Carcinoid-NeuroEndocrine TUMOUR SOCIETY CANADA

SOCIÉTÉ DES TUMEURS Carcinoïdes-NeuroEndocrines DU CANADA

Neuroendocrine Tumours (NETs) Overview

- Neuroendocrine tumours (NETs) are abnormal growth of cells (neoplasms) that arise from cells of the diffuse neuroendocrine system.
- NETs are considered complex tumours because both tumor growth and hormonal function must be addressed.
- NETs are most common in the lung or gastrointestinal system, but they can also originate in other parts of the body such as the thyroid, thymus, pancreas, adrenals, paraganglia, ovaries and testes.
- NETs are classified as functional (produce hormones that cause symptoms), or non-functional **(may or may not produce** hormones and do not cause hormonal symptoms).
- The cause of NETs is currently unknown.

Symptoms and Diagnosis

- NETs can be difficult to diagnose because symptoms are often vague including diarrhea, flushing and nausea, depending on the type of tumour. Symptoms are intermittent and fluctuate in intensity.
- Patients commonly make many visits to the doctor over several years before an actual diagnosis is made; sometimes it takes **5-7** years from first symptoms to diagnosis.
- Many people have symptoms that mimic other conditions; therefore, NETs are often misdiagnosed as something else.
- Because NETs are poorly diagnosed, often by the time of diagnosis, the tumour has spread to other parts of the body (metastasis).
- Diagnosis of this type of cancer is complex and often requires sophisticated laboratory testing and scanning techniques.

Tests for Diagnosis

Blood tests - chromogranin A, gastrin, somatostatin, pancreastatin*, and other hormones specific to the various NET cancers.

Urine tests - 24 hour 5-HIAA - measures the metabolite of serotonin. Decreases may indicate a response to treatment.

Scans - Many of the scans use markers (hormones labeled with radioactive isotopes) that bind to somatostatin receptors to show images of the tumours. Common scans include: CT, MRI, Octreotide scan, MIBG, PET scan/Gallium 68* - improved resolution and higher sensitivity.

Scopes - bronchoscopy, endoscopy, colonoscopy.

Staging and Grading

- The stage and grade of NETs depends on well-defined histological features: size, lymphovascular invasion, mitotic counts, Ki-67 labelling index, invasion of adjacent organs, presence of metastases and whether they produce hormones.
- The grade includes looking at whether the tumour cells are well differentiated (look like normal cells) - grade 1 & 2, or poorly differentiated (abnormal looking cells) - grade 3.

Treatment Options

- NETs almost always become malignant. Surgical removal of very small, localized tumors is the only curative therapy. Regardless of therapy, disease recurs in 84% of patients after five years and 94% by 10 years. Studies have shown that people in rural areas and who are in poverty receive poorer care and treatment. Treatment options include:

Surgery - Debulking is the removal of as much as the tumour as possible. Even if not completely removed, this improves overall survival and quality of life. Having the liver resected increases the chances of living 5 years by 4 times and doubles the chances of living 10 years, after liver resection.

Ablative techniques - Radiofrequency that emits energy to kills the cancer cells and cryotherapy that uses liquid nitrogen or liquid carbon to freeze abnormal cells.

Radiation Therapy - External beam radiation and peptide receptor radionuclide therapy (PRRT)* - radiolabelled octreotide that binds to NETs.

Liver directed therapies – Embolization techniques - block or reduce blood flow to the tumour.

Somatostain analog therapy - octreotide (brand name - Sandostatin) and lanreotide (brand name -Somatuline) - Management of hormonal symptoms resulting from NETs.

Biologically targeted therapies - everolimus (brand name -Afinitor), an mTORC 1 inhibitor and sunitinib (brand name -Sutent), a tyrosine kinase inhibitor, attack cancer cells while limiting damage to normal cells.

Chemotherapy

Interferon Therapy

(* Not available in Canada)