Case Report

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Hysterectomy due to Abnormal Uterine Bleeding in a 15-year Old Girl with Rubinstein-Taybi Syndrome

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Rubinstein-Taybi syndrome is characterized by mental retardation, atypical facial features, broad thumbs and toes, and scoliosis. Polycystic ovaries are associated with chronic anovulation and abnormal uterine bleeding. A 15-year old female patient was diagnosed with Rubinstein-Taybi Syndrome, and had prolonged abnormal uterine bleeding for 2 years, accompanied by a polycystic ovary. As she showed no improvement during hormonal therapy or medical treatment, a hysterectomy was performed to control the bleeding.

Key Words: Hysterectomy, Polycystic ovary, Rubinstein-Taybi syndrome, Uterine bleeding

INTRODUCTION

Rubinstein-Taybi syndrome (RTS) is a disease caused by a chromosomal mutation that occurs in 1 in 100,000 live births. This syndrome includes congenital anomalies that primarily occur in a patient's face and limbs. Patients with RTS have craniofacial and thoracic anomalies, leading to difficult intubation. These features are accompanied by mental retardation, congenital cardiac disease, pulmonary structural anomalies, broad thumbs and big toes, and a high risk of aspiration pneumonia. Abnormal uterine bleeding is reported in approximately 14% of patients with RTS and may be classified as 'anovulatory' or 'ovulatory' bleeding [1]. Chronic anovulatory bleeding occurs irregularly and for a prolonged time; it is caused by factors such as polycystic ovaries, uncontrolled diabetes mellitus, thyroid dysfunction, and antipsychotic medications. Abnormal premenopausal uterine bleeding can be classified into three groups: irregular, non-cyclic, or prolonged/heavy menstrual bleeding. We report a case of a 15-year-old female with a history of RTS accompanied by polycystic ovaries. She presented with abnormal uterine bleeding not responsive to hormonal therapy, so a hysterectomy was performed to control the bleeding.

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CASE REPORT

A 15-year-old female patient weighing 50 kg with a height of 143 cm was admitted for abnormal uterine bleeding that persisted for two years. The patient was diagnosed with Rubinstein-Taybi syndrome when she was one year old. She displayed strabismus, preauricular fistula, and grade II mental retardation. She had a history of thoraco-lumbar spinal fusion to correct scoliosis (Fig. 1); there were no limitations to neck flexion or extension after the fusion. The patient experienced uterine bleeding that persisted for about

20 days every month for 2 years. Ultrasonography findings of the left ovary showed a polycystic ovary (Fig. 2). To treat her symptoms, hormonal treatment, including contraception pills and progesterone therapy, was administered, but there was no improvement in the control of the abnormal bleeding. As there were no other means to control this bleeding, we decided to perform a hysterectomy.

A preoperative ECG showed a premature atrial complex with prolonged QT. The CBC findings showed a hemoglobin level of 13.5 mg/dL and a hematocrit of 38.5%. Electrolyte levels showed a sodium level of 144 mEq and a potassium level of 4.5 mEq. The patient's nasal septum was deflected but there was no abnormality in the oral cavity, and her preoperative airway examination was classified as Mallampati class II. Upon arrival at the operating room, the patient was attached to an electrocardiogram, noninvasive blood pressure monitoring, and pulse oximetry monitoring. Before induction, her vital signs were 120/60 mmHg (blood pressure), SpO2 100%, and 120 beats/min (heart rate). She was preoxygenated for three minutes with 100% O2; then, 200 mg of pentothal sodium and 40 mg of rocuronium were administered via intravenous injection. After the intubation, sevoflurane was maintained at a rate of 2.5% and remifentanyl was infused at a rate of 0.2-0.5 μg/kg/min. During the operation, vital signs were stable with no specific incidents. Laparoscopic hysterectomy along with fistulectomy of the preauricular fistula was performed; total operation time was one hour and forty five minutes. The patient was discharged 5 days later without further complications.

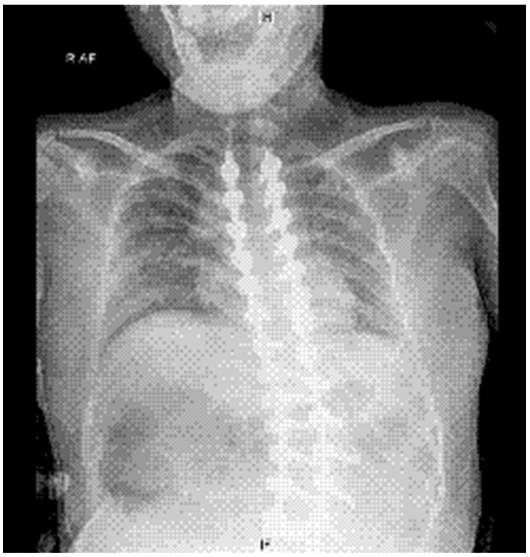

Fig. 1. Preoperative chest PA shows vertebral fusion at the thoraco-lumbar area.

DISCUSSION

Menorrhagia is a condition in which heavy menstrual bleeding (a blood loss greater than 80 mL) persists for several weeks. It is common in premenopausal women. Patients with menstrual bleeding greater than 80 mL are likely to be anemic; however, even if the hemoglobin level is within the normal range, menorrhagia cannot be excluded (sensitivity 43%, specificity 94%) [2]. Menorrhagia primarily occurs due to an intracavitary abnormality, but may also occur without such abnormalities. Excessive, prolonged, and frequent bleeding without an intracavitary structural abnormality is known as dysfunctional uterine bleeding (DUB).

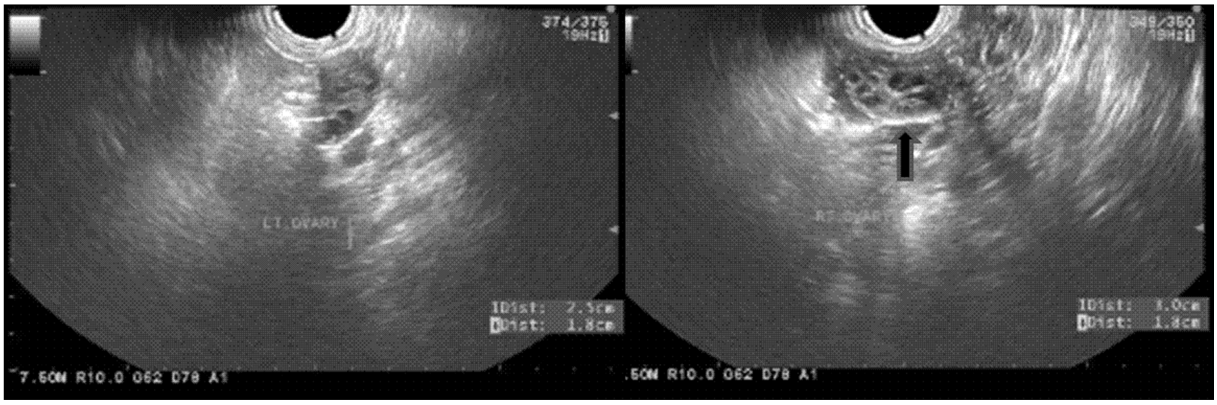

Fig. 2. Ultrasonography shows multiple cysts at the right ovary (arrow).

Ovulatory bleeding comprises 80% of DUB, preceding ovulation and occurring with regular menstruation. Anovulatory DUB usually occurs during the beginning and the end of the reproductive years; patients with anovulatory DUB have irregular menstrual cycles and persistent heavy bleeding.

Treatment for dysfunctional uterine bleeding includes the anti-fibrinolytic tranexamic acid, anti-inflammatory drugs, combined contraception pills, and progesterone administration. Our patient received oral contraceptives and progesterone, but there was no improvement in her bleeding. Surgical methods include endometrial ablation and hysterectomy. Endometrial ablation is a procedure that removes the basal layer of the endometrium, thus controlling uterine bleeding. The definitive treatment for controlling uterine bleeding is hysterectomy.

The cause of polycystic ovary syndrome (PCOS) is not definite, but it is a common endocrine disorder that occurs in 5-8% of patients of reproductive age [3]. PCOS is characterized by features such as hirsutism and irregular menstrual bleeding caused by ovarian androgen production and chronic anovulation. Irregular menstrual bleeding that does not persist for a long time is insignificant; however, if the duration is prolonged, as in our patient, complications such as anemia may occur. PCOS associated with chronic anovulation is known to occur in 6-10% of women with anovulation [4].

RTS is frequently accompanied by orthopedic, ophthalmic, cardiac, and orthodontic disease requiring surgical correction. Our patient had a history of thoraco-lumbar spinal fusion to revise scoliosis and required surgery to correct strabismus and a preauricular fistula. Patients with RTS usually have orofacial deformities, which may lead to difficult tracheal intubation. As our patient had no orofacial deformities or limitations to movement in the cervical spine, there were no difficulties in tracheal intubation. There were also no pulmonary complications. Prolonged uterine bleeding, along with the patient's mental condition (mental retardation grade II), made it harder to manage her uterine bleeding. Therefore, a prophylactic hysterectomy was considered.

RTS is an uncommon genetic disorder that is associated with mental retardation and many congenital anomalies. If such patients with a polycystic ovary have prolonged, irregular, abnormal uterine bleeding, medical treatment or ablation may be the first-line treatment choice. However, if there is an unsatisfactory response to those therapies, a hysterectomy may be effective.

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