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CASE REPORT

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A very rare case report of bilateral maldescended ovaries and müllerian duct anomaly associated with inflammatory myositis, myasthenia gravis and thymic pathology

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Abstract

Background: Maldescended ovaries are a rare condition. Despite its different embryologic development with the uterus, maldescended ovary is usually accompanied by uterine malformations and is found during the course of infertility. In other cases, it may be incidentally diagnosed in examinations due to abdominal pain or in a survey of finding paraneoplastic origin. Probable immune-related developmental conditions are associated with this abnormality; sometimes cross-reaction with other immune-related diseases is possible.

Case presentation: Here, the probable paraneoplastic origin is surveyed for a patient with coexisting inflammatory myositis and myasthenia gravis. According to this survey non recognized Mullerian duct and ovarian anomalies were found.

Conclusions: Knowledge about this anatomical abnormality is helpful for clinicians to prevent misdiagnosis and improper management. Moreover, understanding the probability of accompanying other conditions such as immune-related and neuromuscular junction disorders with Mullerian duct anomalies can offer a comprehensive insight.

Keywords: Congenital anomalies, Maldescended ovaries, Bicornuate uterus, Didelphis uterus, Müllerian duct anomaly, Inflammatory myositis, Myasthenia gravis, Paraneoplastic syndrome

Background

Maldescended ovary is a rare condition. Despite its various embryologic development with the uterus, it is usually accompanied by uterine malformations and can be often found during the course of infertility. This condition can be accidentally found in examinations to find the cause of abdominal pain or in a survey of finding paraneoplastic origin. Developmental abnormalities of the uterus, ovaries, and upper one-third segment of the vagina may cause abnormalities of the ducts. Normally positioned ovaries should be seen in the true pelvic, between the utero-ovarian and infundibulopelvic ligaments, otherwise, the term "maldescended" should be used [1]. Probable immune-related developmental conditions and sometimes cross-reaction with other immune-related diseases can be considered among the possible reasons for this condition [12].

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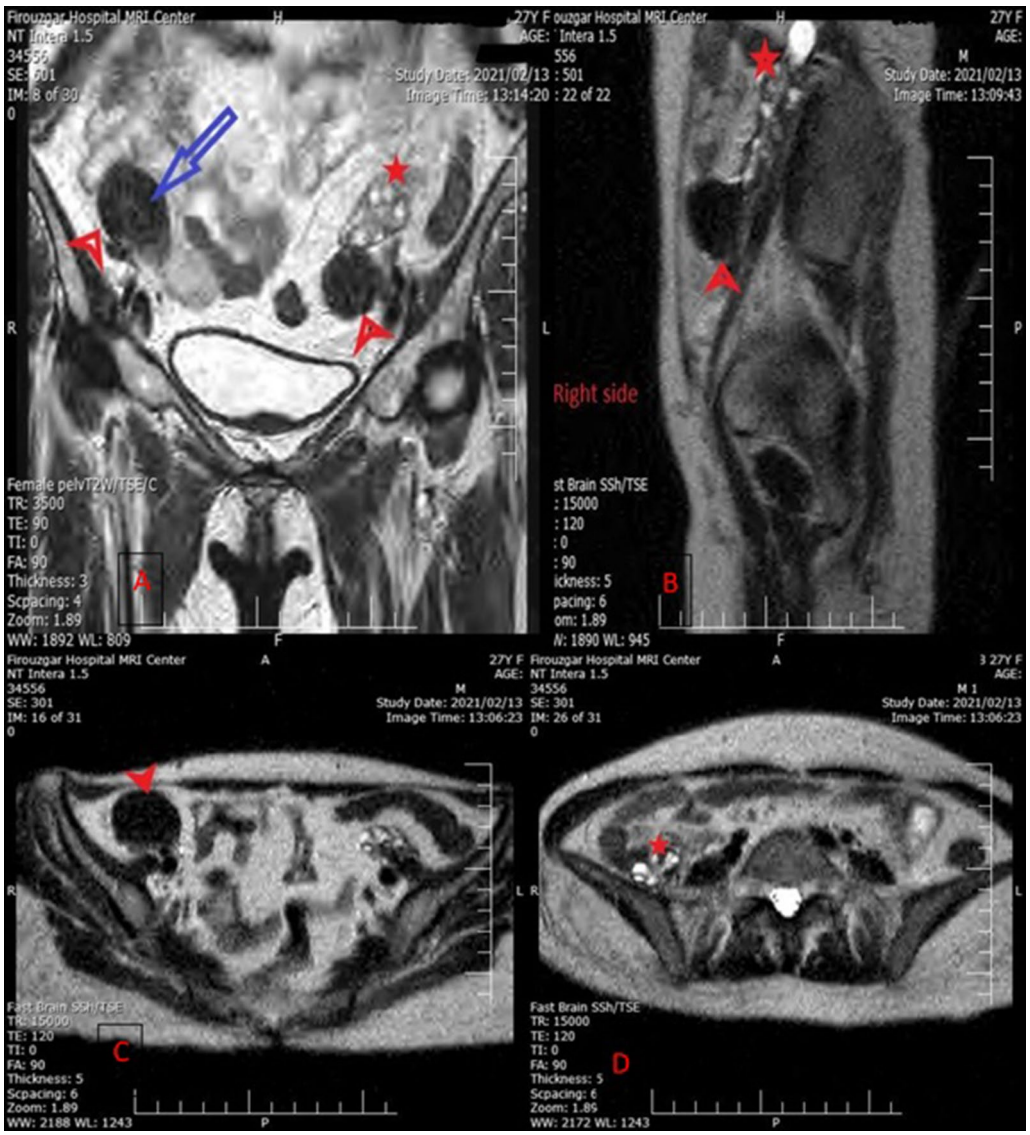

Fig. 1 Coronal and sagittal T2-weighted images in (**a**, **b**) show retroperitoneal low signal structure (red arrow head) under right iliac artery bifurcation with linear high signal endometrial component (blue arrow) in favor of completely separate uterus with separate hypoplastic cervix, Sagittal and axial T2-weighted images in (**b**, **d**) show retroperitoneal structure with a few intralesional cystic components with morphology similar to right ovary (*) in L5-S1 level, **c** right horn of didelphis uterus is shown in axial view (red arrow head)

Case presentation

A 27-year-old woman was admitted to the neurology department of Firouzgar Hospital of Tehran, Iran, with a fluctuating progressive pattern of proximal limb weakness and skin lesions starting from 10 months earlier. Clinical examinations indicated generalized proximal limb atrophy, cachexia, conjunctivitis, nail fold telangiectasia, and cutaneous lesions including upper limb rash with shawl sign pattern, heliotrope lesions, and Gottron papules. The patient had a history of proven myasthenia gravis from 3 years earlier with postsynaptic neuromuscular junction disorder in EMG which was treated with pyridostigmine for 3 years in addition to thymectomy surgery 2 years ago with proven type B pathology thymoma. After evaluating laboratory tests, the patient was considered to have dermatomyositis with recurrent myasthenia gravis.

The whole-body survey was carried out to detect probable tumoral lesions to rule out paraneoplastic syndromes. Brain and chest surveys showed no evidence of significant pathology or thymic hyperplasia or recurrence. Subsequent abdominopelvic CT scan and

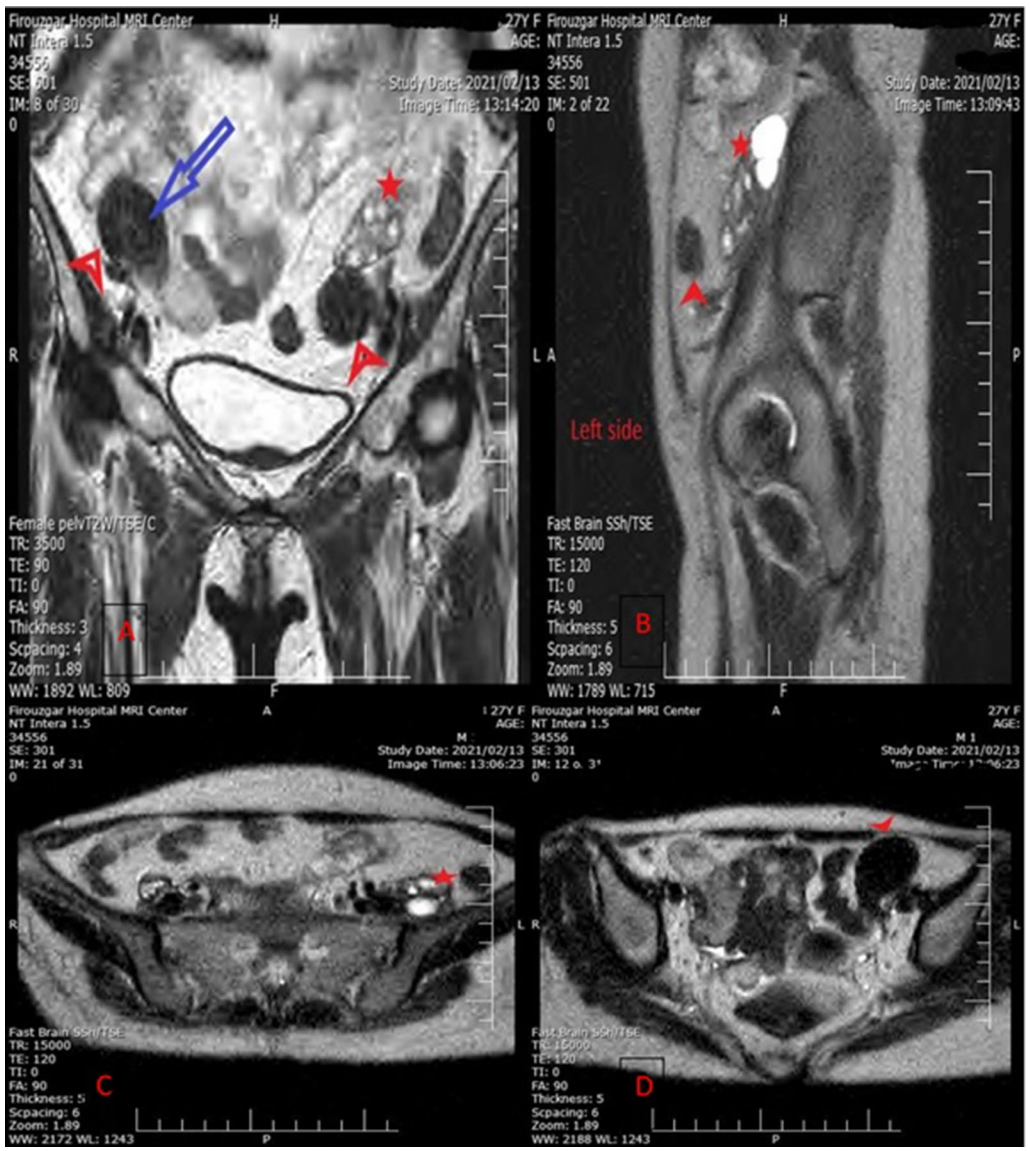

Fig. 2 Coronal and sagittal T2-weighted images in (**a**, **b**) show retroperitoneal low signal structure (red arrow head) under left iliac artery bifurcation, in favor of completely separate uterus with separate hypoplastic cervix, Coronal, sagittal and axial T2-weighted images in (**a**–**c**) show retroperitoneal structure with a few intralesional cystic components with morphology similar to left ovary (*) in pelvic inlet level, **d** left horn of didelphis uterus is shown in axial view (red arrow head)

supplementary pelvic MRI survey exhibited two discrete retroperitoneal structures with a few intralesional cystic components with morphology similar to ovaries at L5-S1 and pelvic inlet levels (Figs. 1, 2). Two discrete uteri with normal endometrial cavity and hypoplastic cervices were also observed in opposite sides of extraperitoneal space of pelvic region just below iliac artery bifurcation, higher than normal site (Figs. 1, 2). The cervices were connected into a single, midline hypoplastic vagina. The mentioned findings were in favor of the widely separated didelphys uterus. Considering her normal regular 28–30 days menstrual cycles, normal feminine appearance, sonographic evaluation, and a 3-year history of infertility, the observed retroperitoneal structures were considered to be bilateral undescended ovaries in the setting of Mullerian duct anomaly. After all, the patient received corticosteroid pulse therapy and was discharged with maintenance corticosteroid. She was recommended to take follow-up sonography in 6- to 12-month intervals for surveillance of probable malignant changes in ovaries due to probable association with the patient's setting.

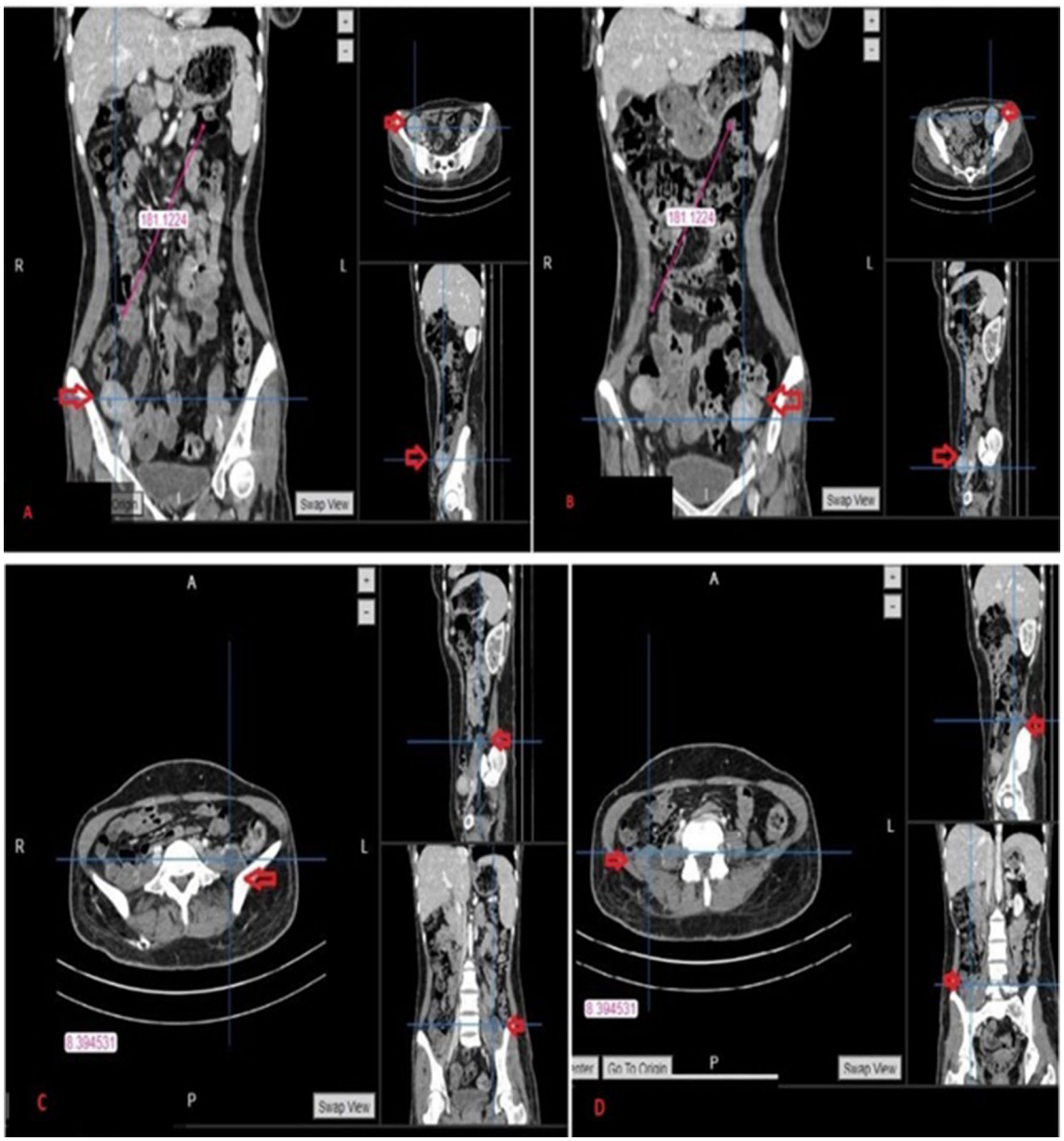

Fig. 3 CT scan with multi-planar reconstruction **a** indicating the right corn of didelphis uterus. CT scan with multi-planar reconstruction **b** indicating the left corn of didelphis uterus (all intended structures are indicated by arrow). CT scan with multi-planar reconstruction **c** indicating the left ovary in abnormal position, also CT scan with multi-planar reconstruction **d** indicating the right ovary in abnormal position (all intended structures is indicated by arrow)

Conclusions

This study presented a patient with dermatomyositis/myasthenia gravis accompanied by undiagnosed Müllerian duct anomaly and bilaterally undescended ovaries. This extremely rare case has not been reported before.

Maldescended single ovary is rare, and the occurrence of bilateral undescended ovaries is extremely rare. According to a mini-review by Jennifer E. Dietrich, the prevalence of maldescended ovaries is 0.3–2%. Seventy-three percent of the reported cases have concomitant Müllerian duct anomalies. The prevalence of bilateral undescended ovaries is 23% with an absolute count of 6 patients as noted in the mini-review [2]. Similar to other cases, an elongated fallopian tube related to the undescended ovaries was detected in this case [3, 4]. Migration disorders can occur both in the ovaries and uterus,

resulting in undescended ovaries and abnormal fusion of the uterus, respectively [5].

A didelphys uterus is formed when a fusion defect of the Müllerian ducts occurs. According to ESHRE/ESGE classification, there is a complete septate uterus with a hypoplastic dual cervix and a normal nonseptate vagina [6]. Our case had a didelphys uterus (Fig. 3) with undescended ovaries (Figs. 1, 2). Her medical history showed normal feminine appearance and menstruation with 3 years of infertility. No coexistent urothelial abnormality was found in this case, although according to other studies urothelial abnormalities are usually seen with Müllerian duct anomalies [4, 7].

The association between neuromuscular junction disorders and/or paraneoplastic syndromes is another interesting point of this case report.

Dermatomyositis coexistent with myasthenia gravis disease is a rare condition [8, 9]. According to Naohiro Uchio, most of the cases of dermatomyositis coexistent with myasthenia gravis had a history of thymoma (about 7 of 10 cases) [9]. The rare coexistence of dermatomyositis and myasthenia gravis is reported to be associated with immune-related adverse events clinically correlated with cardiac involvement and elevated serum CK levels [10, 11]. Association of Müllerian duct anomaly with myasthenia gravis was first reported in SAMUEL ARIAD report who presented a patient with malignant mixed müllerian tumor (MMMT) with sarcomatous differentiation and neuromuscular disorder as a result of the production of autoantibodies against nicotinic acetylcholine receptors in the nerve end-plate, although the diagnosis was made after abdominal metastasis [12].

In our case, dermatomyositis/myasthenia gravis coexistence with Müllerian duct anomaly and undescended ovaries was diagnosed with a history of type B thymoma resection, exertional dyspnea, and elevated creatinine kinase (CK) levels. At the moment we don't find any obvious tumoral growth in the Müllerian duct, but regarding the probability of non-overt and subclinical paraneoplastic features of the patient's condition, abdominopelvic sonographic follow-up surveillance was highly recommended.

This case report is written with the patient's conscious permission, and all personal information is kept confidential.

Abbreviations

EMG: Electromyography; MRI: Magnetic resonance imaging; ESHRE/ESGE: European Society of Human Reproduction and Embryology/European Society for Gynecological Endoscopy; CK: Creatine kinase; MMMT: Malignant mixed Mullerian tumor.

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Not applicable.

Authors' contributions

All authors contributed to the study conception and design. The first draft of the manuscript was written by NR, MAMV, RE and MA commented on previous versions of the manuscript. All authors read and approved the final manuscript.

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Availability of data and materials

All data generated or analyzed during this study are included in this article and are available at Firouzgar Clinical Research center (FCRDC), Iran University of Medical Sciences (IUMS), Tehran, Iran.

Declarations

Ethics approval and consent to participate

This manuscript is a human case report. Informative consent from the patient was obtained before writing the manuscript. Written informed consent was obtained from study participant.

Consent for publication

Written informed consent to publish this information was obtained from study participant. The Publisher has the permission to publish the relevant Contribution.

Competing interests

The authors declare that they have no competing interests.

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