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Case Report

Primary ovarian leiomyoma: Imaging in a rare entity

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ABSTRACT

Primary ovarian leiomyoma is a very rare benign mesenchymal tumor arising from the smooth muscle of walls of ovarian blood vessels. It is usually seen between 20 65 years of age. Being asymptomatic in many patients, these are incidentally detected. Ultrasonography and magnetic resonance imaging are preferred modality while imaging these lesions. Hereby we present a case of a 35-year-old female with incidentally detected right ovarian mass lesion which was hypointense on ultrasonography, hypointense on both T1W and T2W images, and on histopathology confirmed as primary ovarian leiomyoma. T1- and T2-weighted hypointensity on MRI with early homogenous postcontrast enhancement help in its diagnosis, though many a time it is difficult to differentiate it from other mesenchymal fibrous tumors such as fibroma and fibrothecoma. Histopathology and immunohistochemistry remain the mainstay in final confirmatory diagnosis. It is important to keep this entity in the differential diagnosis of solid T1 and T2 hypointense lesions of the ovary.

Introduction

Primary leiomyoma/ fibroid of the ovary is a very rare benign mesenchymal tumor usually seen between 20 and 65 years of age and is usually incidentally detected. These lesions arise from the smooth muscle of walls of ovarian blood vessels. Ultrasonography and magnetic resonance imaging (MRI) are preferred modality while imaging these lesions. T1- and T2-weighted hypointensity on MRI with early homogenous postcontrast enhancement can help in its diagnosis, though many a time it is difficult to differentiate it from other mes-

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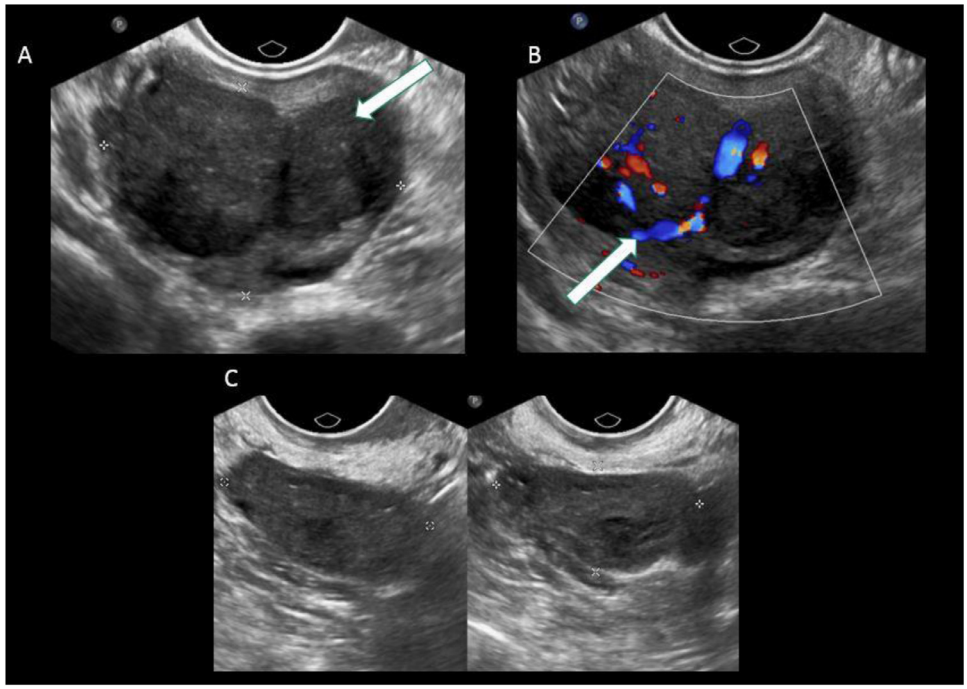

Fig. 1 – (A) Ultrasonography image showing a well-defined lobulated hypoechoic lesion (white arrow) in right ovary with displacement of normal ovarian parenchyma. (B) Doppler image showing predominant peripheral vascularity (white arrow) on Doppler study. (C) Ultrasonography image showing normal left ovary in 2 dimensions.

enchymal fibrous tumors such as fibroma and fibrothecoma. Histopathology and immunohistochemistry remain the mainstay in final confirmatory diagnosis.

Case report

A 35-year-old female presented with the missed menstrual cycle. Urine pregnancy test (UPT) was faintly positive. There was no history of bleeding per vaginum, abdominal pain or fever. The patient was referred to the imaging department to rule out missed abortion. Systemic examinations were within normal limits.

Ultrasonography of pelvis was done which revealed a well-defined solid hypoechoic lesion measuring 4.5 × 3.0 × 4.3 cm in the right adnexa. The right ovary was not seen separately from this lesion. Ovarian tissue was seen displaced laterally by the mass lesion. Doppler study showed predominantly peripheral vascularity (Fig. 1). These features were concerning for benign ovarian lesions with predominant fibrous stroma. The left ovary was normal in size and showed the presence of tiny follicles/cysts arranged peripherally around central echogenic stroma. The uterus was anteverted in position, normal in size and shape. An Lower (uterine) segment caesarean section (LSCS) scar was noted. The myometrial echoes appeared normal.

For further characterization of the lesion MRI was done. There was solitary well-defined lobulated T1 and T2 hypointense lesion in the right ovary. Medial displacement of the normal ovarian parenchyma containing multiple tiny T2 hyperintense follicles was seen. On postcontrast evaluation, this lesion showed similar homogenous enhancement as that of the myometrium (Figs. 2, 3). Based on these imaging findings diagnosis of ovarian fibroma was given with leiomyoma as a potential differential.

The patient underwent laparoscopic removal of the mass with the preservation of ovarian tissue (Fig. 4). Histopathologic evaluation showed the predominant presence of smooth muscle as well as spindle cells. Immunohistochemistry evaluation with alpha-smooth muscle actin (α-SMA) showed positive staining. These features were consistent with ovarian leiomyoma.

Discussion

Primary leiomyoma/ fibroid of the ovary is a very rare benign tumor accounting for about 0.5%-1% of all benign ovarian tumors. This entity was firstly described in 1862 by Sangralli [1]. It is usually seen between 20 and 65 years of age and is usually incidentally detected. Approximately 85% of these are discovered in perimenopausal and postmenopausal women together. Among this 16% are seen after menopause [1–4].

These lesions arise from the smooth muscle of walls of ovarian blood vessels. Other potential theories regarding tissue of origin include smooth muscle metaplasia of cortical

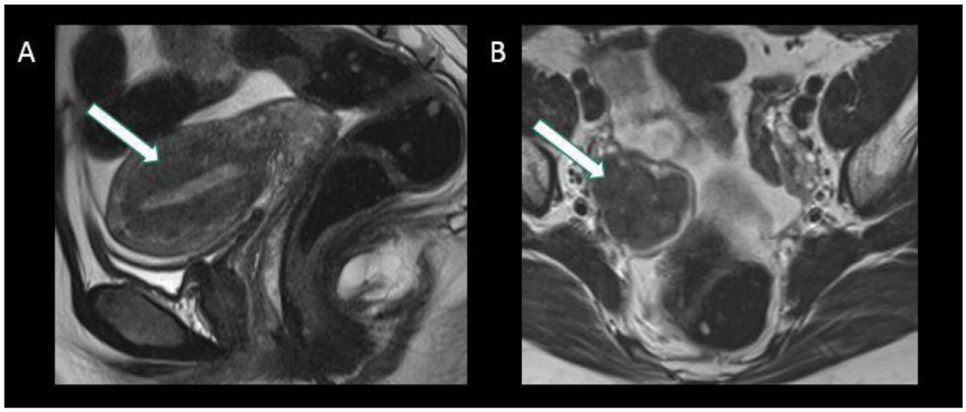

Fig. 2 – (A) Sagittal T2-weighted image showing anteverted uterus with normal endo-myometrial interface. (B) Axial T2-weighted image showing lobulated hypointense lesion in right ovary (white arrow) and normal left ovary (yellow arrow). Signal intensity of the lesion is same as that of the uterine myometrium.

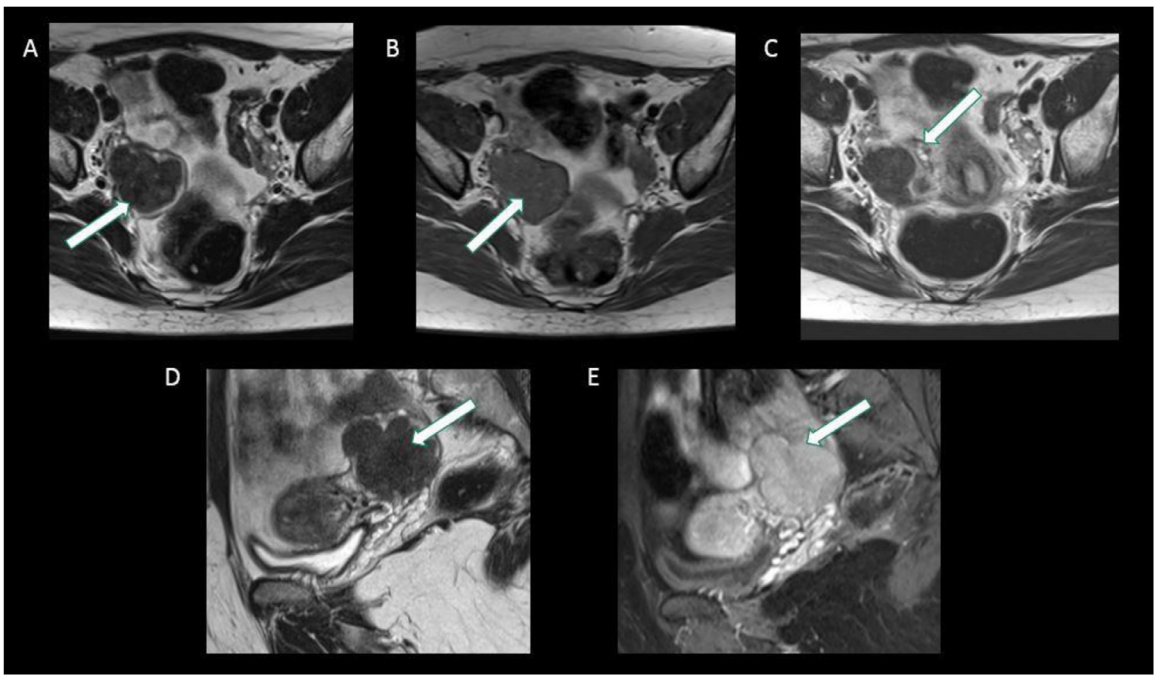

Fig. 3 – (A) Axial T2-weighted image showing well defined lobulated hypointense lesion (white arrow) in right ovary. (B). Axial T1-weighted images showing lobulated iso to hypointense lesion (white arrow) in right ovary. (C) Axial T2-weighted image showing medial displacement of the ovarian parenchyma containing multiple tiny T2 hyperintense follicles (white arrow). (D) Sagittal T2-weighted image showing defined lobulated hypointense lesion (white arrow) in right ovary. (E) Sagittal postcontrast image showing homogenous contrast enhancement of the lesion (white arrow) similar as that of uterine myometrium.

stroma, ovarian ligaments, and multipotent ovarian stromal cells [2,5].

Ovarian leiomyomas are usually unilateral, small in size measuring less than 3 cm in diameter. Associated uterine fibroids can also be identified. Bilaterality of these lesions is common in younger women and coexistence with uterine leiomyomas is usually not seen. Clinically many of the patients are asymptomatic; however, these can present with lower abdominal pain in few cases. Other associated features like Meig's syndrome secondary to ascites, hydrothorax, hydronephrosis, and marginally elevated levels of tumor marker (CA 125) are seen with giant leiomyomas. Secondary degeneration changes like hemorrhage, hyalinization, calcification, and cyst formation are more commonly seen in uterine leiomyomas as that of the ovarian in origin which is usually solid, [1,4–8].

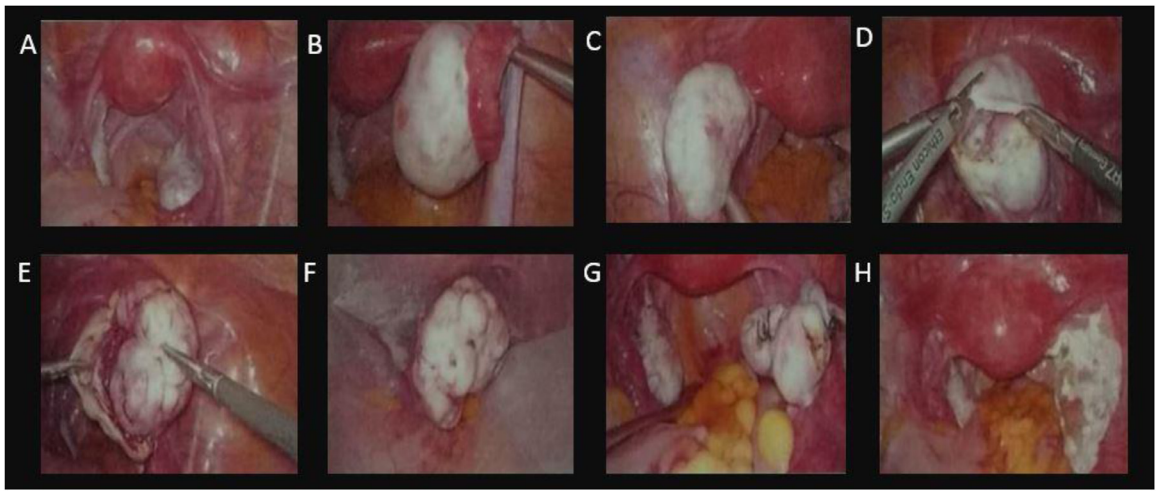

Fig. 4 – Enucleation of right ovarian mass. (A) Normal uterus, (B) normal left ovary, (C) right ovarian mass lesion. (D) Incision over right ovary. (E) Enucleation of right ovarian mass. (F) Right ovarian mass within endobag. (G) Right ovarian reconstruction. (H) Final view after intercede placement.

Ultrasonography due to its ready availability and MRI due to its high soft-tissue characterization are the primary modalities in the evaluation of ovarian lesions [5].

On ultrasonography, ovarian leiomyomas show similar appearance as that of the uterine ones and show echogenicity similar to the myometrium appearing heterogeneous with predominantly hypoechoic in echotexture. Minimal peripheral vascularity can be seen in the Doppler study [4,9,10].

On MRI, these lesions are solid appearing primarily. Primary ovarian origin of the lesion can be assessed with MRI by addressing the following points as ovarian tissue cannot be identified separately from the lesion. If present ovarian follicles can be seen surrounding the lesion. In our case, the ovarian origin of the lesion was confirmed as there was the displacement of the ovarian tissue with small follicles by the mass. Localization and tracking the course of gonadal vessels also helps [5].

Due to the presence of fibrous component, these appear hypointense on both T1- and T2-weighted images. Kobayashi et al. and Troiano et al. worked on the MRI appearance of the ovarian leiomyomas and described predominant T2 hypointense feature which is relatively specific for fibroma and fibrothecoma of ovarian origin [2,5–7].

Differential diagnoses to be considered in cases of T1 and T2 hypointense lesions of ovarian origin include fibroma, fibrothecoma, primary leiomyoma, thecoma, cystdenofibroma, Brenner tumor, and sclerosing stromal tumor. Intravenous leiomyomatosis and leiomyomatosis peritonealis disseminata also shows similar feature and can be considered among differentials in which there is secondary ovarian involvement [2,3,5–7].

Postcontrast evaluation with gadolinium contrast can help to differentiate between primary ovarian leiomyoma and fibroma, fibrothecoma, in which leiomyoma shows early contrast enhancement as that of the delayed and weak contrast enhancement seen in fibroma and fibrothecoma [5].

Imaging can help in narrowing down the differentials however final and confirm the diagnosis is based on the histopathologic and immunohistochemical evaluation. The macroscopic and microscopic appearance of the ovarian leiomyoma is similar to the uterine leiomyoma. It shows the presence of smooth muscle and spindle cells. These stains positively with alpha-smooth muscle actin (α-SMA) [2,3]].

Estrogen plays an important role in the overall development of the ovarian leiomyomas as these are more common in nulligravid women. A rapid increase in the size of these lesions during pregnancy and regression in the postpartum period can also be attributed to the effect of estrogen [2,6].

Surgical intervention is the choice of treatment. Various options include oophorectomy and unilateral salpingo-oophorectomy. Ovary preserving technique, that is, enucleation is preferred in young patients [2,5].

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