Sinus histiocytosis with massive lymphadenopathy (SHML) is a rare, benign disease of unknown aetiology. This disease typically presents with massive, painless cervical lymphadenopathy but may occur in a wide variety of extranodal sites. This report describes a 43-year old man with a left submandibular swelling and left lateral cervical lymphadenopathy clinically suggestive of a malignancy. Initial cytological examination of a fine needle aspirate specimen taken from the mass suggested a chronic granulomatous inflammatory lesion. A definitive diagnosis of SHML was however made only after excisional biopsy of the lesion. The patient was followed up for 6 months and no recurrence of the lesion was observed. SHML may be considered a rare lesion among Nigerians. Knowledge of its clinical presentation with understanding of the differentials diagnosis is important to avoid unnecessary intervention. The diagnosis can be made from FNAC, histopathology and immune-histochemistry. There is no ideal protocol for the treatment. Follow-up is however necessary to detect relapses.

**Keywords:** Sinus histiocytosis; lymphadenopathy; Nigerian