CONDITION

Ganglioneuroblastoma

Ganglioneuroblastoma is a tumour of developing nerve cells that can behave mildly or like neuroblastoma.

WHAT IT IS
Tumour containing immature and maturing nerve cells

BODY SYSTEM
Nervous system

MAIN SYMPTOMS
Lump, abdominal pain, bone pain, weakness

WHAT CAUSES IT
Usually unknown; rarely linked to inherited changes

HOW IT IS DIAGNOSED
Biopsy with tumour pathology; imaging for stage

MAIN TREATMENT
Risk-based surgery, chemotherapy, radiation, and immunotherapy

CAN IT BE CURED?
Sometimes - localized favourable tumours are often curable

IS IT AN EMERGENCY?
Sometimes - breathing trouble, new weakness, or bladder loss

LONG-TERM OUTLOOK
Strongly linked to stage, subtype, age, and tumour genetics

Overview

Ganglioneuroblastoma is a tumour that develops from immature nerve cells. These cells normally form parts of the sympathetic nervous system, which helps control automatic functions such as heart rate and blood pressure.

The tumour most often begins near an adrenal gland above a kidney or alongside the spine in the abdomen, chest, neck, or pelvis. It mainly affects children, although adults can develop it.

Ganglioneuroblastoma belongs to a group called peripheral neuroblastic tumours. The group ranges from ganglioneuroma, which is made mainly of mature cells and is usually non-cancerous, to neuroblastoma, which contains immature cells and can be aggressive.

There are two main forms:

This distinction matters because tumours with the same name may need very different treatment. Doctors consider the pathology subtype, whether the tumour has spread, the person's age, and genetic features within the tumour.

- Intermixed ganglioneuroblastoma contains maturing nerve cells mixed throughout supporting tissue. It often has favourable biological features.
- Nodular ganglioneuroblastoma contains one or more distinct areas resembling neuroblastoma. Its behaviour is strongly influenced by those nodules.

Symptoms

Symptoms depend on the tumour's location, size, and whether it has spread. Some tumours are found unexpectedly during imaging for another reason.

A tumour in the abdomen may cause a firm lump, swelling, abdominal pain, reduced appetite, constipation, or a feeling of fullness. A tumour in the chest can cause cough, chest discomfort, or breathing problems.

A tumour growing near the spine may press on nerves or the spinal cord. This can cause back pain, leg weakness, numbness, difficulty walking, or loss of bladder or bowel control.

If the condition has spread to bones or bone marrow, symptoms may include bone pain, limping, tiredness, pale skin, or easy bruising. Bone marrow is the soft tissue inside bones that makes blood cells.

Some tumours release chemicals called catecholamines. Their breakdown products can often be measured in urine, even when they cause no obvious symptoms. Less commonly, hormone release may contribute to sweating, a fast heartbeat, high blood pressure, or persistent watery diarrhoea.

A neck or upper-chest tumour can sometimes affect nerves supplying the eye. This may cause a drooping eyelid, a smaller pupil, and reduced sweating on one side of the face, known together as Horner syndrome.

Causes and risk factors

Ganglioneuroblastoma starts when developing nerve cells acquire changes that affect how they grow and mature. In most people, it is not possible to identify why this happened.

Most cases are not inherited. The genetic changes are usually found only within the tumour, meaning they developed during the person's lifetime and are not present throughout the body.

Rare inherited changes in genes such as ALK or PHOX2B can increase susceptibility to neuroblastic tumours. A family history of neuroblastoma or related tumours may therefore lead to genetic counselling. Having a genetic change does not mean that a tumour will definitely develop.

There are no well-established lifestyle causes. Nothing a parent did during pregnancy is known to be a usual cause of this condition.

Diagnosis

Diagnosis usually begins with an examination and imaging. Ultrasound may identify an abdominal mass. MRI or CT scans show where the tumour began, its size, nearby structures, and whether it is affecting the spine.

Urine or blood tests may measure the catecholamine breakdown products homovanillic acid, or HVA, and vanillylmandelic acid, or VMA. Raised levels can support the diagnosis, but they do not confirm it by themselves.

A biopsy is needed for confirmation. During a biopsy, a sample of tumour is removed and examined by a pathologist under a microscope. The pathologist identifies whether it is intermixed or nodular ganglioneuroblastoma and looks at how mature the cells appear.

Tumour testing may also examine features that help predict behaviour, including chromosome changes and the number of copies of the MYCN gene. These results can affect risk grouping and treatment.

Further tests establish the stage, meaning how far the condition has spread. They may include:

The healthcare team combines stage, pathology, age, and tumour genetics to place the condition into a risk group. The risk group is more useful for treatment planning than the tumour's name alone.

- MRI or CT scans of relevant body areas
- An MIBG scan, using a radioactive tracer absorbed by many neuroblastic cells
- A PET scan if the tumour does not take up MIBG
- Bone marrow samples when spread to the marrow is possible

Treatment and management

Treatment is planned by a team experienced in neuroblastic tumours. For children, this is

usually a paediatric cancer centre. Adults may need input from both adult cancer specialists and a centre familiar with these uncommon tumours.

Surgery is an important treatment for a localized tumour. The goal is to remove it without causing unacceptable damage to major blood vessels, nerves, or organs. Complete removal is not always safe or necessary.

An intermixed ganglioneuroblastoma with favourable findings may sometimes be managed with surgery alone. In selected low-risk situations, careful monitoring may be appropriate after a biopsy or incomplete removal. Follow-up is important because the remaining tissue still needs to be assessed over time.

Nodular ganglioneuroblastoma is generally managed according to the features of its neuroblastoma component. Treatment may include chemotherapy before or after surgery. Chemotherapy uses medicines that travel through the body to destroy rapidly growing tumour cells.

Higher-risk disease may require several forms of treatment. These can include intensive chemotherapy, surgery, high-dose chemotherapy followed by return of the person's own previously collected stem cells, radiation therapy, anti-GD2 immunotherapy, and treatment that encourages remaining cells to mature.

Radiation uses focused energy to damage tumour cells. It may be used for high-risk disease, a tumour that cannot be safely controlled in another way, or urgent pressure on the spinal cord. Its use is carefully planned to limit effects on nearby growing tissue.

Anti-GD2 immunotherapy helps the immune system recognize a substance found on many neuroblastic cells. It is used in particular high-risk treatment plans rather than for every ganglioneuroblastoma.

If the condition returns or does not respond fully, options may include different chemotherapy, targeted radioactive MIBG treatment, molecularly targeted medicines when the tumour has a relevant change, or a clinical trial.

Complications

The tumour can press on nearby organs, blood vessels, nerves, or the spinal cord. Spinal cord compression can lead to lasting weakness or problems with bladder and bowel control if it is not treated promptly.

Disease that spreads can affect bones, bone marrow, lymph nodes, the liver, or other areas. Bone marrow involvement can reduce normal blood-cell production, leading to anaemia, infections, or easy bleeding.

Treatment can also cause short-term and long-term effects. These depend on the therapies used and the person's age. Possible concerns include hearing loss, kidney or heart problems, thyroid changes, effects on growth, fertility problems, and learning or attention difficulties.

Not everyone develops these complications. Follow-up testing is chosen according to the tumour and the treatments received.

Prevention

There is no known way to prevent most ganglioneuroblastomas. They are not usually linked to lifestyle, diet, or an avoidable exposure.

Routine screening is not recommended for the general population. Screening may be considered by a specialist team when someone has a confirmed inherited susceptibility or a strong family history of neuroblastic tumours.

Genetic counselling can explain whether testing is appropriate and what a result could mean for the individual and their relatives.

Living with it and what to expect

The course can be very different between an intermixed, localized tumour and a nodular tumour with high-risk features. Localized tumours with favourable biology may be cured with limited treatment. Tumours with aggressive pathology, unfavourable genetics, or distant spread are more difficult to treat and may return.

During treatment, daily life may be affected by hospital visits, tiredness, appetite changes, infection precautions, pain, and time away from school or work. Rehabilitation can help with weakness, movement problems, or nerve damage.

After treatment, follow-up usually includes physical examinations and selected imaging or laboratory tests. Visits are often more frequent at first and become less frequent over time, although the exact schedule is individualized.

Follow-up is not only about recurrence. People treated during childhood may need long-term monitoring into adult life because some treatment effects appear years later. Care may include checks of hearing, growth, hormones, heart or kidney function, fertility, emotional wellbeing, and learning.

Many people return to school, work, physical activity, and ordinary family life. Some need ongoing support for treatment effects or uncertainty about recurrence. The likely course is

best understood from the exact pathology report, stage, tumour genetics, and response to treatment.

When to get medical help

Arrange a medical assessment for a new or growing lump, persistent unexplained abdominal or back pain, ongoing bone pain, an unexplained limp, or symptoms that do not settle. These symptoms have many possible causes and do not by themselves mean ganglioneuroblastoma.

During or after treatment, contact the treating team promptly about fever, increasing pain, new swelling, unusual bruising, worsening tiredness, or other changes covered by the care plan.

Seek urgent medical help for:

These symptoms can indicate pressure on the spinal cord or airway and should not wait for a routine appointment.

- New leg or arm weakness
- Difficulty walking or sudden loss of coordination
- New numbness, especially around the groin
- Loss of bladder or bowel control
- Severe or rapidly worsening back pain
- Significant trouble breathing

Common questions

Is ganglioneuroblastoma cancer?

It has the potential to behave as cancer, but its behaviour covers a wide range. Intermixed tumours may act in a relatively mild way, while nodular tumours can behave like neuroblastoma. The pathology and stage are essential for understanding an individual tumour.

Is it the same as neuroblastoma?

No. Both develop from the same family of immature nerve cells, but ganglioneuroblastoma also contains more mature cells. A nodular ganglioneuroblastoma includes a distinct neuroblastoma-like area, which can determine how it is treated.

Does every ganglioneuroblastoma need chemotherapy?

No. Some localized tumours with favourable findings can be managed with surgery and monitoring. Chemotherapy is more likely when the tumour has an aggressive component, cannot be safely controlled with surgery, or has spread.

Can adults get ganglioneuroblastoma?

Yes, but it is much more commonly diagnosed in children. Because adult cases are uncommon, treatment often draws on experience with paediatric neuroblastic tumours while considering adult health needs.

Can ganglioneuroblastoma come back?

It can. The chance is influenced by subtype, stage, tumour genetics, initial treatment, and response. Recurrence is less likely for favourable localized disease than for high-risk disease, but follow-up remains important.

Is ganglioneuroblastoma inherited?

Usually not. Rare families carry inherited changes that increase susceptibility to neuroblastic tumours. Genetic counselling may be considered when there is a family history, more than one affected relative, or another reason to suspect an inherited condition.

Sources

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