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Persistent Müllerian duct structures: Review and report of one case

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Summary

A case of bilateral inguinal hernias, right ectopic testis and left undescended testis with persistent Müllerian duct structures is reported.

Key words: Inguinal hernias, cryptorchidism, Müllerian duct structures.

Introduction

The persistence of Müllerian duct derivates in the male is well recognized and more than 100 cases have been reported [8]. Persistent Müllerian duct syndrome was first desribed by Nilson in 1939 as hernia uteri ingunale [5]. He stated that such patients usually present with cryptorchidism and inguinal hernias and the hernial sac containing the uterus and fallopian tube.

Case Report

A twenty-year-old male was referred for evaluation of bilateral undescended testes and inguinal hernias. His somatic and mental development was normal. He was the youngest of six siblings (three brothers and two sisters) and his youngest brother who was married and had no child had undergone left orchiectomy and herniorraphy for left undescended testis and inguinal hernia when he was nineteen but the details of which were unavailable. Other brothers and sisters who had children were normal. Physical examination revealed right inguinal hernia and absent right testis and left inguinal hernia containing two solid masses and undeveloped empty scrotum. Secondary sex characteristics and external genital development was normal for his age (Fig 1). Seminal analysis showed absent spermatozoa. Serum FSH, LH and testosterone levels were normal for age. Chromosome studies done on lymphocytes revealed a normal male karyotype of 46 XY.

An exploratory laparotomy was performed via left inguinal incision and a testis (3.5x2.5x2 cm), a small uterus (4x2.5x2 cm) with fallopian tube were found in the hernial sac. A total resection and herniorraphy was performed (Fig 2). Histological examination of testicular samples demonstrat-

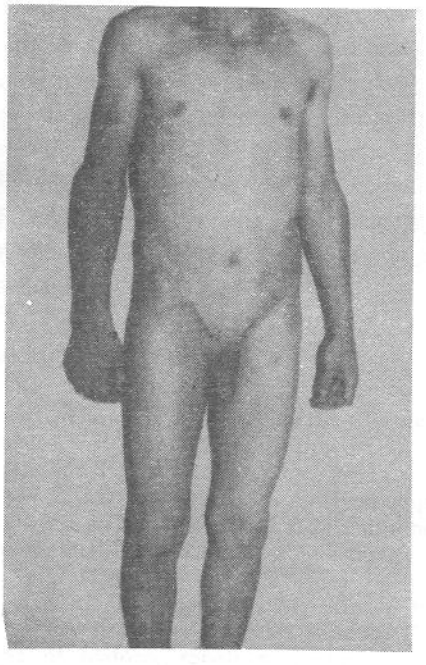

Fig. 1. Patient after both operations. Normal differentiation of external genitalia.

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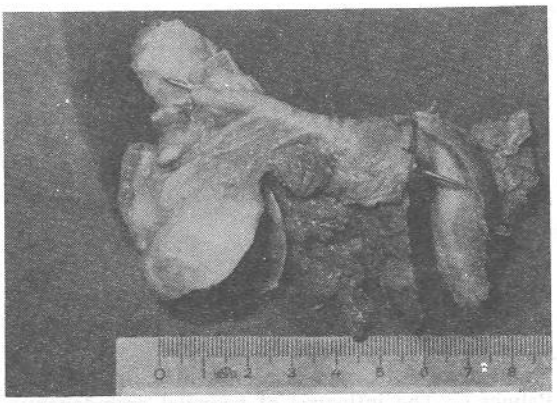

Fig. 2. Gross specimen of left atrophic testis, uterus and fallopian tube that were surgically removed.

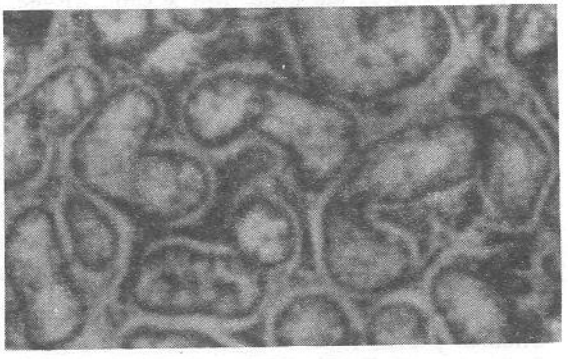

Fig. 3. Left atrophic testis showing peritubular sclerosis with no evidence of spermatogenesis (HE,x40).

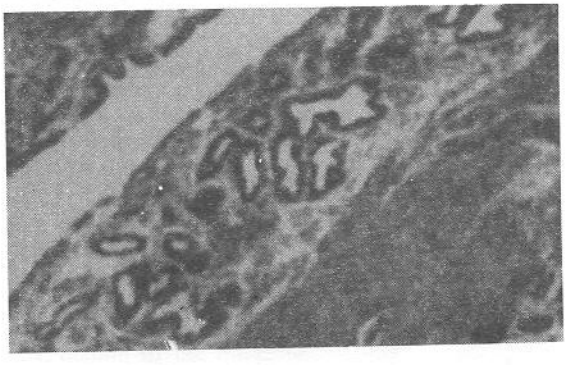

Fig 4. Endometrium and myometrium (HE, x40).

ed peritubular sclerosis, germinal cell aplasia, diffuse hyperplasia of the interstitial cells and sertoli cells lining the seminiferous tubules (Fig 3).

The uterus lined with endometrium with a fallopian tube and a round ligament-like structure (Fig 4-5) and an epididymis with no external orifice was located at the proximal end of this structure. Two months later an exploratory laparotomy was performed through a right inguinal incision. The

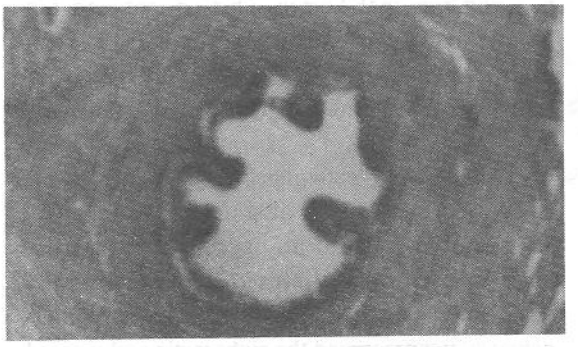

Fig. 5. Fallopian tube (HE,x40)

right testis was found to be located intraperitoneally just behind the internal inguinal ring (2x2x1 cm and of normal consistency). Orchiopexy and herniorraphy was done. Testicular biopsy revealed germinal cell aplasia, no evidence of malignancy was noted.

Discussion

The persistence of Müllerian structures is due to the lacking of the Müllerian inhibitory factor (MIF), defect in the inhibitory factor or the unresponsiveness of the ipsilateral target organs. MIF is a protein secreted by the sertoli cells of the testis from the eight fetal week up to two years of age and acts ipsilaterally [2]. During this period Müllerian ducts undergo regression. In this syndrome, Müllerian structures are not inhibited and uterus and fallopian tubes develop mean while testosterone produced by the fetal testes permits normal development of the male external genital organs and secondary sex characteristics.

Since Müllerian duct remnants may give rise to symptoms in the form of incontinence, retention cysts or tubular utricular structures [4], we performed urography and voiding cystourethrography one month after the first operation and the findings were normal. The view that the syndrome is hereditary is not clear. Autosomal recessive or dominant inheritance with sex limitation [9] or X-linked recessive inheritance is compatible with various reported cases. The syndrome has been reported in eight pairs of siblings suggesting an autosomal recessive pattern of inheritance [9,1].

One brother of our patient had undergone left or-

chiectomy and herniorraphy for left cryptorchidism and inguinal hernia. Our patient and his brother may be an example of familial persisent Müllerian duct syndrome but we could not obtain a detailed clinical information to conform it.

In our case both testicular biopsies revealed no evidence of malignancy although the patient was 20 years old and had no previous operation or hormone therapy. He refused right orchiectomy despite he was cautioned about the risk of malignancy so we preserved the right testis and warned him that his testis still had the tendency to undergo malignant change and he was informed about the importance of periodic controls.

In general it is believed that the incidence of testicular tumors in persistent Müllerian duct syndrome is similar to that of the simple cryptorchidism [6,3,7]. Melman and associates believe that orchiopexy may not protect against future development of testicular tumor even if performed in the neonatal period [3].

In our case seminal analysis and bilateral testicular biopsies revealed azoospermia and germinal cell aplasia. If the diagnosis is made during the first few year of life we believe the surgical procedure should be an orchiopexy to preserve fertility.

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