Rosai-Dorfman disease (RDD), previously also known as sinus histiocytosis with massive lymphadenopathy (SHML), is a rare histiocytic disorder. RDD is characterized by accumulation of abnormal cells (histiocytes) in various tissues/organs of the body. RDD tends to affect skin and lymph nodes most commonly, although any organ system can be involved from head to toe. The reason that these cells over-produce is not known, although many possibilities have been considered, including viral, bacterial, infection, environmental, and genetic causes. The exact incidence of RDD cases is not known, although it does occur worldwide and seems to affect equal numbers of males and females. It can affect children as well as adults. Because this disease is so rare, no large studies have been performed, and there is no approved, widely-accepted treatment.

Symptoms of RDD include skin nodules, lymph node enlargement (with or without pain), and fever.

By frequency, RDD can involve the skin (50%), lymph nodes (30-50%), bones (15%), respiratory system including nasal sinuses and airways (10-20%), skull and nervous system (10%), eye/eye socket (5%), salivary glands (5%), breast (1-2%), and liver/spleen (1-2%).

RDD can also occur in association with autoimmune conditions (lupus, rheumatoid arthritis) or other blood cancers (lymphoma, histiocytic disorders like Erdheim-Chester disease or Langerhans cell histiocytosis).

The diagnosis of RDD is made following a biopsy of the affected tissue. This procedure can be performed of the lymph nodes, skin, bone, liver, lung, or bone marrow. A small piece of the tissue is obtained so that it can be viewed under a microscope by a pathologist.

Learn more about RDD and help us spread awareness!