognized subtypes[1]. Of unknown incidence, it is probably the commonest inheritable connective tissue disorder[2]. Inheritance is autosomal dominant (types I, II, III, IV, VII, VIII), autosomal recessive (types IV, VI, VII, X) or X linked recessive (types V and IX) with variable penetrance[1]. Manifestations include skin laxity, joint hypermobility, hypotonia, poor muscle development, an 'elfin' facies and recurrent hernias, as in our patient. Hollister has reviewed its effects on other body systems[3]. Medullary sponge kidney, polycystic kidney disease, pelvo-ureteric junction obstruction and bladder diverticula are reported in the urinary tract[4,5].

At least 24 cases of bladder diverticula are reported[6-12] since 1942[6]. All are male and aged 18 months to 49 years, with 80% presenting before 16 years. Urinary infection, dysuria, haematuria, abdominal mass, stones and bladder rupture are recognized presentations and it is seen with several subtypes (I, II, III, IV, V, VI and IX)[6-12].

Why do patients with EDS develop bladder diverticula? Eadie[7] and Zalis[8] blamed increased distensibility of connective tissues and Levard[9] thought higher voiding pressures in males explained their absence in female patients. Coexistent bladder outflow obstruction has been suggested[6,10] and posterior urethral valves were found in one case[11], although urodynamics is invariably normal[8,10]. Bladder diverticula are increasingly recognized in X-linked subtypes, as in five out of seven cases of EDS type IX[4] and male to male transference has been seen in five male members of one family[12].

Fifteen cases have been treated by diverticulectomy with histology like our case of attenuated muscle and sclerotic connective tissue. Ten recurrences are documented, even after *v-y* bladder neck plasty for supposed obstruction[6-12]. Surgery should be reserved for severe cases, particularly as complications of delicate friable tissues, per- and post-operative bleeding, poor wound healing and incisional hernias are common in these patients[5].

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Complications of artificial hair implantation

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Synthetic fibre implantation is currently being marketed as a treatment for male pattern baldness. The technique involves implantation of synthetic fibres into the scalp under local anaesthetic. It was discredited over 10 years ago in the USA following the development of severe reactions in the majority of patients undergoing this treatment[1-4]. Claims have been made that revised techniques significantly reduce complications and provide a suitable alternative therapy for baldness[5,6]. We report a case in which severe complications resulted following artificial hair implantation with a revised method.

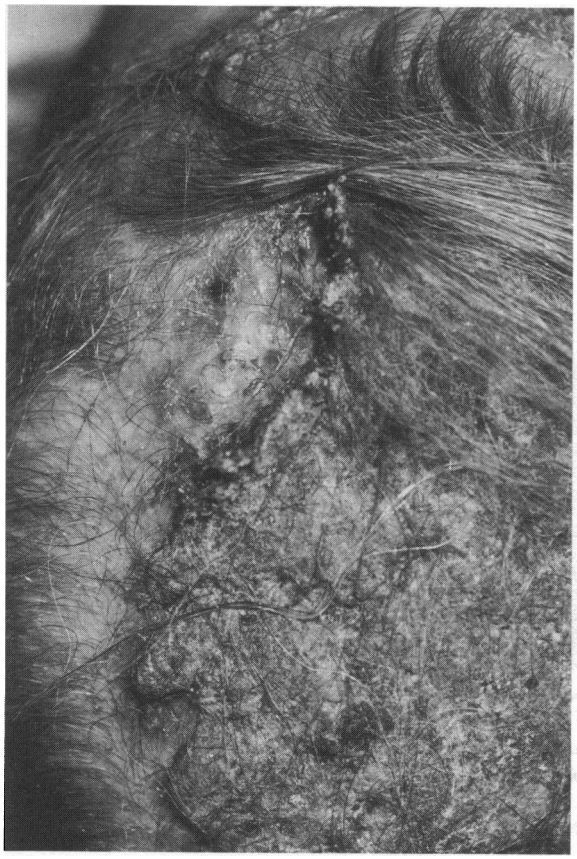

Figure 1. Scalp at presentation covered with thick crust and enmeshed artificial hairs. Removal of the crust revealed extensive cutaneous erosion covered by thick pus and exudate

Case report

In 1984 a 54-year-old man underwent hair transplantation with autologous punch grafting as a treatment for his male pattern baldness. After several years he decided to have the punch grafts removed and replaced with a flap of hair bearing skin (Juri flap) which was taken from the right side of his scalp and transposed over the frontal scalp to form a frontal hairline. He was reasonably happy with the result but felt that the hairline looked unnatural.

In early 1991, in response to an advertisement in a daily newspaper, a clinic consultation and a persuasive brochure he underwent the procedure of synthetic fibre implantation in order to improve the appearance of his hairline, as well as increasing the coverage of hair on the vertex. Over three 2½ h sessions at weekly intervals multiple synthetic polyester fibres were implanted individually under local anaesthetic into the scalp both behind and in front of the existing surgical flap. The cost of the procedure was £4000. At no stage was there any mention of potential complications. Following the third and final visit he was discharged without follow-up.

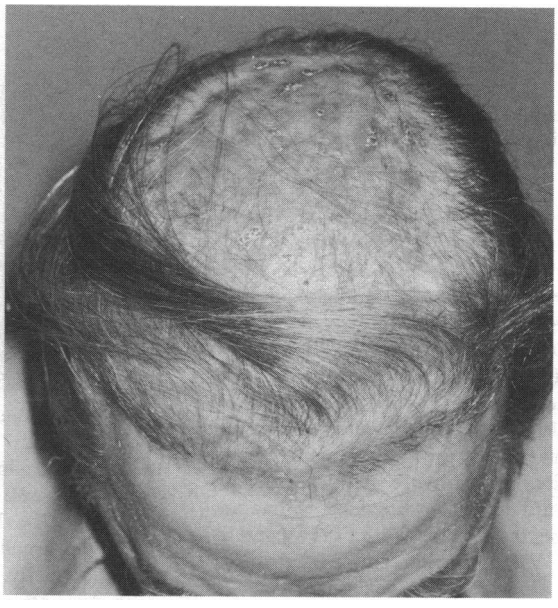

Figure 2. Extensive residual scarring in the areas of synthetic fibre implantation after resolution of the inflammation