

## CASE REPORT

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### ADNEXAL TORSION FOLLOWING GONADOTROPIN-RELEASING HORMONE ANALOG THERAPY: A CASE REPORT

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Adnexal torsion may occur in girls and adolescents. Often it is associated with ovarian diseases resulting in ovarian enlargement. Adnexal torsion may involve the ovary, fallopian tube or both, and the main symptom is acute pelvic pain.

An 8-year-old girl complaining of acute pelvic and abdominal pain, who was previously diagnosed with precocious puberty and who received treatment with a GnRH analog, is reported.

Ultrasound demonstrated a normal-sized uterus and bilaterally enlarged ovaries with multiple internal cysts. At laparotomy, we found a complete torsion in the right adnexa.

The histological examination revealed massive edema associated with multiple antral follicles and reduction of the follicular reserve.

**KEY WORDS: Ovarian neoplasms. Adnexal torsion. Precocious puberty. GnRH analog.**

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Ovarian torsion is an uncommon cause of abdominal pain in children and teenagers. However, in several series of children with ovarian masses, torsion was at least a component of the final diagnosis in 5 to 17 percent of the cases. Torsion of the ovary initially interferes with venous and lymphatic circulation, so there is sudden growth of the adnexa because of congestion and edema. If the torsion is unrelieved, the arterial supply may eventually be compromised—a condition that can lead to ovarian necrosis and peritonitis, mandating early surgical intervention<sup>2</sup>.

Adnexal torsion may involve the ovary, fallopian tube,<sup>2</sup> or both. Ovarian enlargement of any cause is a predisposing condition for adnexal torsion. It may occur in any age group, including prepubertal girls,<sup>2-5</sup> and the

diagnosis must be included in the differential diagnosis for girls and teenagers with acute pelvic pain.

The right side is affected more frequently than the left<sup>2</sup>. Because symptoms are nonspecific, the diagnosis may be confused with pelvic inflammatory disease or appendicitis.<sup>2</sup> Nausea and vomiting are frequent symptoms, and fever and leukocytosis are occasionally present.

A case of adnexal torsion following GnRH analog treatment for pre-

ocious puberty in a patient with unsuspected McCune-Albright syndrome is reported.

#### CASE REPORT

An eight year girl, height 145 cm, was referred to our hospital complaining of acute pelvic and abdominal pain. There was no history of fever, urination changes, weight loss, or diarrhea. In the previous 12 hours she had presented some episodes of nausea and vomiting.

The mother reported that the girl had experienced breast development since the age of 4.

Five months previously, serum estradiol had been measured at 23 pg/mL. The diagnosis of precocious pu-

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berly was made, and the patient was treated with leuprolide acetate 3.6 mg once a month. Three days after the third injection of leuprolide, the patient developed abdominal pain, nausea and vomiting, and abdominal enlargement, and she was then referred to our institution. At that time, an ultrasound exam revealed bilateral ovarian enlargement.

On physical examination, she had a protuberant pelvic mass that extended above the umbilicus as well as diffuse café-au-lait spots in the forearm and tibial regions. With these findings, we suspected McCune-Albright syndrome complicated with adnexal torsion. Ultrasound reveals a uterine volume of 28 cm<sup>3</sup>, a right adnexal mass of 18 cm in diameter and volume of 857 cm<sup>3</sup>, and multiple internal cysts.

The patient underwent laparotomy. The right adnexa had a complete torsion (Figure 1), and the right ovary presented areas of necrosis and multiple cysts (Figure 2). The left ovary was also enlarged (about 7 x 5 x 4 cm) with multiple cysts, but without torsion or areas of necrosis. A right salpingo-oophorectomy was performed. The histological examination revealed massive edema associated with multiple antral follicles and reduction of the follicular reserve. The patient recovered well from the surgery.

One month later, a follow-up ultrasound exam revealed that the remaining left ovary was still enlarged, and we chose to treat her with medroxyprogesterone 150 mg, intravenously, every 2 months for the next 2 years. During this period, the patient presented 2 episodes of uterine bleeding. At the age of 10, the medroxyprogesterone regime was interrupted, and the patient experienced 1 year of amenorrhea.

At the age of 11, irregular menses began, and we started a regime of norethindrone acetate from the 15th to 24th day of the menstrual cycle. Two years later, the patient has eumenorrhea and continues with this regime. She is 155 cm in height and weighs 52 kg.

## DISCUSSION

McCune-Albright syndrome is characterized by irregular café-au-lait spots, polyostotic fibrous dysplasia, and GnRH-independent precocious puberty. It is caused by a mutation in the alpha-subunit of the stimulatory G-protein, producing constitutive activation.<sup>2</sup> Mutations of the GS-alpha subunit of the G protein, which couples extracellular hormonal signals to the activation of adenylate cyclase, are

responsible for the autonomous hyperfunction of the endocrine glands and, presumably, for the other defects present in this disorder.<sup>2</sup>

Sexual maturation in these instances may be due to extrapituitary secretion of human chorionic gonadotropin (HCG) or sex steroid secretion independent of hypothalamic-pituitary gonadotropin stimulation.

Girls develop sexual precocity as a result of functioning ovarian cysts. Breast development and vaginal bleeding, which may be periodic, are related to intermittent increases in estradiol levels in association with 1 or more ovarian cysts. LH and FSH levels are prepubertal. Excessive hormone production by the thyroid, adrenal, and parathyroid may occur. The diagnosis is made on the basis of skin pigmentation and demonstration of bone lesions or pathologic fractures. Treatment of the McCune-Albright syndrome involves use of testolactone, spironolactone, and ketoconazole.<sup>2</sup>

In spite of the rarity of this syndrome, it represents almost 10% of the precocious puberty cases. The bilateral increase of the ovarian volume can result in adnexal torsion, abdominal pain, hemorrhage, necrosis of the ovary, and loss of the gonads. The use of a GnRH analog, does not relieve the symptoms. At first, the GnRH analog

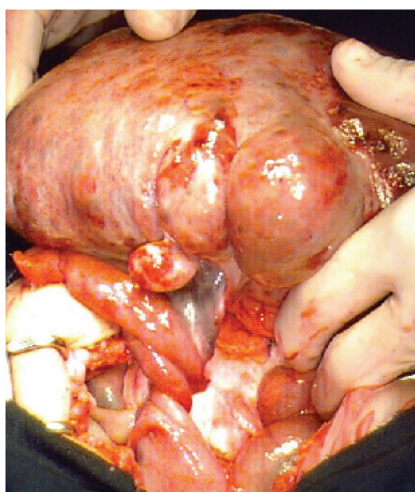


Figure 1 - Right adnexa with torsion.

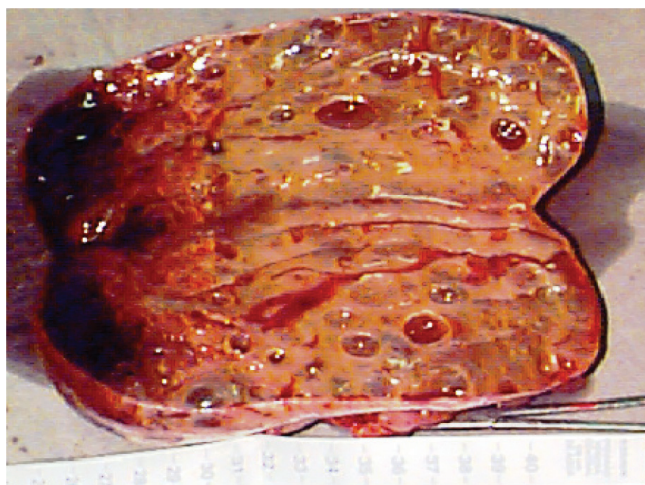


Figure 2 - The enlarged ovary with areas of necrosis and multiple cysts.

may stimulate FSH and LH secretion, resulting in ovarian stimulation, cyst formation, increase of volume and adnexal torsion.

The use of 150 mg medroxyprogesterone acetate every 2 months until the expected beginning of the menses and a regime of norethindrone acetate from the 15th to 24th day of the menstrual cycle after the establish-

ment of the periods seems to be a good option for treatment.

### CONCLUSION

McCune-Albright syndrome is a very rare disease characterized by irregular café-au-lait spots, polyostotic fibrous dysplasia, and GnRH-inde-

pendent precocious puberty. The enlarged ovaries predispose the individual to develop adnexal torsion. It is very important to recognize this syndrome before GnRH analog treatment for precocious puberty because treatment with a GnRH analog can increase the risk of adnexal torsion by the initial stimulatory effect of this drug on the ovaries.

### RESUMO

CARVALHO JP e col. Torção Anexial após tratamento da Síndrome de McCune-Albright com Agonista de GnRH. Relato de Caso. **Rev. Hosp. Clin. Fac. Med. S. Paulo** 59(3): 128-130, 2004.

Torção anexial pode ocorrer em crianças e adolescentes do sexo feminino. Frequentemente está associada com doenças ovarianas que resultam em cres-

cimento da gônada. A torção anexial pode comprometer os ovários isoladamente, as tubas uterinas ou ambos e o sintoma principal é dor pélvica aguda.

Descrevemos um caso de dor pélvica aguda em uma menina de 8 anos de idade, com diagnóstico prévio de puberdade precoce e que estava em tratamento com análogo de GnRH.

O exame ultra-sonográfico demonstrava útero de tamanho normal

com ovários aumentados bilateralmente e múltiplos cistos. Na laparotomia foi encontrado torção completa do anexo direito. O exame histológico demonstrou edema maciço de ovário associado com múltiplos cistos antrais e redução da reserva folicular.

**UNITERMOS: Neoplasia de ovário. Torção anexial. Puberdade precoce. Agonista de GnRH. Cirurgia.**

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