Rare ovarian tumours

Ovacome is a national charity providing advice and support to women with ovarian cancer. We give information about symptoms, diagnosis, treatment, research and screening. Ovacome also runs a telephone support line and works to raise awareness and give a voice to all those affected by ovarian cancer.

This fact sheet describes rarer kinds of ovarian cancer and their treatments.

There is a glossary with this fact sheet, to explain words you may not be familiar with.

Normal ovaries are made up of:

- epithelial cells, covering the surface
- germ cells from which ova (eggs) develop
- sex-cord stromal cells which provide supportive tissues and are responsible for producing hormones.

A range of tumours which behave differently and have different results can develop from each of these tissues.

About 90 per cent of malignant (cancerous) ovarian tumours develop from the epithelial cells that cover the surface of the ovaries. This is the common type of ovarian cancer affecting about 6000 women each year in the UK. Its behaviour is well understood, and there are reliable guidelines about treatment and results.

Each year hundreds of women develop one of the rarer ovarian cancers which can be divided into:

- germ-cell tumours
- sex-cord stromal tumours
- other rare tumours.

**Basic principles**

The best care and result for a woman with a rarer tumour depends on accurate staging of the cancer. This
means seeing how advanced the cancer is and if it has spread. You can get more information in our Fact sheet 3 The stages of ovarian cancer.

If you have a rare ovarian tumour, there may be some uncertainty about the best treatment for you. This is because research into different treatments cannot be carried out on the small numbers of women affected.

You and your multidisciplinary team (MDT) of specialist surgeons, oncologists and pathologists should make decisions about your treatment. Sometimes the MDT asks for opinions from other expert pathologists. Often, carefully using tried and tested treatment is the best option.

**Germ-cell tumours**

These tumours begin in the reproductive cells or egg cells of the ovary. They tend to appear in teenage girls or young women. The diagnosis is usually made after you develop symptoms similar to all types of ovarian tumour, such as pain in your pelvis and abdomen, a swollen abdomen, difficulty eating and feeling full quickly.

First investigations include a physical examination and a CT scan. If you are a young woman with a suspected ovarian tumour, you should have an extra test to measure the hormones βHCG and AFP in your blood. If you have a high level of one or both of these in your blood, this is strong evidence that you have a germ-cell tumour.

This will help your surgeon choose the most appropriate operation. As most women with an ovarian germ-cell tumour are cured, the surgeon will carefully consider what to do during the operation, so you may still be able to have children in the future. After surgery the pathologist can identify the type of germ-cell tumour. These include:

- dysgerminoma
- endodermal sinus tumour
- embryonal carcinoma
- choriocarcinoma
- teratoma.

Correct treatment depends on the type and stage of the disease, and whether any tumour is left after surgery.

Because these tumours are relatively rare, you should be referred to a specialist as soon as possible after diagnosis. Sometimes, chemotherapy is the preferred option rather than immediate surgery. A special combination of chemotherapy drugs called BEP is commonly used for germ cell tumours. (BEP refers to the constituent drugs: bleomycin, etoposide, and platinum). This regimen is usually highly effective, but requires careful supervision. Once your treatment is complete, regular follow-up will be organised.

**Sex-cord stromal tumours**

The supporting tissues and cells that produce hormones in the ovary can produce many different tumours. The usual types are:

- granulosa cell tumour
- sertoli-leydig cell tumour
Granulosa cell tumours
These are the most common kind of sex-cord stromal tumours and tend to appear in middle-aged women. They sometimes produce the hormone oestrogen, which can result in symptoms such as those in early puberty or menstrual disorders.

First you will have an operation to confirm the stage of the disease and to remove as much tumour as possible, if it has spread beyond the ovary to the lining of the abdomen, the peritoneum.

Most patients have a tumour just in the ovary (stage1). You will not need any more treatment after surgery and the outlook in most cases is very good. However, in rare cases, granulosa cell tumours come back very late (up to 30 years) after apparently successful surgery. When granulosa cell tumours come back, you may need more surgery to control the disease for longer.

If the disease is more advanced (stages 2, 3 or 4), you may need more treatment after surgery, usually involving drugs. This also applies if your granulosa cell tumour comes back after surgery, and another operation is not likely to help. The aim is to try and control the disease. Because granulosa cell tumours can be cancer that grows slowly, it is sometimes appropriate to keep an eye on the tumour by having repeated scans.

This means you can wait to have treatment until the condition is causing you problems.

Before you start chemotherapy treatment, you may want to have a trial of hormonal therapy with medroxyprogesterone acetate (provera). Some granulosa cell tumours will shrink for a while if you take this medicine.

If you have an advanced granulosa cell tumour and need chemotherapy to control it, there is a range of options available. Aggressive combination chemotherapy can sometimes help, but you are at risk of significant side effects.

Another approach is to use the same type of chemotherapy as is used for common epithelial ovarian cancer which is Carboplatin. There is some evidence that using Paclitaxel (Taxol) and Carboplatin may achieve a better result than Carboplatin on its own.

If you have a granulosa cell tumour, you and your oncologist should decide what treatment you are going to have. You should think about the benefits and the possible side effects. If you have symptoms from specific tumours, radiotherapy can be helpful.

Sertoli-leydig cell tumours
These tumours can produce male hormones, and may be detected if you develop symptoms such as a lot of body and facial hair, or baldness. Most sertoli-leydig cell tumours are not malignant (cancerous) and can be cured by surgery.
Other rare tumours
There are some uncommon but important ovarian tumours that do not fit into the categories above.
- Small-cell carcinoma of the ovary
- Brenner tumour
- Krukenberg tumour

Small-cell carcinoma of the ovary
Small-cell carcinoma of the ovary tends to appear in young women, and behaves like an aggressive form of the more common epithelial ovarian cancer.

If the disease is stage 1 (just in the ovary) some patients will be cured with surgery. But there is a significant risk that the cancer will come back, even at this early stage. It is not clear whether giving chemotherapy immediately after surgery reduces this risk.

If you have small cell ovarian cancer which is advanced, or which comes back, the benefits of chemotherapy are limited. However you can have times where you do not have cancer (remission).

Brenner tumours
Brenner tumours of the ovary are often discovered by chance. They probably arise from the surface of the ovary. Most are not cancerous, so no other treatment is needed except surgery to remove them.

Krukenberg tumours
Krukenberg tumours are not ovarian cancers. They are formed when seedlings of cancer from another organ (usually the stomach, but sometimes other parts of the gut) grow on the ovary.

These tumours can be confusing if under the microscope they look similar to common epithelial ovarian cancer. However, special tests (immunostains) can find where the primary cancer is and improve the diagnosis.

After surgery to remove the affected ovary, women with Krukenberg tumours should receive treatment specifically for the area of the body the cancer first affected.

You can get more information in Fact sheet 10 Borderline ovarian cancer and Fact sheet 11 Clear cell carcinoma of the ovary.

If you would like more information on the sources and references for this fact sheet, please call us on 0800 008 7054. If you would like to discuss anything about ovarian cancer, phone our support line on Freephone 0800 008 7054 Monday to Friday 10am to 5pm.
You can also visit our website at www.ovacome.org.uk.

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Glossary

Benign  Not cancer - used to refer to tumours which grow slowly in one place and which, once removed by surgery, tend not to come back.

βHCG and AFP  These are hormones. The levels of these hormones can be measured with a blood test.

CT scan  A CT (computerised tomography) scan uses x-rays to produce images of the body.

Malignant  Malignant tumours are able to spread to and destroy surrounding tissues and other organs in the body.

Pathologist  A specialist who examines parts of the body affected by disease.

Radiotherapy  Treating cancer using radiation.

Staging  The process for deciding how far the cancer has spread, by using internationally recognised and agreed standards. This can be done using x-rays, scans, blood tests or surgery.