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Case Report

Twisted large cellular fibroma of the ovary with pelvic effusion-revisiting incomplete demons Meigs' syndrome

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ABSTRACT

Fibromas of the ovary are benign tumours arising from the stromal component of the ovary. They represent 1% of ovarian neoplasms and are generally benign. The cellular subtype accounts for around 10% of these and is a tumour of uncertain malignant potential. Recurrence may occur or it could be associated with peritoneal implants. Usually these are solid tumours, sometimes with small areas of cystic degeneration. Occasionally, large fibromas may undergo torsion causing acute abdominal pain and generally tends to occur in post-menopausal women. The solid areas, ORADS and ascites simulated a malignant ovarian tumour on both MRI, USG and gross pathologic examination. Here we present a case of a postmenopausal woman presented with abdominal pain and postprandial vomiting with a large twisted ovarian fibroma leading to diagnostic and therapeutic challenges masquerading as a malignant ovarian neoplasm.

Keywords: Fibroma, Cellular, Spindle cell, Ovary, Ascites, Solid, Twisted, Postmenopausal, Meigs

INTRODUCTION

Ovarian fibroma is the commonest benign tumour of the ovarian stroma (4% of all ovarian neoplasms), and it can occur at any age.[1] The cellular subtype exhibits hypercellularity, increased mitotic activity, and mild-to-moderate nuclear atypia. The average age is in the fifth and sixth decades. Seen bilateral in 4-8% of patients and giant fibromas can undergo torsion.[3] Gorlin's, Maffucci and Sotos syndromes have associated fibromas.[4,5] Although the subject had ascites, commonly found in women with ovarian fibroma, both serum CA 125 and MRI-Thorax did not show the presence of pleural effusion and hence incomplete Meigs in 66-year-old postmenopausal lady with a large twisted ovarian fibroma and pelvic effusion.

CASE REPORT

A 66-year-old post-menopausal, multiparous woman presented with a 10-day history of worsening abdominal pain started in the right lower quadrant and subsequently spread to the whole abdomen. In her past medical history were intermittent episodes of abdominal discomfort, postprandial vomiting and a sensation of abdominal heaviness during the preceding months. Her vital signs were all within normal limits. CA-125 was 38 U /ml.

Physical examination revealed a palpable abdominal mass in the lower abdomen and involuntary guarding. Vaginal examination revealed a normal sized uterus and a large irregular mass of 12 cm size, firm in consistency.

Pelvic ultrasonography showed a large complex mass (12×14 cm) in the abdominopelvic location converging to right adnexa with twisted pedicle.

MRI revealed a 15.2×11.2×8.5 right sized predominantly solid lesion with multiple variable sized cystic areas note in the pelvis, in the midline. Solid components of the lesion show T1 hypointense T2 heterogenously hyperintense

areas, postcontrast enhancement and diffuse restriction. Relatively largest cystic component 7.5×6.9×7.8 cm noted in the superior aspect of the lesion. Transient twisting of vascular pedicle of the lesion noted. Uterus 5.9×3.4×4.2 anteverted 1.2×1.5 cm fibroid noted. Endometrial cavity distended 1.4×2.1×1.8 cm polyp. Mild ascites noted extending to pelvis and upper abdomen. Bilateral ovaries not separately visualised. No pelvic or paraaortic lymphadenectomy. ORADS -MRI-4 (Figure 1 A and B).

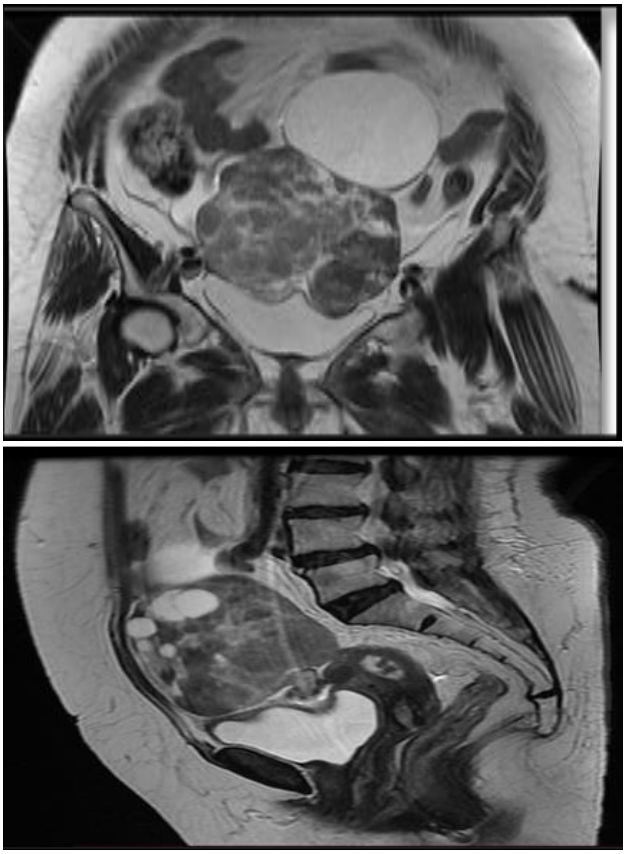

Figure 1 (A and B): MRI revealing an enlarged right ovary with predominantly solid lesion 15.2×11.2×8.5 cm predominantly solid lesion with multiple variable sized cystic areas noted midline in the pelvis. Solid components of the lesion show T1 hypointense T2 heterogenously hyperintense areas, postcontrast enhancement and diffuse restriction. Solid components of the lesion show T1 hypointense T2 heterogenously hyperintense areas, postcontrast enhancement and diffuse restriction. Relatively largest cystic component 75×6.9×7.8 cm noted in the superior aspect of the lesion.

MRI THORAX-no pleural space fluid (Figure 2).

An exploratory laparotomy had showed a black bluish encapsulated mass arising from the twisted right adnexa, measuring 150×110×80 mm with solid cystic irregular surface and attached to the right ovary with a once twisted pedicle (Figure 3 and 4). Both the uterus and left adnexa appeared atrophic. Total abdominal hysterectomy and bilateral salpingo-oophorectomy were performed. The operating time was about 110 min. There was no significant blood loss and no anaesthesia-related complications. Intra- or post-operative period was unremarkable and she was discharged on the sixth day following the surgery. She is still being followed up and there is no recurrence. Pathologic study shows gross pathologic findings: the ovarian tumour measured 15×12×10 cm and weighed 1230 gm, with smooth white external surface and absence of macroscopic rupture. According to pathological and immunohistochemical findings-positive for Inhibin and negative for desmin, the pathological diagnosis of ovarian cellular fibroma was confirmed (Figure 5-7). In addition presence of cellularity and < 4 mitoses under 10 HPF with no nuclear atypia. ruled out fibrosarcoma. Microscopically, the tumour showed alternating solid and cystic areas. Part of the solid areas were slightly hypercellular with nonatypical spindle cells arranged in storiform pattern, compatible with fibroma (Figure 4 B). Frequently small foci were observed with incipient cystic degeneration.

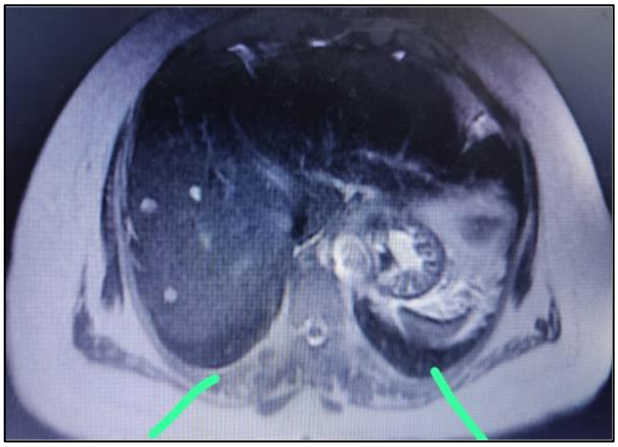

Figure 2: MRI-no evidence of pleural space fluid-dependent space is marked.

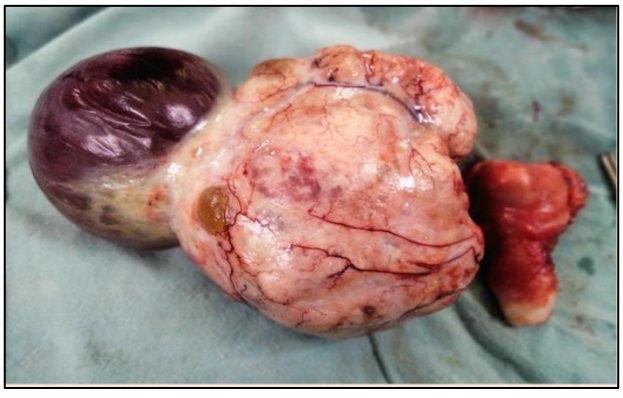

Figure 3: Intraoperative picture showing a black bluish encapsulated mass 1230 gm arising from the twisted right adnexa, measuring 150×110×80 mm with solid and cystic areas with largest cystic component 75×6.9×7.8 cm noted in the superior aspect of the lesion

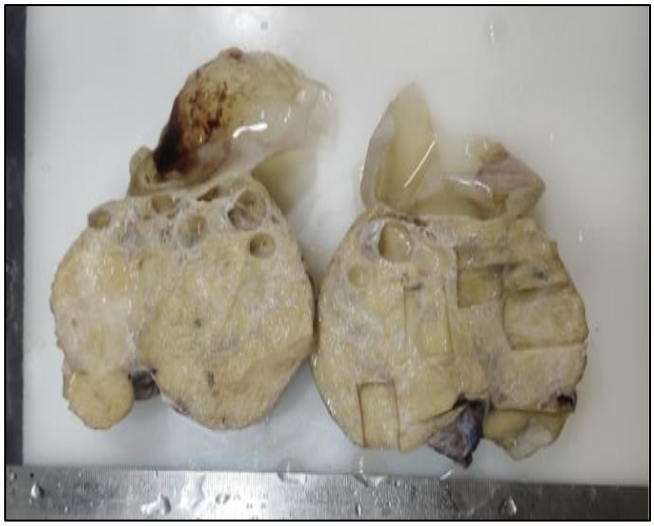

Figure 4: Macroscopic appearance of the resected ovarian fibroma.

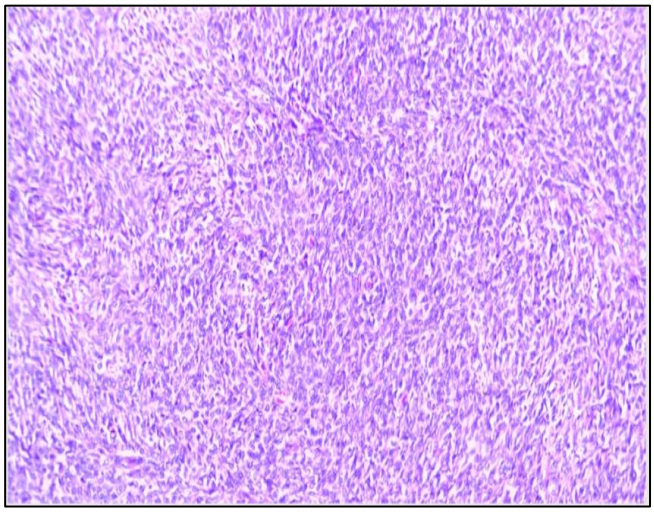

Figure 5: Histopathology of ovarian cellular fibroma showing benign spindle cell proliferation (X4) Representation of the solid area compatible with cellular fibroma, noting evident high cell density.

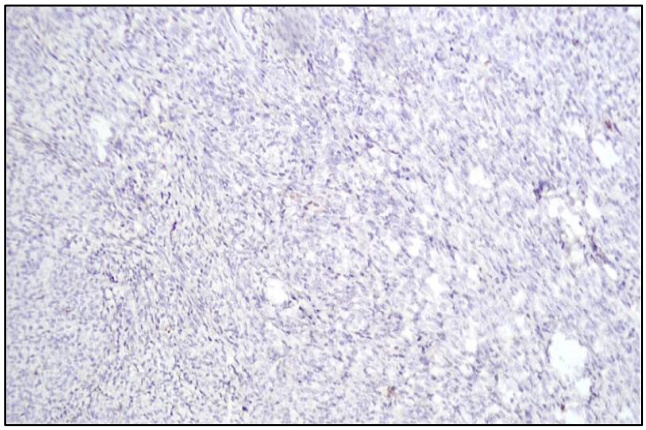

Figure 6: Immunostaining (400x) showing Desmin negative.

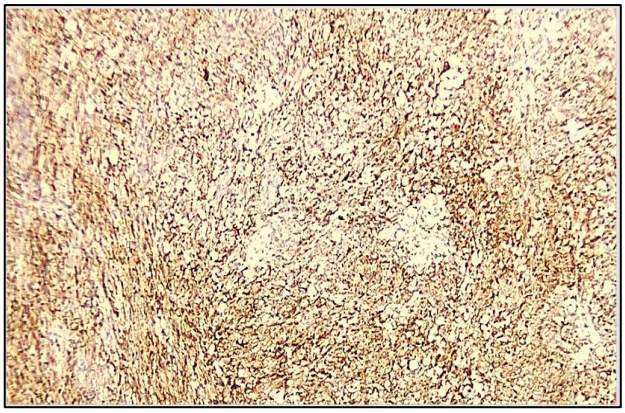

Figure 7: Immunostaining (400x) showing inhibin positive.

DISCUSSION

An ovarian fibroma, with pleural and ascitic fluid that disappears after removal of the tumour, is classified as Meigs' syndrome when the primary tumour is fibroma, thecoma, granulosa cell tumour, or Brenner tumour.[8] With other tumour types, such cases are defined as pseudo-Meigs' syndrome (PMS).[9] Generally, both pleural effusion and ascites are recognized, but there are reported atypical cases with only pleural effusion or only ascites, which called incomplete Meigs' syndrome.[9,10] Those who present with symptoms of systemic lupus erythematosus and high CA-125 in pleural effusion and ascites are called pseudo-pseudo Meigs' syndrome (PPMS)/ Tjalma syndrome.[11-13] Cellular fibromas contain one to three mitoses per 10 high-power fields but display ordinary nuclei and usually behave in a benign fashion.[7]

Although in the present case report, the subject had ascites, commonly found in women with ovarian fibroma, both serum CA 125 and MRI-Thorax did not show the presence of pleural effusion and hence the diagnosis of incomplete Meigs in 66-year-old postmenopausal lady with a large twisted ovarian fibroma and pelvic effusion.

		Meigs' syndrome related terms							
		Demons-Meigs' syndrome		Pseudo Meigs' syndrome		Incomplete Meigs' syndrome			Pseudo-pseudo Meigs' syndrome
	Diagnostic criteria	Classic Meigs' syndrome	Non-classic Meigs' syndrome	Benign pseudo Meigs' syndrome	Malignant pseudo Meigs' syndrome	Incomplete Demons Meigs' syndrome	Incomplete benign pseudo Meigs' syndrome	Incomplete malignant pseudo Meigs' syndrome	
TUMOR RELATED CRITERIA	Benign fibroma or fibroma-like ovarian tumor					OR			
	Benign ovarian or broad ligament tumor other than fibroma or fibroma- like								
	Benign extra-ovarian and extra broad ligament pelvic or abdominal tumor								
	Malignant pelvic or abdominal tumor								
NON-TUMOR RELATED CRITERIA	Ascites*					OR	OR	OR	
	Pleural effusion*								
	Resolution of ascites and pleural effusion after tumor removal								
	Systemic lupus erythematosus								
	Elevated serum CA-125 concentration								

*Without signs of malignant involvement. Please note that Demons–Meigs' syndrome includes all cases classified as classic Meigs' syndrome and nonclassic Meigs' syndrome, whereas incomplete Demons Meigs' syndrome combines all cases classified as incomplete classic Meigs' syndrome and incomplete nonclassic pseudo-Meigs' syndrome. Due to space limitations the subgroups of incomplete Demons–Meigs' syndrome were not presented on the diagram. CA-125 = carbohydrate antigen 125.

Figure 8: Meigs' syndrome related terms.

Acute abdomen due to torsion, heavy menstrual bleed, gastrointestinal, urinary symptoms fatigue/malaise could be the symptoms in decreasing frequency in cellular fibroma ovary.[14,15] Incidentally detected cases on physical examination, ultrasound, or intraoperatively (caesarean section, etc.) are also reported in literature.

Irritation of the peritoneal surface could explain the increased CA 125 levels associated with fibroma. Volume of ascites was positively associated with the expression level of CA-125 but not linearly associated with tumour dimension and it regressed following surgical removal of the tumour.[16-17]

Gross

Fibromas are usually solid, spherical, slightly lobulated, encapsulated, grey-white masses covered by a glistening, intact ovarian serosa. The cut surface of a fibroma is white and slightly spiralled and may manifest areas of cystic degeneration.[18]

Microscopy

Tumours consist of cellular bundles of fibroblastic tumour cells having spindle-shaped nuclei with no signs of atypia and intersecting strips of hyaline-appearing fibro-collagen tissue.[18] Cellular fibromas are benign with one to three mitoses per 10 high-power fields with ordinary nuclei.

Prognosis

Cellular fibromas with excellent prognosis may recur locally but are rare.

Variants

Fibro sarcomas have more than 4 mitoses, display nuclear atypia and they are malignant.[19] There are few cases with benign features but high mitosis count, named as "mitotically active cellular fibroma" (more than four mitotic figures) but behave as benign.[20-22]

Bilateral in 5% of cases, seldom more than 10-15 cm in maximum diameter, with only one third of them were smaller than 3 cm.[23]

Theories

Mechanical compression or partial torsion of fibromas or other benign ovarian solid tumours leads to the ascites production by the lymph released from the surface of the tumour.[24] Recently, a probable active secretion by the tumour of the growth factors mediating the hyperpermeability in the ovary or the peritoneal vessel was suggested.[25] Pleural effusion is thought to occur secondarily via congenital defects or diaphragmatic lymphatic vessels that tend to be common on the right side allowing ascitic fluid to pass through the diaphragm into the pleural cavity. Pleural effusion of PMS is often right unilateral, followed by bilateral. Pleural effusion was exudative in 70-80% cases.[26]

Sonography

Stripy shadows on USG can be explained by the cellular bundles and intersecting strips of hyaline-appearing collagen and fibrous tissue which are responsible for the 'spiral pattern' seen on the cut surface of typical fibromas on macroscopic examination The major difficulty is to differentiate between ovarian fibroma and other solid ovarian tumours.

Colour doppler findings are variable, but most fibromas manifest minimal to moderate vascularization.[28,29]

On CT, ovarian fibroma usually appears as homogeneous solid tumours with delayed enhancement. While, diagnostic criteria for torsion include an adnexal mass in the midline, rotated toward the contralateral side of the pelvis; deviation of the uterus to the side of the affected ovary and ascites.[30]

MRI is often needed for further differentiation of ovarian fibromas and fibro-thecomas from other solid ovarian masses, especially pedunculated or broad ligament leiomyomas.[31] Most ovarian fibromas were isointense to hypointense to uterine myometrium on T1- and T2-weighted images.[32] MRI features of fibromas depend on the size of the lesion. Fibromas larger than 6 cm were more likely to show the presence of capsule, degenerative changes, peripheral subcapsular cystic areas, heterogeneous T2 signal, and heterogeneous enhancement.[31] Correct diagnosis of these tumours on imaging can decrease patient anxiety and avoid unnecessary invasive procedures.

Diagnosis

It is confirmed only on histopathology. Ovarian fibromas must also be differentiated from leiomyosarcomas and fibrosarcomas.[33,34] To do so, pathologists use criteria such as the number of mitoses, cytologic atypia, and tumour necrosis. In our case, none of these criteria were detected.[2,7]

Immuno-histochemistry

Because of its rarity, the immunohistochemical analysis with Desmin, inhibin or histochemical staining with Masson´s trichrome is recommended to rule out the differential diagnosis especially leiomyosarcoma and sex-cord stromal tumours (thecoma and sclerosing stromal tumour)[32]

Surgical removal

Surgical removal of these solid ovarian tumours is recommended because of the low probability of malignancy and recurrence.[29] Cystectomy only can be

performed in young women while TAH with BSO the treatment of choice in elderly patients.[35-37]

CONCLUSION

Ovarian cellular fibromas are usually solid. Here, both the rate of mitosis (maximum 3 mitoses/10 HPF), inhibin immunohistochemistry along with alternating areas of low and high cellularity and non-aggressive clinical course clinches the diagnosis of cellular fibroma.

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REFERENCES

1. Kitajima K, Kaji Y, Sugimura K. Usual and unusual MRI findings of ovarian fibroma: correlation with pathologic findings. Magnetic Resonance Med Sci. 2008;7(1):43-8.
2. Prat J, Scully RE. Cellular fibromas and fibro-sarcomas of the ovary: a comparative clinicopathologic analysis of seventeen cases. Cancer. 1981;47(11):2663-70.
3. Macciò A, Madeddu C, Kotsonis P, Pietrangeli M, Paoletti AM. Large twisted ovarian fibroma associated with Meigs' syndrome, abdominal pain and severe anaemia treated by laparoscopic surgery. BMC Surg. 2014;14:38.
4. Howell CG, Jr, Rogers DA, Gable DS, Falls GD. Bilateral ovarian fibromas in children. J Pediatric Surg. 1990;25:690-1.
5. Christman JE, Ballon SC. Ovarian fibrosarcoma associated with Maffucci's syndrome. Gynecol Oncol 1990;37:290-91.
6. Chen CP, Yang YC, Lin SP, Wang W, Chang CL, Chang KM. Bilateral calcified ovarian fibromas in a patient with Sotos syndrome. Fertil Steril. 2002;77:1285-7.
7. Irving JA, Alkushi A, Young RH, Clement PB. Cellular fibromas of the ovary: a study of 75 cases including 40 mitotically active tumors emphasizing their distinction from fibrosarcoma. Am J Surgical Pathol. 2006;30(8):929-38.
8. Meigs JV. Fibroma of the ovary with ascites and hydrothorax: Meigs' syndrome. Am J Obstet Gynecol. 1954;67:962-85.
9. Hardy E. Meigs' syndrome and pseudo-Meigs' syndrome. J R Soc Med. 1987;80:537.
10. Hou YY, Peng L, Zhou M. Meigs syndrome with pleural effusion as initial manifestation: A case report. World J Clin Cases. 2021;9(21):5972-9.
11. Schmitt R, Weichert W, Schneider W. Pseudo-pseudo-Meigs' syndrome. Lancet. 2005;366:1672.
12. Tjalma WA. Tjalma's syndrome. Lancet. 2006;367:567-8.
13. Ural UM, Kilic¸ A, Gu¨ngo¨r T. Tjalma's or pseudo-pseudo-Meigs' syndrome: a case report. Clin Exp Dermatol. 2008;33:363-4.
14. Smith LH. Early clinical detection of ovarian cancer: a review of the evidence. Expert Rev Anticancer Therapy. 2006;6(7):1045-52.
15. Adad SJ, Laterza VL, Dos Santos CDT. Cellular Fibroma of the Ovary with Multiloculated Macroscopic Characteristics: Case Report. Case Rep Med. 2012;2012:5.
16. Hall H, Weirich E, Gruda A, Haddad C, Malviya V. Revisiting Meigs Syndrome: A Typical Case Report. Gynecol Obstet Case Rep. 2021;7(8):146.
17. Riker D, Goba D. Ovarian mass, pleural effusion, and ascites: revisiting Meigs syndrome. J Bronchol Interventional Pulmonol. 2013;20:48-51.
18. Nucci R, Oliva E, Goldblum JR. Foundations in Diagnostic Pathology. In: eds. Gynaecologic Pathology. 1st ed. New York: Elsevier Churchill Livingstone. 2009;446-8.
19. Scully RE, Young RH, Clement PB. Tumours of the ovary, maldeveloped gonads, fallopian tube, and broad ligament. In: Rosai J ed. Atlas of Tumour Pathology, 3rd series. Washington, DC: American Registry of Pathology. 1998.
20. Cinel L, Taner D, Nabaei SB, Oguz S, Gökmen O. Ovarian fibrosarcoma with five-year survival: a case report. Eur J Gynaecol Oncol. 2002;23:345-6.
21. Wu H, Xie J, Huang W, Wu J. Mitotically active cellular fibroma of the ovary: a case report and a review of the literature. Eur J Gynaecol Oncol. 2014;35:81-3.
22. Bi R, Zhao Y, Yang WT. Mitotically active cellular fibroma of ovary: a clinicopathologic analysis. Zhonghua Bing Li Xue Za Zhi. 2013;42:660-4.
23. Sivanesaratnam V, Dutta R, Jayalakshmi P. Ovarian fibroma-clinical and histopathological characteristics. Int J Gynaecoln Obstet. 1990;33:243-7.
24. Abad A, Cazorla E, Ruiz F, Aznar I, Asins E, Llixiona J: Meigs' syndrome with elevated CA125: case report and review of the literature. Eur J Obstet Gynecol Reprod Biol. 1999'82:97-9.
25. Abramov Y, Anteby SO, Fasouliotis SJ, Barak V. Markedly elevated levels of vascular endothelial growth factor, fibroblast growth factor, and interleukin 6 in Meigs' syndrome. Am J Obstet Gynecol. 2001;184:354-5.
26. Maskey-Warzechowska KR, Korczynski M, Zielinska-Krawczyk P, Klimiuk M, Chazan J, Light R et al. Pleural Effusion in Meigs' Syndrome-Transudate or Exudate? Systematic Review of the Literature. Medicine. 2015;94(49):e2114.
27. Leung SW, Yuen PM. Ovarian fibroma: a review on the clinical characteristics, diagnostic difficulties, and management options of 23 cases. Gynaecol Obstetr Invest. 2006;62(1):1-6.
28. Chang HC, Bhatt S, Dogra VS. Pearls and Pitfalls in Diagnosis of Ovarian Torsion. Radiographics. 2008;28(5):1355-68.
29. Paladini D, Testa A, Van Holsbeke C, Mancari R, Timmerman D, Valentin L. Imaging in gynaecological disease: Clinical and ultrasound characteristics in fibroma and fibro-thecoma of the

ovary. Ultrasound Obstet Gynecol. 2009;34(5):188-95.
30. Jung SE, Rha SE, Lee JM, Park SY, Oh SN, Cho KS et al. CT and MRI Findings of Sex Cord–Stromal Tumor of the Ovary. AJR Am J Roentgenol. 2005;185(1):207-15.
31. Shingare AB, Meylaerts LJ, Laury AR, Mortele KJ. MRI Features of Ovarian Fibroma and Fibro-thecoma With Histopathologic Correlation. AJR Am J Roentgenol. 2012;198(3):W296-303.
32. Rajabi P, Hani M, Bagheri M, Mirzadeh F. Large ovarian leiomyoma in young woman. Adv Biomed Res. 2014;3:88.
33. Kaku S, Takeshima N, Akiyama F, Furuta R, Hirai Y, Takizawa K. A unique fibrous tumour of the ovary: fibrosarcoma or mitotically active cellular fibroma? Anticancer Res. 2007;27:4365-9.
34. Yıldırım N, Saatli B, Akalın F, Ulukus C, Obuz F, Saygılı U. Mitotically active cellular fibroma of the ovary: A case report J Turk Soc Obstet Gynecol. 2015;1:53-5.
35. Najmi Z, Mehdizadehkashi A, Kadivar M, Tamannaie Z, Chaichian S. Laparoscopic Approach to a Large Ovarian Fibroma: A Case Report. J Reprod Infertil. 2014;15(1):57-60.
36. Târcoveanu E, Dimofte G, Niculescu D, Vasilescu A, Moldovanu R, Ferariu D, Marcovici I. Ovarian Fibroma in the Era of Laparoscopic Surgery: a General Surgeon's Experience. Actachirbelg. 2007;107(6):664-9.
37. Boujoual M. Large twisted ovarian fibroma in menopausal women: a case report. Pan Afr Med J. 2015;20:322.

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