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PRIMARY BONE TUMOUR

AMELOBLASTOMA (A NON-CANCEROUS TUMOUR)

BONE CANCER RESEARCH TRUST
UNTIL THERE'S A CURE

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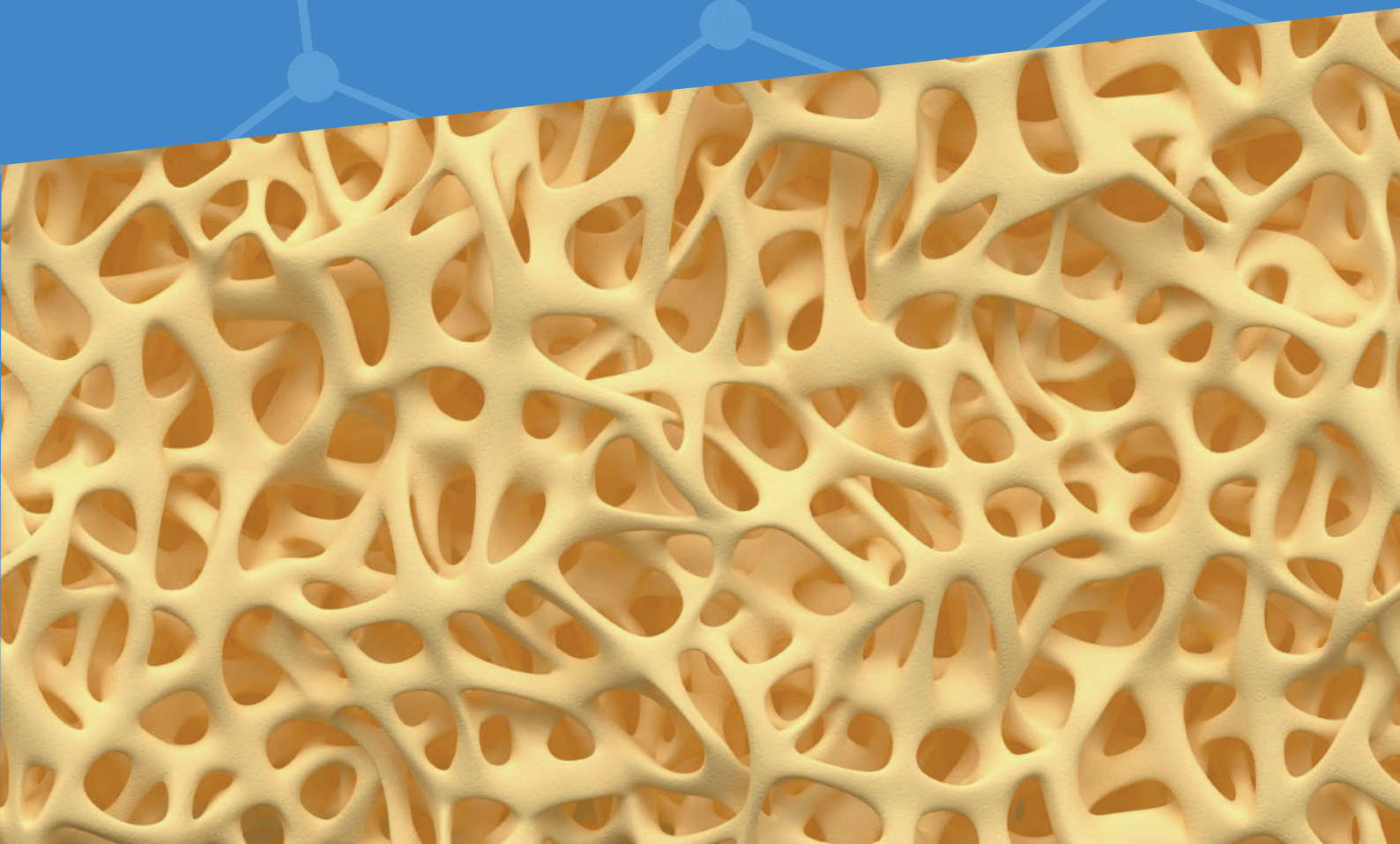

Ameloblastoma is a rare, benign (non-cancerous), tumour arising in and around the jaw bone. Only 1% of all jaw tumours are ameloblastomas. This tumour involves excess tissue growth in and around the jaw, which can appear to be solid or cyst-like.

WHAT IS IT?

Ameloblastoma is a rare, **non-cancerous**, tumour which can occur in the lower or upper jaw bone. It is very common for this tumour to occur around the position of the wisdom tooth.

This tumour most frequently occurs in the **lower jaw bone** (known as the mandible), with approximately 80% of ameloblastomas occurring here. The remaining 20% of cases occur in the upper jaw bone (known as the maxilla).

Although ameloblastomas tend to grow slowly and are non-cancerous, they can occasionally affect tissues surrounding the jaw area - such as the sinuses or the eye-sockets – causing destruction of the bone and some facial distortion.

WHO DOES IT AFFECT?

Ameloblastoma is known to affect males and females in equal proportion and can affect any age group or ethnicity.

Although ameloblastoma can occur at any age, this tumour is most commonly seen in patients around **30 to 40 years of age** and is relatively uncommon in children under the age of 10.

WHAT ARE THE SYMPTOMS?

Some ameloblastoma patients experience no symptoms at all until the tumour is of a larger size. Doctors describe this as being '**asymptomatic**'. Therefore, in many cases, this rare tumour type is discovered during routine dental examinations and X-rays.
The most commonly reported symptoms of ameloblastoma are:

Patients may not experience any of these symptoms, or may only experience a few of the ones listed.

TYPES OF AMELOBLASTOMA

There are a number of different types of ameloblastoma, which vary due to their structural make-up.

It is important to determine the correct form of ameloblastoma that is presenting to ensure the best treatment plan is put into action for the individual patient.

The four main types of ameloblastoma are:

- **MULTI-CYSTIC OR CONVENTIONAL AMELOBLASTOMA**
- **UNI-CYSTIC OR CYSTIC AMELOBLASTOMA**
- **PERIPHERAL OR EXTRAOSSEOUS AMELOBLASTOMA**
- **DESMOPLASTIC AMELOBLASTOMA**

CAUSES AND RISK FACTORS

The cause of ameloblastoma is unknown, and due to the rarity of this tumour it is hard to assess a possible cause from the small number of cases that have been reported.

Recent research suggests that there is an association between abnormalities in certain genes (known as gene mutations) and the development of ameloblastoma. These genes, known as **BRAF** and **SMO**, are involved in controlling cell growth, division and survival.

Mutations in genes BRAF and SMO have been seen in over **80%** of ameloblastoma cases and so further investigation into these genes is currently taking place.

MUTATIONS IN GENES BRAF AND SMO HAVE BEEN SEEN IN OVER 80% OF AMELOBLASTOMA CASES

DIAGNOSING AMELOBLASTOMA

Further tests to confirm an ameloblastoma diagnosis include:

- **A CT SCAN**
- **AN MRI SCAN**
- **A BIOPSY OF THE BONE**

Many ameloblastomas are detected at the dentist and are discovered accidentally during routine dental X-Rays.
A CT scan cannot definitively diagnose an ameloblastoma. However, using a CT scan alongside an MRI scan can provide important information on the exact location of the tumour and the size of the tumour.

Taking a biopsy of the bone is needed to confirm the diagnosis of an ameloblastoma. This specialist procedure takes a small sample of the tumour so it can be examined under a microscope.

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

AN ALTERNATIVE DIAGNOSIS?

When diagnosing an ameloblastoma, it is important to be able to tell the difference between this rare tumour and any other health conditions that may present in a similar manner - in terms of signs, symptoms and diagnostic appearance. Conditions other than ameloblastoma which can be the cause of particular symptoms are known as '**differential diagnoses**'.

Other conditions which can present in the same way as ameloblastoma include:

- **KERATOCYSTIC ODONTOGENIC TUMOUR -** another benign (non-cancerous) tumour of the jaw
- **A HARD ODONTOMA -** a tumour of the dental region
- **AMELOBLASTIC FIBROSARCOMA -** a rare but aggressive tumour of the dental region

TREATING AMELOBLASTOMA

If the presence of ameloblastoma 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.

Due to the rarity of ameloblastoma, and therefore the smaller number of reported cases, it is difficult to work out the most effective and safest treatment for this tumour. Research suggests that chemotherapy is of little-to-no benefit in treating this tumour type and is used purely to relieve patients' symptoms.

SURGERY

The most effective treatment method for ameloblastoma is the surgical removal of the tumour using '**wide surgical margins**'. This means the tumour is removed alongside a small amount of healthy tissue to ensure all tumour cells are removed and to lower the risk of the tumour returning at a later date.

On rare occasions, it may be necessary for surgeons to perform a procedure known as a '**hemimandibulectomy**' - which is the removal of part of the lower jaw.

Once the tumour has been removed, surgical reconstruction will be carried out using either a **metal implant** or an **autograft** – which is a graft that uses bone taken from elsewhere in the patient's body to reconstruct the jaw once the tumour is removed.

RESEARCH INTO AMELOBLASTOMA

Research is being carried out to investigate genes which appear to be commonly mutated in ameloblastomas - such as **BRAF** and **SMO** genes. The mutations observed in these genes appear to be associated with a lack of control over the cells growth and division – which ultimately leads to the development of a tumour.

There are developed and approved drugs to target these mutations, which are used in the treatment of other tumour types. Therefore, it is hoped that in the near future these targeted drugs (such as Vemurafenib which directly targets the BRAF mutation) can be tested for their effectiveness in the treatment of ameloblastoma.

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

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 ran 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:

CALL 0113 258 5934

OR VISIT BCRT.ORG.UK

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