A 41-year-old man who presents for evaluation of right arm cyst (1 cm in diameter). The cyst has been present for 3 months and grown slightly since then. No associated bleeding or drainage from the lesion.
Mass, right arm, excisional biopsy (H&E, x20)

Contributed by Dr. Donald Tschirhart
Mass, right arm, excisional biopsy (H&E, x200)

Contributed by Dr. Donald Tschirhart
Mass, right arm, excisional biopsy (H&E, x400)

Contributed by Dr. Donald Tschirhart
Mass, right arm, excisional biopsy (H&E, x400)

Contributed by Dr. Donald Tschirhart
Mass, right arm, excisional biopsy (IHC)

CD68 (KP-1)  

S100  

Contributed by Dr. Donald Tschirhart
LYMPH NODE, RIGHT ARM, EXCISIONAL BIOPSY:
- NODULAR HISTIOCYTIC PROLIFERATION, COMPATIBLE WITH ROSAI-DORFMAN DISEASE.
Sinus histiocytosis with massive lymphadenopathy

• This rare disease was initially described by Destombes in 1965. Rosai and Dorfman recognized this disease as a distinct clinicopathological entity in 1967 and coined the term *sinus histiocytosis with massive lymphadenopathy* (SHML).

• Although first described as “massive, bilateral and mostly cervical lymphadenopathy”, this disease in our case presented as a single slow growing mass of the right upper arm. It has a small orifice clinically suggestive of a benign inclusion cyst.

• Therefore, not every SHML has “massive lymphadenopathy”.
Histology & Immunohistochemistry of SHML

• This case shows the classical morphology of Rosai-Dorfman disease with sinus histiocytic proliferation and emperipolesis of small lymphocytes and plasma cells.

• The histiocytes usually have a single nucleus with centrally located distinct nucleolus, but in our case the binucleated histiocytes with engulfed lymphocytes are also seen (pointed with red arrow).

• The histiocytes are positive for all the histiocytic markers including CD4, CD14, CD68 (KP1), and CD163. They almost always express S100, but not CD1a.
Treatment & prognosis

• It is considered a benign lymphadenopathy which is self-limited and eventually recedes.
• It occasionally collide with other malignant lymphomas in the same lymph node.
• It can be fatal if it is locally aggressive and involves vital organs or associated with immune dysfunctions.
• Combinations of corticosteroids, chemotherapeutic agents, and radiotherapy have been used for aggressive diseases, but no effective treatment is currently available.