PRIMARY BONE CANCER
OSTEOSARCOMA

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Osteosarcoma is the most common form of primary bone cancer affecting children and young adults. The most effective way to treat osteosarcoma is by combining the use of surgery and chemotherapy.

**WHAT IS IT?**

‘Osteo’ comes from an ancient Greek word for bone and ‘sarcoma’ is the name given to cancers that start in connective or supporting tissues - such as the bone, fat, cartilage, blood vessels or muscle.

Osteosarcoma forms when a bone cell becomes abnormal and grows out of control to form a tumour. The cells in the tumour act like immature bone and try to create new bone as they grow and divide. If bone cells can be seen in a tumour sample when it is assessed under the microscope, it helps the doctor to confirm a diagnosis of osteosarcoma.

The three areas most commonly affected by osteosarcoma are the lower **thigh bone** (known as the femur), the upper **shin bone** (known as the tibia) and the upper **arm bone** (known as the humerus). In rarer cases, osteosarcoma can also be found in the jaw, the spine or the pelvis.

Very rarely, osteosarcoma can start in more than one bone at the same time. This is called **multi-focal osteosarcoma**.

**WHO DOES IT AFFECT?**

Osteosarcoma can affect all ages but is the most common form of primary bone cancer in children and young adults, aged **10 to 24** years of age. There is also a smaller group of people **over the age of 55** who develop osteosarcoma. This cancer is known to affect males slightly more than females.

In the UK alone, around 160 people are diagnosed with osteosarcoma each year.
WHAT ARE THE SYMPTOMS?

The symptoms of osteosarcoma are general and can be similar to sports injuries, growing pains, or common conditions such as tendonitis or arthritis.

- **INTERMITTENT BONE PAIN**
  - which can be constant or come and go, and may be worse at night

- **A LUMP OR SWELLING**

- **PROBLEMS WITH MOBILITY**
  - such as stiff joints or reduced movement

- **A BONE FRACTURE**

- **BRUISING EASILY**

- **TIREDNESS, FEVER OR WEIGHT LOSS MAY ALSO BE EXPERIENCED**

These symptoms may vary in each patient and can present alone or in combination with one another.
By looking closely under a microscope at what kind of cells are making up the tumour, doctors can class each patient’s tumour as one of five main subtypes of osteosarcoma. These are:

• **OSTEOBLASTIC OSTEOSARCOMA**

• **CHONDROBLASTIC OSTEOSARCOMA**

• **FIBROBLASTIC OSTEOSARCOMA**

• **TELANGIECTATIC OSTEOSARCOMA**

• **SMALL CELL OSTEOSARCOMA**

These five subtypes account for around 90% of all osteosarcomas and are all treated in the same way - using a combination of surgery and chemotherapy.

The remaining 10% of osteosarcomas are less aggressive forms, known as ‘parosteal’ or ‘periosteal’ osteosarcomas, which may require only surgery to treat these tumours.
We still don’t understand the cause of osteosarcoma. However, in around 10-15% of cases, there are possible causes and factors that have been identified which may increase the individual’s risk of developing osteosarcoma.

These possible causes and risk factors include:

- **UNDERGOING PREVIOUS RADIOTHERAPY TREATMENT**

- **INHERITING A DAMAGED GENE FROM YOUR PARENTS**

- **HAVING UNDERLYING BONE ABNORMALITIES** – such as Paget’s disease.

- **HAVING A RARE, INHERITED, CANCER KNOWN AS RETINOBLASTOMA** – this childhood eye cancer develops when a baby inherits a damaged copy of a gene called ‘Rb’, which usually prevents cells from dividing too quickly. Around 12% of retinoblastoma patients may develop an osteosarcoma.

- **HAVING AN INHERITED CONDITION KNOWN AS LI-FRAUMENI SYNDROME** this syndrome is caused by inheriting a damaged gene known as p53, which is one of the most important cancer-preventing genes. Inheriting a damaged p53 gene can increase an individual’s risk of developing various cancers - including osteosarcoma.

A risk factor for older patients (over the age of 55 years) is **Paget’s disease of the bone** - a disorder that results in the bones becoming misshapen and painful. Around 2% of patients with Paget’s disease of bone will develop osteosarcoma.

Other rarer conditions that create an increased risk of developing osteosarcoma are **Rothmund-Thomson Syndrome** and **Werner’s Syndrome**.
The first step in diagnosing any primary bone cancer is a trip to the doctor, where a clinical examination and an X-ray will be carried out. There is no clear sign that doctors can easily look for to make a diagnosis of osteosarcoma. A CT scan and MRI scan provide crucial information on the exact location of the tumour, the stage of the tumour and the presence of an osteosarcoma spreading elsewhere in the body.

Taking a biopsy of the bone is very useful when diagnosing osteosarcoma. This specialist procedure takes a small sample of the tumour so it can be examined under a microscope.

Results from a biopsy can take up to two weeks to analyse but they enable doctors to confirm the presence and specific type of osteosarcoma.

Further tests to confirm a osteosarcoma diagnosis include:

- A CT SCAN
- AN MRI SCAN
- A BIOPSY OF THE BONE
- BLOOD TESTS
When diagnosing osteosarcoma, it is important to distinguish this tumour from other health conditions, or more common occurrences such as ‘growing pains’ or a sports injury.

There may be health conditions that present similarly to osteosarcoma - in terms of symptoms and signs - but it is important the correct diagnosis is made to ensure the treatment provided is suitable. Distinguishing a disease from a similarly presenting condition or disease is known as ‘differential diagnosis’.

AN ALTERNATIVE DIAGNOSIS?

If the diagnostic tests show that the patient does not have osteosarcoma, there are a number of other conditions that may be presenting, such as:

- **OSGOOD SCHLATTERS DISEASE** - which is found in physically active adolescent boys and girls, and is caused by stress on the tendons that connect to the knee-cap. This causes pain in the leg bones, similar to osteosarcoma.
If the presence of osteosarcoma is confirmed the patient will be referred to the nearest Bone Cancer Centre where the specialist medical team will design the best possible treatment plan for the individual patient.

The survival rates for osteosarcoma have improved dramatically as treatments have been standardised throughout much of the world. To treat osteosarcoma, chemotherapy and surgery are used alongside one another.

Surgery

The aim of surgery is to remove the primary tumour to prevent its growth and spread to other areas of the body. The surgical removal of an osteosarcoma requires ‘wide-surgical margins’; this means some healthy tissue is removed alongside the tumour to ensure all tumour cells are removed and there is a lower risk of the tumour returning at a later date.

The surgical procedure carried out to treat an osteosarcoma is known as ‘limb-salvaging surgery’, which aims to remove the tumour while preserving as much of the normal function and cosmetic appearance of the limb as possible.

Common limb-salvaging surgical procedures performed are:

- **RESECTION**: the affected area of bone is removed

- **AN AUTOGRAFT/AUTOLOGOUS GRAFT**: the affected area of bone is removed and reconstructed using the patients’ own tissue from another area of their body.

- **AN ALLOGRAFT**: donated tissue is used to reconstruct the affected area of the bone once the tumour has been removed

- **A METALLIC REPLACEMENT**: once the tumour is removed the area of damaged bone is replaced with a metal implant known as a prosthesis. This procedure usually requires rehabilitation therapy after surgery

- **IRRADIATION/REIMPLANTATION**: damaged bone is removed and treated with radiation, destroying the cancer cells, before being put back into the body

- **AMPUTATION**: on very rare occasions, the removal of the whole limb is required due to the size or location of the tumour. If possible, a prosthetic limb will be made for the patient
CHEMOTHERAPY

In most cases, chemotherapy is used before surgery to kill cancer cells and shrink the tumour to make surgery easier; this is known as ‘neo-adjuvant chemotherapy’. The number of chemotherapy drugs used can vary from 2 to 4 and these can be given for between 6 and 12 weeks.

Chemotherapy can also be used after surgery to destroy any remaining cancer cells in the location of the tumour or in the rest of the body; this is known as ‘adjuvant chemotherapy’. Most treatment courses last for a further 16 -30 weeks after surgery and tend to combine the use of 3 drugs.

The chemotherapy drugs used to treat osteosarcoma are called:

- METHOTREXATE
- DOXORUBICIN
- CISPLATIN
- IFOSFAMIDE
- ETOPOSIDE

Mifamurtide (also known as Mepact) is also used for the treatment of osteosarcoma in the UK and Ireland. It was licensed following an American clinical trial that proved Mifamurtide to improve the long-term survival aspects for osteosarcoma patients. The Bone Cancer Research Trust was active in supporting the approval of Mepact for patients in the UK, and were consulted during the appraisal process of this drug.

RADIOThERAPY

Not all osteosarcomas are found in the limbs. Tumours in the pelvis, skull, spine and jaw can often be difficult to remove completely by surgery. Therefore, radiotherapy is occasionally used in situations where it is not possible to remove the whole tumour surgically.
FOLLOW-UP CARE

After finishing treatment, many patients will require follow-up care.

Follow-up care at the hospital will allow healthcare professionals to keep an eye on a patient’s general health and ensure the patient hasn’t suffered any ‘late effects’ from their treatment. Late effects of a patient’s treatment include effects on the patient’s kidney function, fertility or risk of developing a secondary cancer.

Follow-up care can continue for months, or even years, and allows patients to discuss any concerns they may have with their doctor. Tests may be carried out during these appointments to ensure the patient is healthy and the cancer is not at risk of returning.

REHABILITATION AND SUPPORT

Following treatment, many patients benefit from further support and rehabilitation services.

Rehabilitation is a form of therapy that enables patients to regain strength, tackle day-to-day activities and return to normal life as quickly as possible following a disease. These services are available both during and after treatment and include:

- **Physiotherapists:** help patients return back to an active lifestyle as quickly as possible to restore strength, movement and function.
- **Occupational therapists:** help patients to complete day-to-day activities in order to regain their independence.
- **Dietician:** offer advice on the most appropriate nutrition for patients during and after their treatment.
- **Prosthetists:** specialists who design and create prostheses following amputations to match as closely as possible to the individual patient’s removed limb.
- **Orthotists:** specialists who provide aids for patients following surgery, such as splints or special footwear.

Patients, or their family and friends, may benefit from discussing any feelings of anxiety or concerns they may have following a cancer diagnosis or treatment. Many services are available for this form of support, such as:

- **Psychological support and services:** psychologists will support patients through any feelings of anxiety or depression to overcome the concerns that often come with a cancer diagnosis.
- **Local support groups:** many support groups are organised and run locally. It is best to ask your clinical nurse specialist for information on these local services.
THE BONE CANCER RESEARCH TRUST IS THE LEADING CHARITY DEDICATED TO FIGHTING PRIMARY BONE CANCER.

OUR MISSION IS TO SAVE LIVES AND IMPROVE OUTCOMES FOR PEOPLE AFFECTED BY PRIMARY BONE CANCER THROUGH RESEARCH, INFORMATION, AWARENESS AND SUPPORT.

WE RECEIVE NO GOVERNMENTAL FUNDING, SO RELY ENTIRELY ON THE SUPPORT OF THE PUBLIC TO CONTINUE OUR LIFE SAVING WORK.

FOR INFORMATION AND SUPPORT CONTACT US:

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