Abstract: Atherosclerosis, a chronic inflammatory disease of the vessel wall and the underlying cause of cardiovascular disease, is initiated and maintained by innate and adaptive immunity. Accumulating evidence suggests an important contribution of autoimmune responses to this disease. Plasmacytoid dendritic cells (pDCs), a specialized cell type known to produce large amounts of type I interferons (IFNs) in response to bacterial and viral infections, have recently been revealed to play important roles in atherosclerosis. For example, the development of autoimmune complexes consisting of self-DNA and antimicrobial peptides, which trigger chronic type I IFN production by pDCs, promote early atherosclerotic lesion formation. pDCs and pDC-derived type I IFNs can also induce the maturation of conventional DCs and macrophages, and the development of autoreactive B cells and antibody production. These mechanisms, known to play a role in the pathogenesis of other autoimmune diseases such as systemic lupus erythematosus and psoriasis, may also affect the development and progression of atherosclerotic lesion formation. This review discusses emerging evidence showing a contribution of pDCs in the onset and progression of atherosclerosis.