Tagraxofusp Treatment

Implications for patients with blastic plasmacytoid dendritic cell neoplasm

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BACKGROUND: Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare, clinically aggressive, and often fatal hematologic malignancy. BPDCN is not a new entity, but it has been renamed and reclassified, which, in part, contributes to it being underrecognized. In 2018, tagraxofusp became the first U.S. Food and Drug Administration–approved therapy for BPDCN.

OBJECTIVES: This article aims to educate oncology nurses about tagraxofusp's dosing regimen, side effects, and how to manage patients undergoing treatment in inpatient and outpatient settings.

METHODS: The authors reviewed content related to the safety and clinical management of tagraxofusp, as well as content that supports patient and provider education.

FINDINGS: Capillary leak syndrome (CLS) is the most serious adverse event reported with tagraxofusp; therefore, nurses should stop tagraxofusp administration until all CLS-related symptoms have resolved. Hypersensitivity reactions and hepatotoxicity have also been observed in patients treated with tagraxofusp and should be monitored during treatment cycles.

KEYWORDS
capillary leak syndrome; blastic plasmacytoid dendritic cell neoplasm; tagraxofusp

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BLASTIC PLASMACYTOID DENDRITIC CELL NEOPLASM (BPDCN) is a clinically aggressive and often fatal hematologic malignancy that involves bone marrow, peripheral blood, the central nervous system, and skin (Pemmaraju, 2017; Riaz et al., 2014). This malignancy of plasmacytoid dendritic cell precursors progresses rapidly and carries a poor prognosis (Pagano et al., 2016; Pemmaraju, 2017). BPDCN was originally known as agranular CD4+ natural killer (NK) cell leukemia in 1995 (Brody et al., 1995), and naming conventions were updated to blastic NK-cell lymphoma in 2001 with the World Health Organization (WHO) classification based on blastic appearance and CD56 expression (Chan et al., 2001). One other name, noted in 2005 by the WHO and the European Organisation for Research and Treatment of Cancer, was CD4+CD56+ hematodermic neoplasm, which was based on immunophenotype and skin involvement (Willemze et al., 2005). In 2008, the WHO solidified the name as BPDCN after decades of changing nomenclature and placed it under acute myeloid leukemia and related family of neoplasms (Facchetti et al., 2008). BPDCN is commonly mistaken for other hematologic malignancies and has only recently been named in a consistent manner for classification purposes (Arber et al., 2016). The exact incidence of BPDCN is unknown, and registries likely underestimate the true incidence of BPDCN (Pagano et al., 2016; Sweet, 2020); however, one study suggests that the overall incidence of BPDCN is 0.04 cases per 100,000 population (Guru Murthy et al., 2018).

Key facts related to BPDCN are presented in Figure 1. Diagnosis of BPDCN occurs via flow cytometry and/or immunohistochemistry on a skin biopsy or bone marrow aspirate to detect the triad of immunophenotype markers (CD123, CD4, CD56) followed by morphologic assessment (Pagano et al., 2016; Riaz et al., 2014). Bone marrow, peripheral blood and skin, central nervous system, lymph nodes, and viscera are commonly involved in BPDCN (Riaz et al., 2014). BPDCN clinical presentation includes skin lesions that are often asymptomatic, which represents the cutaneous pathway. Leukemic dissemination is where blasts are present in the bone marrow and peripheral blood (Pagano et al., 2016; Pemmaraju, 2017; Reichard, 2013).

Patients with BPDCN have been typically treated with combination chemotherapy regimens generally used for lymphoma or acute leukemia and, historically, standard frontline therapy has not been established