—Case Reports—

Microscope-assisted Reduction Clitoroplasty Used to Treat Two Patients with Clitoromegaly

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Abstract

We report 2 cases of clitoromegaly, 1 in a patient with true hermaphroditism, and the other in a patient with adrenogenital syndrome. Both were treated surgically with reduction clitoroplasty. There are 3 different clitoroplasty procedures: clitorectomy, clitoral recession, and reduction clitoroplasty. Reduction clitoroplasty with preservation of the neurovascular bundle is considered superior in terms of formation of the external genitals and sensation. However, the disadvantages are that detachment of the neurovascular bundle from the clitoral shaft is difficult and that there is a high possibility of sensory and blood flow disorders in the clitoris. In an attempt to achieve safe and reliable surgical manipulation, we used a surgical microscope (OPMI 6-SDFC, Carl Zeiss Surgical GmbH, magnification ×8) to detach the neurovascular bundle from the clitoral shaft in our 2 patients. Our impression is that our efforts were extremely effective. Furthermore, our experience leads us to believe that the procedure for neurovascular bundle detachment required in reduction clitoroplasty is not particularly difficult if performed with a surgical microscope by a plastic surgeon who regularly performs microsurgery. Because the procedure can be performed simply and safely, we believe that reduction clitoroplasty with preservation of the neurovascular bundle is the best overall of the 3 clitoroplasty procedures.

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Key words: clitoromegaly, reduction clitoroplasty, microscope, true hermaphroditism, adrenogenital syndrome

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Introduction

Clitoromegaly has various causes, but whatever the cause, surgery is needed when the patient is 2 or 3 years old so that he or she can acquire a sexual identity. We have treated 2 patients with clitoromegaly, and we describe here the case histories and our findings regarding the useful role of a surgical microscope in detaching the neurovascular bundle from the clitoral shaft and in allowing appropriate selection of operative procedures.

Case Report

Patient 1 was 2 years old and was recorded in the family register as a girl.

Medical history: Nothing of note

Family history: The older of 2 daughters; family history, including that of the younger sister, was unremarkable.

Prenatal history: The patient was delivered by caesarean section, with no other findings of note.

History of present illness: Clitoromegaly was recognized at birth, but the patient was raised as a girl. She was referred to our hospital for clitoroplasty. Findings of the initial examination included a subcutaneous mass under the right labium majus and urogenital sinus in addition to clitoromegaly **(Fig. 1)**. Suspecting hermaphroditism, we conducted human chorionic gonadotropin (HCG) loading and chromosomal tests. The former revealed elevated testosterone, whereas the latter showed a normal female 46XX pattern. Therefore, we performed a laparoscopic biopsy of the gonads to determine whether the diagnosis should be true hermaphroditism or XX-male syndrome at the beginning of the surgery. Thereafter, we removed the subcutaneous mass from the right labium majus and performed reduction clitoroplasty under a surgical microscope (OPMI 6-SDFC, Carl Zeiss Surgical GmbH, Oberkochen, Germany, magnification ×8), preserving the neurovascular bundle **(Fig. 2)**; we also performed a cut-back procedure for vaginal construction. An intraperitoneal gonad tissue sample was sent for rapid intraoperative histopathological examination and was found to be an ovary, so the remainder was preserved. Permanent histopathological findings showed that the gonads in the peritoneal cavity were ovaries, as also revealed by the rapid intraoperative histopathological test, but that the subcutaneous mass was an ovotestis. The diagnosis was therefore confirmed as true hermaphroditism **(Fig. 3)**.

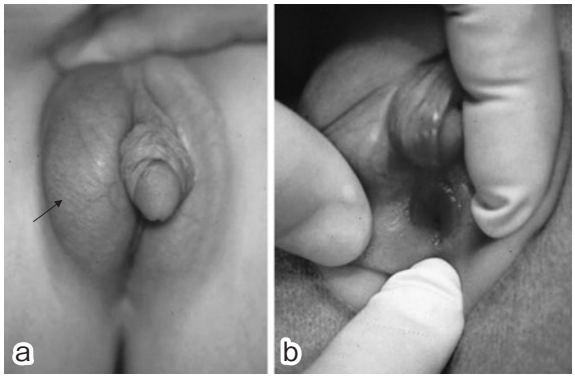

Fig. 1 Preoperative view
a: Clitoromegaly and subcutaneous mass **(arrow)**
b: Urogenital sinus

One year after surgery, there was a slight excess of skin resembling scrotal skin, and the clitoris was a little larger than normal, but the shape of the external genitalia had improved **(Fig. 4)**, and the patient was living as a girl, as she had before surgery. Moreover, no elevation in testosterone was seen with an HCG loading test.

Patient 2 was a 5-year-old girl.

Medical history: Nothing noteworthy

Family history: The older of 2 daughters; family history, including that of the younger sister, was unremarkable.

Prenatal history: The patient was delivered by vacuum extraction because of fetal distress; the Apgar score was 9.

History of present illness: After birth, the patient was referred to a pediatrician because of poor sucking, and at that time brownish skin and clitoromegaly were observed. Congenital adrenal hyperplasia was suspected, and various tests were performed. The resulting diagnosis was simple andromorphous congenital adrenal hyperplasia, and

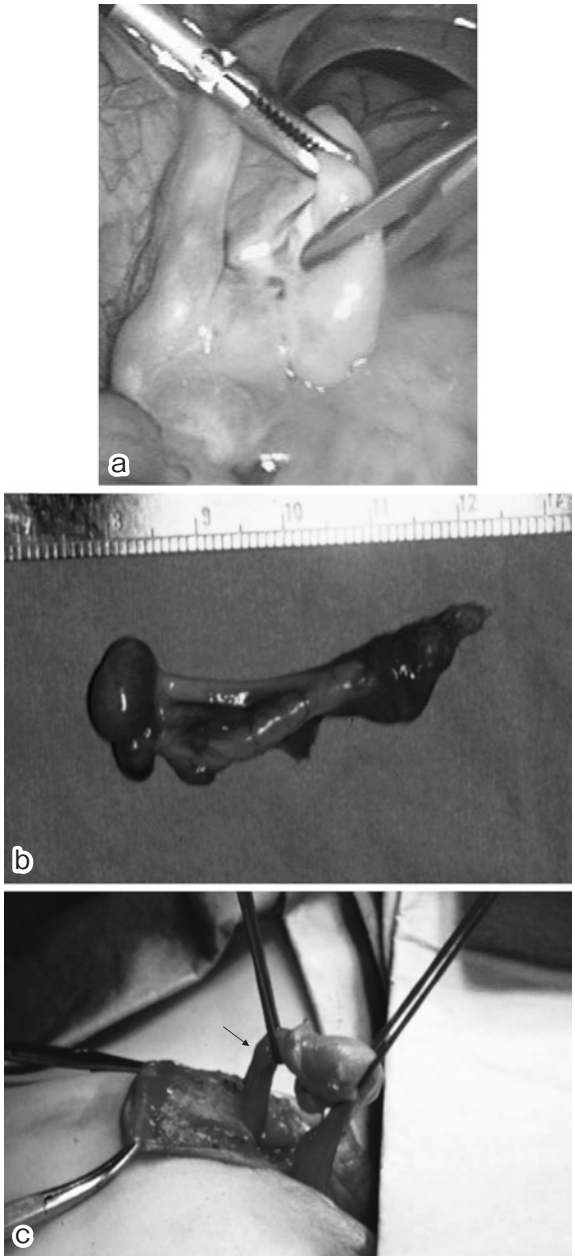

Fig. 2 Intraoperative view
a: Intraperitoneal gonad tissue
b: Subcutaneous mass
c: Neurovascular bundle **(arrow)**

oral administration of hydrocortisone was started at 20 to 30 mg/day. The patient had previously undergone incomplete clitoral recession and reduction clitoroplasty for clitoromegaly, but the appearance of the external genitalia remained poor **(Fig. 5)**. We performed reduction clitoroplasty with preservation of the neurovascular bundle under a surgical microscope **(Fig. 6)**. The result, as of 2 years after surgery, has been a clear improvement in the appearance of the external genitalia **(Fig. 7)**.

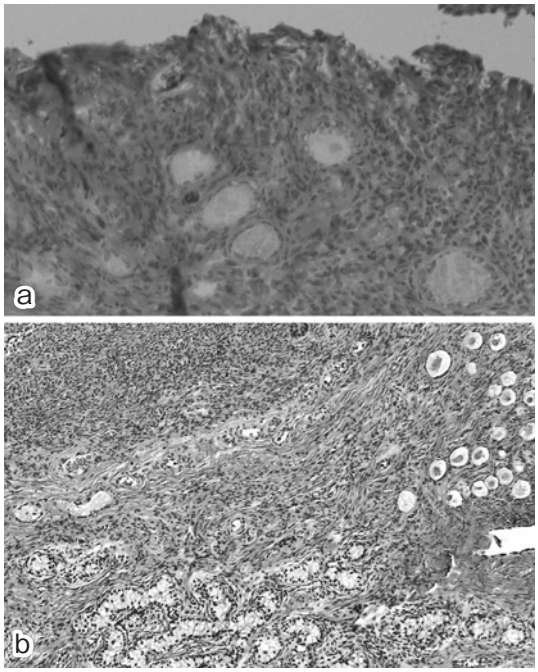

Fig. 3 Histopathological findings
a: Only ovarian tissue is found.
b: Both ovarian tissue and testicular tissue are found.

Discussion

More than 160 cases of true hermaphroditism have been reported in Japan[1], but it remains a rare condition. The formation of the external genitalia is important in the acquisition of a sexual identity, so the best time for surgery is usually when the patient is 2 to 3 years old. The legal sex of the child must be determined before surgery from an overall investigation based on the results of chromosome or endocrine tests and the status of the external and internal genitalia. On the other hand, because a child's sex must be selected so that he or she can be accepted by the family and society, sexual function is unfortunately sacrificed in some cases. Some reports suggest that in cases where there is any question about which sex to select, opting for the female sex is preferable[2].

Patient 1 was recorded as a girl in her family register, and the family also strongly wished to continue bringing her up as a girl. Selection of the female sex was also appropriate from the point of view of external appearance and the results of various tests. Because the decision had been made in

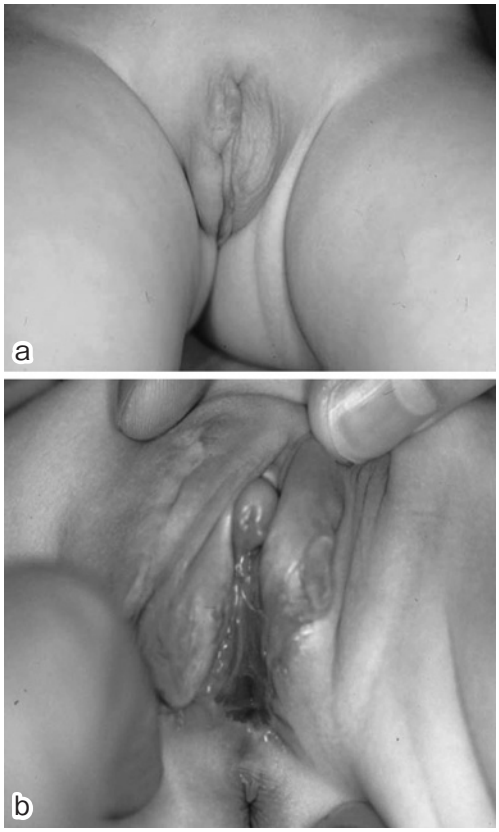

Fig. 4 1 year after operation
a: Appearance of the external genitalia
b: Appearance of the clitoris, urethral meatus and vaginal ostium.

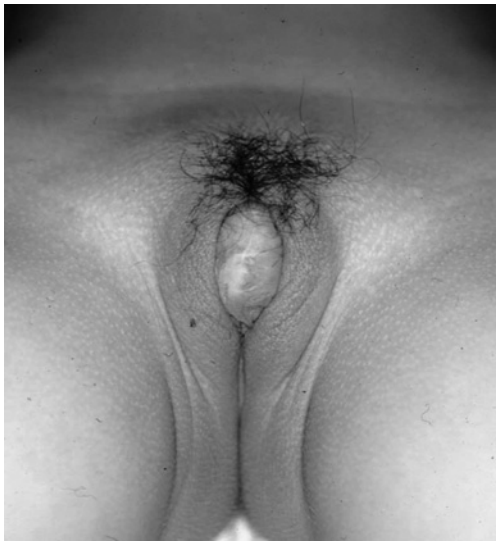

Fig. 5 Preoperative view

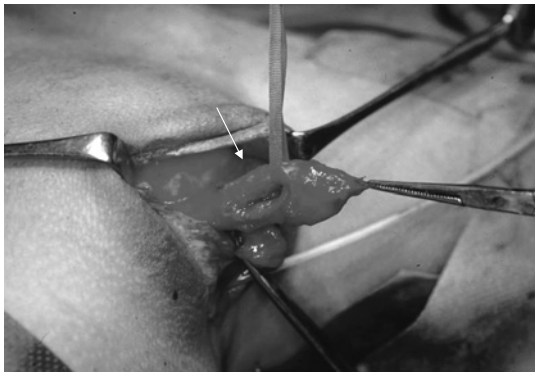

Fig. 6 Intraoperative view (**arrow**: neurovascular bundle)

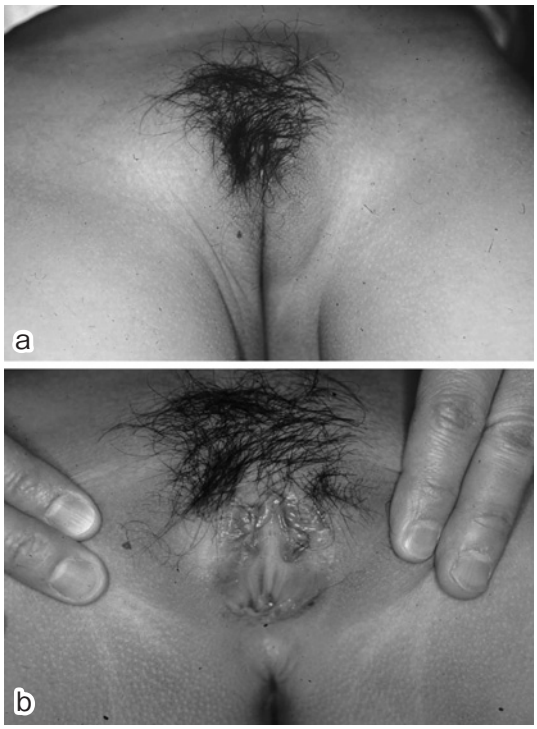

Fig. 7 Two years after operation
a: Appearance of the external genitalia
b: Appearance of the clitoris.

advance that the female sex would be selected even if the child were an XX-male, which remained a possibility until the results of the rapid intraoperative histopathological tests were received, no problems regarding sex selection were encountered. However, if, for example, the patient had been recorded in the family register as a boy with severe hypospadius, problems could well have occurred.

Reduction clitoroplasty with preservation of the neurovascular bundle was selected as the operative procedure not only for the sake of clitoris formation and sensitivity, but also because we believed it would achieve the best possible appearance of the external genitalia overall. A precondition for this

decision was the use of a surgical microscope to aid in separating the neurovascular bundle from the clitoral shaft to overcome the usual shortcomings of this method. The urogenital sinus was about 5 mm long, and a cut-back procedure was selected to form the vagina because a low vaginal entry had been confirmed preoperatively by intravenous urography.

In patient 2, because congenital adrenal hyperplasia had already been diagnosed, there were no questions about which sex to select. Because surgery had been performed twice previously, we decided to perform reduction clitoroplasty only after confirming with Doppler flowmetry that the dorsal artery of the clitoral shaft had been preserved. Detachment of the neurovascular bundle was performed under a surgical microscope, as with patient 1.

Three main procedures are available for clitoroplasty: clitorectomy, clitoral recession, and reduction clitoroplasty[3–6]. Each has advantages and disadvantages. Clitoral recession, which had previously been performed for our second patient, is a safe procedure, but it had not sufficiently reduced the size of the clitoris or its shaft. As a result, further surgery involving reduction clitoroplasty was needed. It is possible that after the previous operation ineffective hormone therapy had led to further hypertrophy of the residual clitoris, but intraoperative findings did not rule out the possibility that the clitoris had not been adequately reduced in size by the operation itself. Classic reduction clitoroplasty always results in sensory loss in the clitoris. Reduction clitoroplasty with preservation of the neurovascular bundle is a difficult procedure and is also considered to pose a high risk of sensory and circulatory disorders of the clitoris[7]. We found, however, that the procedure is safe when a surgical microscope is used to aid in the detachment of the neurovascular bundle from the clitoral shaft. We observed absolutely no clitoral circulation problems after surgery in either of our patients.

With regard to sensation, good postoperative circulation may be taken to indicate that sensory impairment has been minimized, although we could not confirm this in our patients because they were young children. It must be admitted that in the first patient the size of the clitoris was insufficiently reduced but could probably have been reduced further if both the reduction and the neurovascular bundle detachment had been performed under a surgical microscope.

Morphological abnormalities of the external genitalia can have a great effect on the psychological development of children. We believe that reduction clitoroplasty with preservation of the neurovascular bundle, which enables the best reconstruction of vulvar morphology with minimal loss of clitoral sensation, is the optimal choice for plastic surgeons who regularly perform microsurgery. Our experience with the two patients reported on here shows that microsurgery is useful for overcoming the disadvantages of this procedure and that, from a general perspective, reduction clitoroplasty with a surgical microscope to aid in detaching the neurovascular bundle is the best procedure for clitoroplasty.

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