Aretha Franklin's Cancer Diagnosis Clarified

The renowned singer died of a neuroendocrine tumor in the pancreas.

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Last month, it was widely reported by Cancer Health and others that Aretha Franklin, the "Queen of Soul," had died of pancreatic cancer at age 76. But the reality is a bit more complicated. Franklin had a pancreatic neuroendocrine tumor (pNET), according to her doctor, Philip A. Philip, MD, PhD, of the Karmanos Cancer Institute in Detroit.

Neuroendocrine tumors in the pancreas originate in the pancreatic islets (also known as islets of Langerhans), clusters of endocrine cells that produce hormones, including insulin and glucagon, which regulate blood sugar. NETs account for only around 5 percent of all cancers in the pancreas.

Pancreatic cancer, in contrast, typically refers to adenocarcinoma of the exocrine cells and ducts that produce and transport pancreatic enzymes that help digest food. This type of cancer is often diagnosed at an advanced stage, is difficult to treat and has a high mortality rate.

Diagnosis, treatment and prognosis of NETs in the pancreas differ from those of pancreatic adenocarcinoma. "Unlike the common variety pancreatic cancer, patients with pNETs on average survive significantly longer and have an increasing number of treatment options," Philip said.

J. Leonard Lichtenfeld, MD, deputy chief medical officer of the American Cancer Society, elaborated on the differences in a blog post following Franklin's death. He said the organization will revise its widely used informational website, cancer.org, to separate out pNETs from pancreatic cancer.

"The typical pancreatic cancer is almost always fatal, and unfortunately life is too frequently short following a diagnosis," Lichtenfeld wrote. "NETs can be cured or if extending beyond the pancreas can still be associated with a survival much longer than typical pancreatic cancer. The treatment is different, the outcome is different, the cure rate is different."

Advocates for people with neuroendocrine cancer called attention to the distinction in an effort to bring more awareness to this under-recognized disease. Like Franklin, Apple cofounder Steve Jobs also had neuroendocrine cancer in the pancreas that was widely reported as pancreatic cancer.

NETs can also develop in other endocrine glands including the adrenal glands (known as pheochromocytoma) and thyroid, as well as in neuroendocrine cells in the skin (known as Merkel cell carcinoma), gastrointestinal system (known as carcinoid tumors) and lungs. Neuroendocrine tumors can produce abnormally high amounts of hormones, leading to a wide variety of symptoms.

Although rare, the incidence of neuroendocrine cancer appears to be rising. More than 12,000 people in the United States are diagnosed with neuroendocrine tumors each year, according to the American Society of Clinical Oncology (ASCO). The average time to diagnosis is around five years due to a lack of awareness about the disease in the medical community.

“Awareness about this disease needs to occur at all levels from physicians, to nurses, to the general public,” said Eric Liu, MD, of Sarah Cannon Cancer Institute in Denver, cofounder of the Healing NET Foundation. “The number of people with neuroendocrine cancer is on the rise and they can only get the best care when there is a better understanding that it is a different type of cancer. When well treated, people can live quite a long time. But in some cases, it is very aggressive. Good care starts with more education.”

Click here to learn more about neuroendocrine tumors from ASCO.

For more information:

Carcinoid Cancer Foundation

Healing NET Foundation

Neuroendocrine Cancer Awareness Network

Neuroendocrine Tumor Research Foundation

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