

MEIG'S SYNDROME CAUSED BY A GRANULOSA CELL TUMOUR

(Report of a Case)

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The following case is reported because of the relative rarity of the condition.

Meig's syndrome is a relatively uncommon condition which mimics the terminal phase of a metastasizing malignant tumour, far advanced cirrhosis or cardiac failure, but curable by surgical treatment. The syndrome is a triad of an ovarian tumour, ascites and hydrothorax. The true syndrome is confined to fibromas, thecomas, granulosa cell tumours and Brenner tumours.

The first report of such a case was made by Cullingworth of England in 1879 in an autopsy protocol. Since that time there have been several reports in the literature, by Lawson Tait, Spencer Wells, Gaillard Thomas and others. Three cases of fibroma of the ovary with ascites and hydrothorax were first discussed in "Tumours of the female pelvic organs" in 1934 by J. V. Meigs. In 1937, Rhoads and Terrell reported a case from the University of Pennsylvania Hospital in the *Journal of the American Medical Association*, and the eponym

"Meig's Syndrome" was first used. Many more case reports have appeared in the medical literature, so that by 1954, J. V. Meigs was able to collect 84 cases, that were considered to be definitely representative of the syndrome, though many other cases may have remained unreported.

Case Report

On June 9, 1962, a female patient (P.K.T.) aged 40 years was admitted to the gynaecological wards of the hospital with the presenting symptoms of distension of abdomen and breathlessness. She looked extremely ill and found it difficult even to walk. Marked dyspnoea and a huge swelling of the abdomen were at once obvious. She had noticed the distension of abdomen for the past two years and suffered from breathlessness on accustomed exertion for the last 6 months. The latter symptom had become distressing during the previous month. She gave no history of any other illness. Her menstrual cycles were normal till one year ago, since when she gave history of amenorrhoea. She had had 7 full-term normal deliveries, last delivery being 10 years ago.

On examination, there was pallor of the skin and conjunctivae, but no cyanosis. The veins of the neck were engorged. The temperature was 98°F and pulse rate 100. Blood pressure was 130/80. There was no pitting oedema over the ankles. The apex beat was in the 5th space and well inside the midclavicular line. The heart sounds

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were normal. The trachea was deviated to the right.

Examination of the respiratory system revealed a massive left-sided pleural effusion, extending to the 2nd. intercostal space. Movements of the left side of the chest were markedly impaired and percussion revealed a dull note. The vocal resonance was markedly diminished, so also was the air entry.

The abdomen was markedly distended, with prominent veins over it. A marked degree of ascites was present. It was difficult to feel the liver and spleen. A lump with ill-defined margins and reaching just above the level of the umbilicus was felt in the lower abdomen. The lump seemed to be arising from the pelvis. On bimanual examination, it was not possible to feel the uterus. All the fornices were bulging but no well-defined lump could be made out.

The presence of ascites, together with pleural effusion and a lump in the lower abdomen at once suggested a possible diagnosis of Meig's Syndrome.

On June 10, investigations showed: Hb — 7.2 gms.% W.B.C. — 7200/cmm, E.S.R. — 25 mm. at the end of one hour (Wintrobe). Urine contained no albumen or sugar. A chest radiograph, on June 10, showed a massive pleural effusion on the left side, with mediastinal shift to the right. A plain X-ray of the abdomen showed a soft tissue shadow in the right half.

On June 11th., the peritoneal cavity was tapped. About 2 pints of blood-stained fluid were removed. An examination of the fluid showed: Proteins — 4.4 gms%, cobweb absent, coagulum present, R.B.C.s ++. A cytological examination of the fluid failed to show any malignant cells.

On June 14th., the patient became very breathless, hence a paracentesis of the left pleural cavity was done. One pint of blood-stained fluid was removed. An examination of this fluid showed essentially similar constituents as the ascitic fluid.

On June 15th., a peritoneal tapping was repeated and 2 pints of fluid removed to lessen the distension.

At operation on June 18th., about 4 pints of blood-stained fluid was removed from the peritoneal cavity. Laparotomy revealed a large, irregular ovarian tumour arising

from the right ovary filling the pelvis and reaching above the umbilicus. The tumour showed adhesions to the parietal peritoneum (possible secondaries in view of the subsequent pathological report). The peritoneum showed secondary implants. The other ovary was normal. A total hysterectomy with bilateral salpingo-oophorectomy was performed. At operation, the liver, gall-bladder and stomach were felt to be normal.

Gross appearance:—The ovarian tumour measured 19 x 16 x 8 cm. It was globular, the surface being irregular at some places and smooth at others.

There were fibrous fasciculae in one area. There was destruction of the surface in another area which measured 6 x 4 x 2 cms. The tumour was soft in consistency. The cut surface showed a soft yellowish substance with an area of chocolate colour (10 x 6 cms). The tumour showed numerous blood-vessels.

The other ovary was small and appeared to be normal.

Histo-pathological report:— On microscopic examination, the tumour turned out to be a granulosa cell tumour. At the periphery there was a fibrous capsule, infiltrated by tumour cells, which in some areas, had reached up to the outermost part of the capsule. The tumour was highly cellular. The cells were round or polyhedral or even plump and elongated and spindle-shaped. In some areas, the cytoplasm was scanty. The cells showed hyperchromatic nuclei. Mitotic figures were seen in small numbers. The cells were arranged in a variable number, either in sheaths or groups, the general pattern being diffuse or parenchymatous. At many places, the cells were arranged in clusters or rosettes—Call Exner bodies could be seen in some areas, resembling primordial follicles. At the periphery of such collections were seen some elongated spindle shaped cells. They showed plenty of cytoplasm which was vacuolated (Theca Cells). Their cytoplasm contained fat, as shown by Sudan III stain. Some vessels were well formed, while some were ill-formed. These vessels did not show tumour cells in their lumen. The stromal fibrous tissue was hyalinized at some places.

The endometrium showed a proliferative phase—the myometrium being normal.

Post-operatively, the patient was in profound shock for 2 days. However, with blood transfusions and other appropriate therapy, she rallied round and the rest of the post-operative period was uneventful.

On June 25th., a fluorescent examination of the chest showed that the pleural effusion had almost subsided, except for a little fluid at the base.

On July 3rd, repeat fluorescent examination showed a thickened pleura at the left base but no fluid.

In view of the pathological report of the removed tumour, it was decided to give the patient a course of post-operative deep X-rays. 30 sittings were given in all.

AP—Lower Abdomen—200 B x 10 Sit-tings.

Posterior Side-Right Oblique—200 B x 10 Sittings.

Posterior Side-Left Oblique—200 B x 10 Sittings.

On July 19, the patient was discharged from hospital in a good condition.

She was seen again on August 20. A chest screening still showed a thickened pleura at the left base. In other respects, the examination revealed nothing abnormal. The patient has been advised to return for regular check up to detect any evidence of secondaries as early as possible.

Discussion

Although fibromas of the ovary are not nearly so common as cysts and cancers, they are much more frequently responsible for Meig's Syndrome. The interesting feature of the case under review is that the causative tumour was a granulosa cell tumour. Despite the histopathological report, no malignant cells could be detected in either the ascitic or the pleural fluid, and both the fluids disappeared rapidly after the removal of the tumour. In the present case, the fluid was blood-stained. Meigs, however, thinks that the presence of blood-stained fluid is not necessarily

an evidence for the malignant nature of the tumour.

It has been felt by many that the fluid is always in the right chest. Though, in the majority of instances, this is true, in our case it was on the left side. Meigs gave the following figures: Right side, 62%; Left side 11%; both sides 24%, and unknown 3%.

Granulosa cell tumour belongs to the functioning group of ovarian tumours. This patient gave history of amenorrhoea of one year's duration. This long period of amenorrhoea without any episodes of bleeding is difficult to explain. A histopathological study of the endometrium revealed a proliferative phase without any evidence of cystic hyperplasia.

Two factors of interest emerge from the present case, firstly the importance of a gynaecological examination in every case of pleural effusion, especially in women where the causative factor may be in doubt (of course, in the present case, the lump was palpable per abdomen), and secondly the fluid may disappear completely after the removal of the tumour, even though the latter be a malignant one.

Various theories have been put forward to explain the existence of fluid in the peritoneal and pleural cavities in cases of Meig's Syndrome. Also, the reason why the chest and abdominal fluids disappear suddenly after the tumour is removed, is unknown.

Fluid in the abdomen has been explained by:

(1). Low serum proteins. This

has not been borne out by laboratory data.

(2). Selye's alarm reaction. His explanation is that repeated trauma to the peritoneum first provokes a resistance of the tissues, but after some time the resistance disappears. There then appears a histamine toxicosis or anaphylactic shock with occurrence of peritoneal and pleural exudates.

(3). Obstruction of the azygos veins.

(4). Inflammation in the pelvis. In most of the cases, there is no evidence of inflammation.

(5). Twisting of the pedicle of the tumour with consequent exudation.

There has been difficulty, too, in explaining the fluid in the chest. It is conceivable that fluid may pour into the chest through the pleuro-peritoneal canal. Studies of Cowan, Cron, Burgess and Karioris with radioactive gold showed that some form of rapid trans-diaphragmatic lymphatic transport appears to be the most feasible explanation for the presence of hydrothorax.

Summary

A case of Meig's Syndrome caused by a granulosa cell tumour has been described. Removal of the tumour by operation resulted in complete disappearance of the peritoneal and pleural fluids. It is difficult to say whether the cure obtained in the present case will be a lasting one, as granulosa cell tumours are known to recur after surprisingly long periods.

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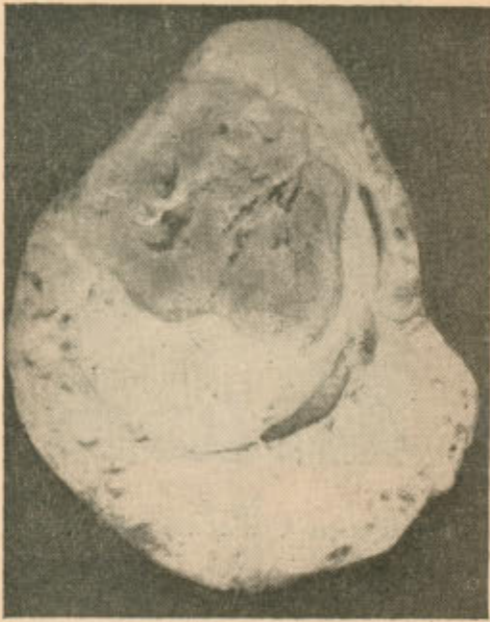


Fig. 1.
Cut surface showing a haemorrhagic area.



Fig. 2.
An oblique view of the tumour.

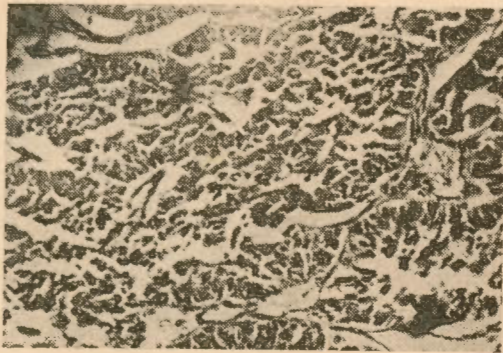


Fig. 3.
Low power view showing the granulosa cells
arranged in clusters.

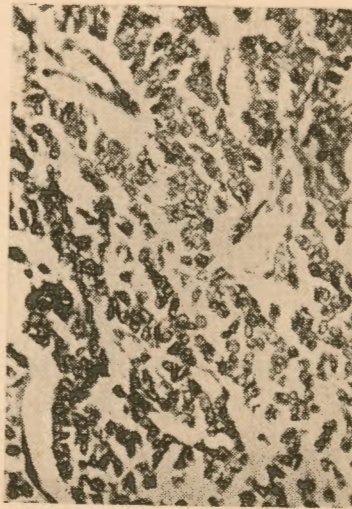


Fig. 4.
High power view of the field above.



Fig. 5.
Low power view of another portion of the
tumour showing a different pattern.



Fig. 6.
High power view of the field above.