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Opinion

Meigs' syndrome: how we mistook the diagnosis in a tertiary oncology centre in India; an opinion piece on the surgeon's responsibility in minimizing the stress of the cancer patient and family

M. D. Ray*, J. R. Jeena Josephin, Shivani Kapila, Premanand N.

Department of Surgical Oncology, Dr BRA IRCH, AIIMS, New Delhi, India

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***Correspondence:**
Dr. M. D. Ray,
E-mail: drmajormdrayaiimsdelhi@aiims.edu

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ABSTRACT

Typical Meigs syndrome may mislead the surgeon as advanced ovarian cancer. CECT can identify the huge ovarian mass, ascites and pleural effusion. Mostly Surgeon's mind focuses towards the pleural effusion with elevated CA 125 as stage IVa ovarian cancer because of the rarity of this syndrome. But it is Surgeon's responsibility to diagnosis of Meigs syndrome beforehand to alleviate the pain of the patient and her family. Meigs' syndrome typically presents with the triad of an ovarian mass, ascites and pleural effusion. The latter two resolved after removal of the mass. As this syndrome is a rare entity, this presentation of an ovarian mass may mislead the surgeon, biasing his or her mind towards advanced ovarian malignancy. Along with these if the CA-125 is also raised, the patient is usually labelled as stage IVa ovarian cancer, causing immense distress to the patient and family. This makes it even more imperative that the surgeon diagnoses this syndrome beforehand to avoid the pain of the patient and family.

Keywords: Meigs syndrome, Ovarian cancer, Diagnostic dilemma, Surgeons responsibility

INTRODUCTION

Typical Meigs syndrome is a rare presentation of an ovarian mass. Only 2-5% of surgically removed ovarian tumours are fibromas, out of which less than 1% percent as typical Meigs' syndrome.[1]In our vast experience of 600 ovarian cancer cases in surgical oncology department, we encountered typical Meigs syndrome for the first time.

Meigs in 1945 described four criteria for this syndrome. These are (i) The tumour is a benign fibroma or fibroma like tumour ovary (Thecoma, granulosa cell tumour). (ii) ascites, (iii) pleural effusion-unilateral or bilateral and (iv) Removal of tumour resolves all.[2]

Other benign tumours of ovary like teratoma, mucinous cystadenoma, Leiomyoma, Krukenberg tumour ovary with ascites and hydrothorax (pleural effusion) are termed as "pseudo-Meigs syndrome".

The 10-15% of women with ovarian fibroma have ascites. The 1% have hydrothorax. About 70% pleural effusions are right sided, 15% left sided and 15% bilateral.[3,4] Incidence increases with the age, starting from the third decade.

Meigs' syndrome is a deceptive entity. Our patient, a 40 years old lady having two small children, presented to us with lower abdominal discomfort, irregular menstrual cycles and a large abdomino-pelvic mass. Her performance status was good (ECOG1). Abdominal examination revealed a 12×10 cm firm abdominopelvic mass reaching above the umbilicus superiorly.

We evaluated her further with imaging and lab investigations. Her CT and MRI abdomen and pelvis revealed a 15×13 cm solid-cystic mass with enhancing septae in umbilical region in right paramedian location. Left ovary was not separately seen. Right ovary was normal and her uterus had an intramural fibroid. She also

had moderate ascites and right sided pleural effusion. Her CA-125 was 895 U/mL. Pleural fluid was aspirated twice. Both the times cytology was negative for malignant cells.

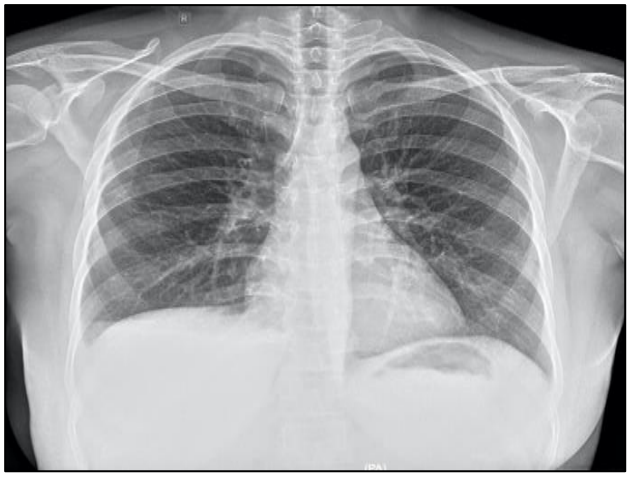

Figure 1: Chest x-ray showing right pleural effusion.

We made a provisional diagnosis of ca ovary, FIGO stage IVa based on clinical and radiological findings. We started her prehabilitation three to four weeks prior as we planned her for primary cytoreductive surgery (CRS). The patient was so depressed that she started visiting us frequently with folded hands requesting for early surgery. We could understand the agony of the patient and her family members including two children but we were following our protocol for primary CRS as we strongly suspected carcinoma ovary considering the large complex adnexal mass with ascites and effusion and raised CA-125. The agony of the immediate family members and her internal struggles are usually unknown to the whole treating team!

On laparotomy we found an 12×12 cm left ovarian mass with hard solid component and minimal cystic component, twisted on its axis. 300 ml of ascitic fluid was present. There were no peritoneal or omental deposits and no enlarged lymph nodes found.

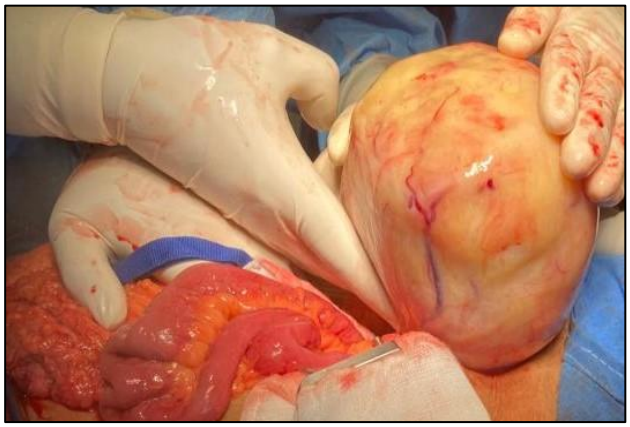

Figure 2: Picture of the edematous large globular mass in left ovary.

On the operation table, we came to the conclusion that it was a typical case of Meigs syndrome, considering her ovarian mass and her clean peritoneum. We just removed the adnexal mass, keeping the age of the patient in mind. We sent the mass for frozen section and it showed fibroma ovary. Then only we were able to make out that it was a case of typical Meigs' syndrome, not ca ovary, though CA 125 was high.

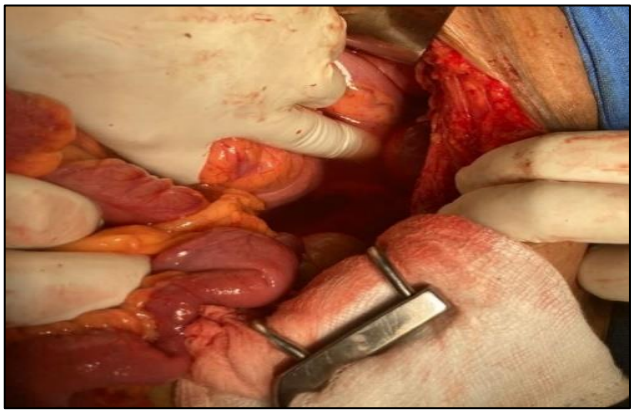

Figure 3: Picture of ascites.

When we disclosed to the family members that it was not a cancer ovary rather benign fibroma, they literally burst into tears. Lead surgeon Dr. Ray says," being a 'so called' experienced surgeon I felt guilty, not diagnosing the syndrome before placing her on the operation table. The agony I caused to the patient for more than a month is not acceptable to me. We can conceal our professional fault from other people but it is difficult to convince own self"

Though the patient and her family were extremely happy with the good news and satisfied recovery, still we like to share our experience so that other oncosurgeons may not face the same scenario.

Her post-operative period was uneventful and she was discharged on 3rd post-operative day.

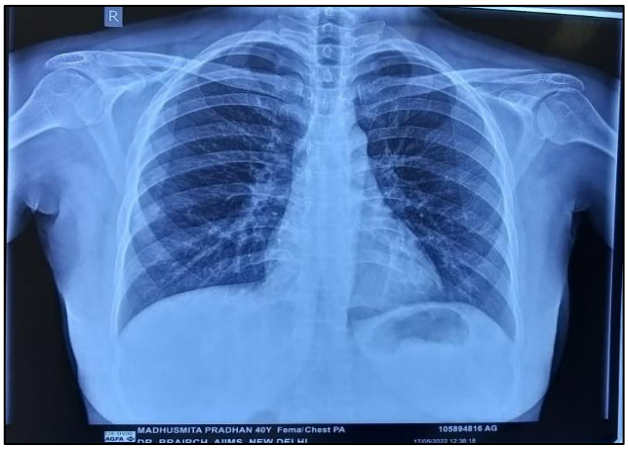

Figure 4: Chest X-ray shows resolution of effusion on post-operative day 10.

On post operative day 10 chest X-ray shows no pleural fluid.

And on 14 days USG abdomen shows no ascites.

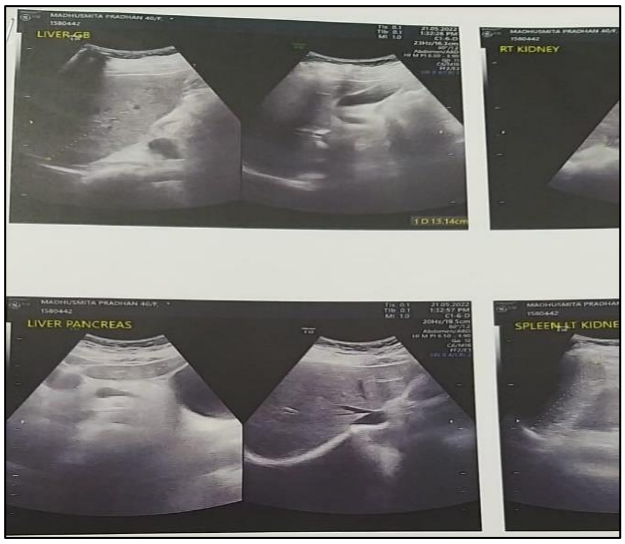

Figure 5: Ultrasound shows resolution of ascites on postoperative day-14.

Histopathology showed left ovarian mass comprising of a well encapsulated, well circumscribed globular mass measuring 13×12×9 cm. capsule appears congested. Cut surface shows a grey-white to yellowish tumour measuring 12.8×11.8×0.8 cm. necrosis and hemorrhage are not seen. It lies 0.1 cm from the capsule. Normal ovarian parenchyma is not identified. Sections examined, showed a tumour comprising of spindle cells with mild nuclear atypia. There is increase in cellularity. Mitosis of 3-4/10 hpf is noted. In addition, some of the portion shows thecomatous areas. Overall features are those of fibro thecoma with foci of cellular fibroma, left ovarian mass.

Overall features including fibro thecoma, ascites and pleural effusion confirmed the diagnosis of Meigs syndrome.

DISCUSSION

The combination of ovarian mass, ascites with pleural effusion along with raised CA125 in a middle aged lady raise the suspicion as ovarian cancer usually.

The painful thing is that when patient and close family members come to know about the advanced ovarian cancer and its prognosis, it becomes a nightmare for them. Meigs' syndrome is at least a better diagnosis than an ovarian carcinoma from the patient's perspective. It is true that because of rarity of this syndrome, surgeon's mind usually does not think of this syndrome rather prepares for the worst case of ovarian cancer.

But from critical point of view, expert mind could diagnosis the case beforehand by USG itself as it shows well demarcated adnexal mass without increased vascularity.

MRI/CECT can find out edema, degeneration, hemorrhage may diagnose this syndrome without features of metastasis.

Anyway, we missed the diagnosis, despite of typical features. Thereby we could not counsel the patient in better way.

Every moment of the lady, who was having two small kids, was so distressed along with whole family could have been relieved on exact diagnosis beforehand. It is important to diagnose this benign condition which we were focusing only as advanced ovarian cancer.

The possible hypothesis of the manifestations of Meigs syndrome are-(i) Diffusion of interstitial fluid in peritoneal space, subsequently into the pleural cavity through the lymphatic channels.[4,5] (ii) Imbalance between blood supply and its lymphatic and venous drainage to a large tumour may responsible for stromal edema and transudation.[6] (iii) Fluid accumulation may be due to VEGF induced raise capillary permeability and (iv) Raising of CA125 may be secondary to mesothelial expression of CA125.[7]

CONCLUSION

Meigs syndrome is a rare clinical entity without any doubt. Early diagnosis may alleviate the depression level of the patient and her whole family members. It is the surgeon's responsibility to diagnose the syndrome beforehand which we missed out in this case despite of large experience in tackling ovarian tumours. We cannot deny our responsibility to diagnose the right disease in right way and right time.

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