The Challenge of Rare Cancers in Current and Advancing Treatment

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The term *rare cancers* is relatively new in oncology, with significant presence in cancer literature occurring only since the early 2000s. Epidemiologic studies in Europe and the United States show that rare cancers account for about 25% of adult cancers (Eslick, 2012). However, rare cancer types have been traditionally understudied, with an associated lack of progress in survival and challenges in decision making for patients, physicians, and policy makers (Greenlee et al., 2010). Initiatives led within the European Union and with the United States are intended to boost progress in treatment for rare cancers through collective research and shared databases (Keat et al., 2013). Nurses see patients with rare cancers struggle for access to quality, evidence-based care, as well as the isolation they endure as they seek the information and support needed to cope with the diagnosis of a life-threatening illness.

Background

The definition of rare cancer varies internationally. Surveillance of Rare Cancer in Europe defines a rare cancer as having an estimated annual incidence rate of 108 cases per 100,000 people for all rare cancers. This incidence corresponds to 541,000 new diagnoses per year, representing 22% of all cancer diagnoses and 24% of total cancer prevalence in Europe (Gatta et al., 2011). In the United States, rare cancers are defined as fewer than 15 cases per 100,000 people or 150 cases per 1,000,000 people per year, corresponding to fewer than 40,000 new cases per year. Sixty of 71 cancer types are considered rare, accounting for 25% of all adult tumors (Greenlee et al., 2010). Rare cancers include all pediatric cancers, all hematologic cancers, and solid tumors of low incidence.

Associated Factors for Rare Cancer Incidence

Rare cancers occur with greater relative frequency among people who are of a younger age and of Hispanic ethnicity, as well as those who are non-Caucasian. Five-year survival rates are generally lower than for common cancers and worsens with age, which is thought to be associated with factors inherent to the disease, as well as inadequacies of care or treatment, delayed diagnosis, lack of effective therapies, or lack of evidence-based treatment guidelines (Gatta et al., 2011).

Dilemmas With Rare Cancers

A study by Wagland, Levesque, and Connors (2015) highlighted the dilemmas of a rare cancer diagnosis compounded by social and geographic factors and limited access to specialized care. The study design included retrospective interviews with women treated for multiple myeloma. Participants were a mean age of 48 years, had dependent children living at home, lived in a rural area, and traveled a distance of 140–3,000 km to a specialist metropolitan hospital for treatment. All spent one to five months away from home and had at least one stem cell transplantation.

Analysis of the interviews revealed three themes: isolation from living with an unknown cancer, isolation within the myeloma population, and isolation because of disease and treatment effects. According to the researchers, the impact of having a rare cancer emerged as a core issue in the study. Most participants had never heard of their illness before diagnosis and found a nonexistent understanding among the public, family members, and local healthcare professionals. This impeded their ability to comprehend complex treatment options and an uncertain prognosis. They felt isolated from others with more common cancers who they witnessed receiving far greater support from their community, their physicians, and their family members. In addition, because of their younger age, they were unable to find support from other myeloma patients. Extended travel to specialized hospitals was burdensome on families and relationships, but the hospitals offered support and recognition from an educated healthcare team and other informed patients. Wagland et al. (2015) made recommendations to diminish disease isolation that included ongoing support from the specialized treatment team, public and professional education to increase awareness of the rare cancer to improve support services and decrease stigma, and strategies to connect patients with comparable peers via the Internet and face-to-face meetings to maximize available social support.

Search for Effective Treatments

Effective treatment for rare cancers has lagged because of a lack of significant numbers for widespread development of clinical expertise, as well as a lack of adequate funding and interest in clinical trials. Recognition of the public health burden of rare cancers in Europe and the United States (i.e., that they may be rare individually but not collectively) is leading to greater focus on improving treatment, prevention, and control. In Europe, discussion is underway for

ONF, 42(5), 566–567.
doi: 10.1188/15.ONF.566-567