Rosai-Dorfman disease is a rare disorder characterized by overproduction (proliferation) and accumulation of a specific type of white blood cell (histiocyte) in the lymph nodes of the body (lymphadenopathy), most often those of the neck (cervical lymphadenopathy). In some cases, abnormal accumulation of histiocytes may occur in other areas of the body besides the lymph nodes (extranodal). These areas include the skin, central nervous system, kidney, and digestive tract. The symptoms and physical findings associated with Rosai-Dorfman disease vary depending upon the specific areas of the body that are affected. The disorder predominantly affects children, adolescents or young adults. The exact cause of Rosai-Dorfman disease is unknown. Some studies suggest that Rosai-Dorfman disease affects males more often than females. The disorder can affected individuals of any age, but most often affects young adults under the age of 20. Cutaneous Rosai-Dorfman disease occurs more often in females in their 20s or 30s. More than 650 cases have been reported in the medical literature since the disorder's first description in the medical literature in 1969. The diagnosis of Rosai-Dorfman disease may be confirmed by a thorough clinical evaluation, a detailed patient history and a variety of specialized tests, such as surgical removal and microscopic examination of affected tissue (biopsy).