Case Report

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Management of cyclical pelvic pain by multiple ultrasound-guided superior hypogastric plexus blocks in a rare case of Mayer-Rokitansky-Küster-Hauser syndrome - A case series of three blocks in a patient

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Summary **Mayer-Rokitansky-Küster-Hauser syndrome is an uncommon disorder of mullerian agenesis where patients face multiple challenges like difficulty or inability to conceive and have sexual intercourse and chronic abdominal pain. This is a case report of a patient with Mayer-Rokitansky-Küster-Hauser syndrome who presented to the pain clinic with severe cyclical pelvic pain unresponsive to conservative treatment. This case was successfully managed with three ultrasound-guided superior hypogastric plexus blocks. This case illustrates that acute pelvic pain in MKRS patients can be effectively treated with bedside ultrasound-guided superior hypogastric plexus blocks. However, a GnRh analogue or hysterectomy is recommended for definitive treatment.**

Keywords: Superior hypogastric plexus block, ultrasound, Mayer-Rokitansky-Kuster-Hauser syndrome, pelvic pain

1. Introduction

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is an uncommon disorder of mullerian agenesis with a reported incidence of 1 in 4,500 live births and is diagnosed when a female presents with primary amenorrhoea with normal secondary sexual characteristics and a normal female genotype around puberty (*1*). It is caused by embryologic mullerian agenesis or atresia of the vagina, uterus, or both. MRKH is classified as type I or type II (*2*). MRKH type I is restricted to abnormalities of the reproductive system whereas MRKH type II has associated system anomalies (*2*). Patients face multiple challenges like difficulty or inability to conceive and have sexual intercourse; if rudimentary mullerian structures with active endometrium are present, they experience severe cyclical or chronic abdominal pain due to ovulation or endometriosis.

According to searches of PubMed and Google Scholar, management of abdominal pain in these patients has not been previously described. Reported here is the case of a patient with MRKH who presented to the pain clinic with severe cyclical pelvic pain unresponsive to conservative treatment. This case was successfully managed with repeated ultrasound-guided superior hypogastric plexus blocks.

2. Case Report

A 27-year-old married nulliparous female with a weight of 50 kg and height of 155 cm who had previously been diagnosed with MRKH type II was referred to the pain clinic for management of pelvic pain. She had a history of cyclical pain of 3-4 days duration every month. The pain was severe (8-9) on a numerical rating scale (NRS), aching in nature, infraumblical, and radiated to the back and perineal region. Pain was not present in any other region of the body. There was no relief after oral antispasmodic and non-steroidal anti-inflammatory drugs and partial relief with intravenous diclofenac.

The patient's past history indicated that pubarche occurred at age 12 with primary amenorrhoea and

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complete development of secondary sexual characteristics. She underwent vaginal dilatation at another hospital a year earlier, after which she developed a rectovaginal fistula and subsequently underwent diversion colostomy. Contrast-enhanced computerized tomography (CECT) of the abdomen revealed an ectopic right kidney with a left renal cortical cyst and uterine agenesis with an absent right ovary. Routine blood tests including serum estradiol and levels of follicular stimulating hormone, luteinizing hormone, and testosterone were within normal limits.

The woman was seen as an outpatient by Gynecology at this Hospital for severe pelvic pain (8 on the numerical rating scale). Pain had started in the morning and was not relieved with oral pain medications or injectable diclofenac. She was referred to the pain clinic for further management. A bedside ultrasound-guided superior hypogastric plexus block was planned for immediate pain relief. An intravenous line was inserted, and routine monitors were attached. Her heart rate was 120 beats/minutes and blood pressure was 134/84 mmHg. The procedure was explained to the patient and consent was obtained. She was then placed in a supine position in an sterile environment. A 2-5 MHz curvilinear probe (FUJIFILM SonoSite Edge) was used to identify the division of the abdominal aorta into common iliac arteries, which occurs at the fifth vertebral body. After subcutaneous local anesthesia with 2% lignocaine, a 9-cm 22-gauge spinal needle was inserted 3 cm below the umbilicus *via* the out-of-plane technique until bony contact at the anterior part of the fifth lumbar vertebral body. After hitting the vertebral body, the needle was withdrawn 1-2 mm, and 20 ml of 0.2% ropivacaine and 30 ug of clonidine was given after negative aspiration. The uniform spread of each drug was confirmed sonographically. Post-procedure, pain on the NRS decreased to 1-2. Her pulse rate decreased to 88 beats/min and blood pressure was 110/70 mmHg. She was observed in the recovery room for two hours. Her vital signs remained stable. She was satisfied after the procedure and did not complain of excessive sedation or drowsiness. There was no bruising or soreness at the injection site.

She was discharged after two hours and remained pain free for the full month but was seen again after a month with pain of 8-9 on the NRS. The block was repeated using a similar procedure and dose (Figure 1 and Figure 2). She had immediate pain relief post-procedure. MRI revealed slight deviation of uterine tissue to the left with an endometrial thickness of 5.8 mm. The right ovary and atretic cervix and upper vagina were not evident. MRI findings differed from those of a previous MRI at another hospital that found no uterine tissue. She was advised to receive injectable leuprolide (a GnRH analogue) to decrease the plasma level of estradiol in order to provide longer relief of abdominal pain. After a month, she was seen again

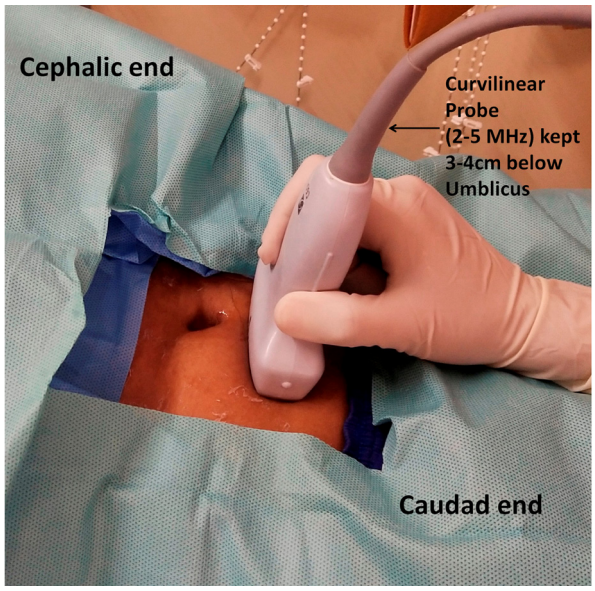

Figure 1. Curvilinear ultrasound probe (2-5MHz) kept in the transverse plane 3-4 cm below the umbilicus.

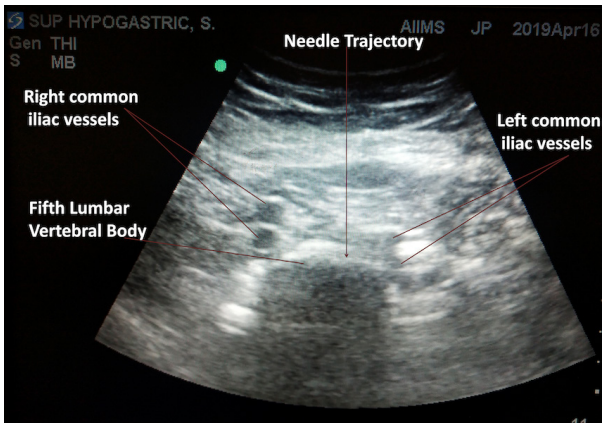

Figure 2. Transverse ultrasound image showing the body of the fifth lumbar vertebra along with the common iliac vessels and needle trajectory out-of-plane.

for a third time with pelvic pain of 7 on the NRS. A third block was similarly administered with ultrasound guidance and yielded excellent results. The patient has been followed up for the last three months and has been pain-free.

3. Discussion

MRKH syndrome is a rare condition with a reported incidence of 1:4,500. It represents the second most common cause of primary amenorrhea (*3,4*). MRKH syndrome is a complex malformation of complete agenesis of any of the structures derived from the paramesonephric müllerian duct (uterus, cervix, and upper 2/3 of the vagina) in females with a normal genotype, phenotype, and endocrine status (*5*). The typical syndrome (type I) is characterized by abnormalities restricted to the reproductive system. MRKH syndrome type II is associated with kidney

abnormalities in 40% of cases, with hearing problems in 10%, and with skeletal abnormalities in 10-12%. The most important step in the effective management of müllerian agenesis is correct diagnosis of the underlying condition, evaluation for associated congenital anomalies, and psychosocial counseling in addition to treatment or intervention to address the functional effects of genital anomalies. Abnormalities in the reproductive tract include a shortened vaginal canal, single midline uterine remnant, or uterine horns (with or without an endometrial cavity). The challenge is to recognize a remnant uterus and rudimentary müllerian structures, which are difficult to interpret on ultrasonography and may be particularly misleading before puberty (*6*). Thus, MRI should always be advised for assessment and should be interpreted by an expert radiologist since identification of uterine remnants is sometimes difficult, as it was in the current case (*7*). Severe abdominal pain is present in 70-80 % of patients. Abdominal pain in these patients is either because of endometriosis from retrograde menstruation due to obstructed uterine horns or monthly ovulation (*2*). Severe abdominal pain in patients with MRKH is described in the literature, but to the extent known no source has described pain management for those patients.

The superior hypogastric plexus (SHP) is located at the aortic bifurcation anterior to the peritoneum (*1,8*). The superior hypogastric plexus is an inferior continuation of the prevertebral sympathetic trunk and extends from the level of L-4 to S1. Primary visceral afferent nociceptive fibers from the upper vagina, cervix, uterus, fallopian tubes, bladder, and right colon travel through the SHP to the dorsal horn of the spinal cord. An SHP block (SHPB) is generally performed for relief of pain arising from lower abdominal structures (*9*). It is usually performed under fluoroscopy and is reported to be effective in relieving about 70-80% of pain (*9*). However, a special area is required for this. Other disadvantages include radiation hazards, discitis in a transdiscal approach, and inability to identify vascular and other peritoneal structures. Recently an ultrasound-guided technique for SHPB has been described for patients with cancer (*10*). The procedure was verified in a cadaveric study and confirmed using fluoroscopy (*11*). The advantages are that it is simple, fast, and non-invasive, it causes less pain since it is performed in the supine position, and it spares somatic nerve roots. To minimize the risk of injury to the bowel, patients are advised to take a tablet of dulcolax a day prior to the procedure. This same protocol was followed in the current case for every block except the first.

A previous report described successful management of interstitial cystitis in these patients *via* SHPB using pulsed radiofrequency (*12*). For chronic pelvic pain, ablation of the uterosacral nerve *via* a laparoscopic or open technique has also been reported (*13,14*). PubMed and Google Scholar were searched using the keywords "superior hypogastric plexus block," "ultrasound," and "chronic abdominal pain," but yielded no hits regarding SHPB for noncancer abdominal pain in females with MRKH syndrome. In the current case, cyclical abdominal pain did not respond to conventional oral or intravenous pain medications, but an SHPB provided complete pain relief. An immediate bedside ultrasound-guided superior hypogastric plexus block was performed since cyclical pain was present for only 2-3 days, and delay of another day would have been inappropriate in this patient.

Injectable leuprolide, a GnRh analogue, prolongs activation of GnRH receptors, which leads to desensitization and consequent suppression of gonadotrophin secretion and results in good pain relief when pain is secondary to endometriosis. However, inability to identify uterus remnants is common, as in the current case, so this treatment is frequently delayed. Definite treatment of cyclical abdominal pain is removal of the uterus, for which the current patient is scheduled.

In conclusion, acute pelvic pain in patients with MKRH syndrome can be effectively treated with a bedside ultrasound-guided superior hypogastric plexus block. However, a GnRh analogue or hysterectomy is recommended for definitive treatment.

Informed consent: Informed consent was obtained from the patient prior to publication of this article

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