MULTICENTRIC RETICULOHISTIOCYTOSIS ELECTIVE EXCISION OF SYMPTOMATIC HAND NODULES WITH ONE-YEAR FOLLOW-UP

Presenter: Matthew Mead, MD
Authors: Matthew Mead, MD; Travis Baes, MD; George Dass, MD

Background: Multicentric reticulohistiocytosis (MRH) is a rare, non-Langerhans histiocytosis that is characterized by multiple skin lesions and destructive polyarthritis. To date, there have been approximately 300 reported cases in the literature and the pathogenesis remains unclear. MRH has a female predominance (3:1) and most commonly begins during the 4th decade, lasting 5-10 years before remittance. During this time, there is significant destruction of multiple joints with potential involvement of the skin, muscle, heart, lungs and gastrointestinal tract as well. It is an extremely destructive inflammatory polyarthritis and can result in arthritis mutilans in over 45% of cases. Currently, there is no diagnostic serologic test and no consensus on management.

CASE: We describe a case of a 56-year-old right-hand dominant female who presented with multiple soft nodules to bilateral hands which had developed in the last 3-4 years. She was being treated by a Rheumatologist with a current regimen of leflunomide, infliximab, and ibuprofen, resulting in only moderate control of her symptoms. Overall, the nodules in her hands were not painful, with the exception of several larger nodules which were causing mechanical irritation with pinch and writing. After discussion of risks, including recurrence of these nodules, the patient elected to have three of these excised surgically. These were sent for pathology revealing a diffuse proliferation of histiocytic cells, characterized by vesicular chromatin, prominent nucleoli, and abundant amphophilic cytoplasm with associated multinucleated giant cells. The histiocytes were CD68 and lysozyme positive, and S100, Melan-A, and AE1/AE3 negative. The patient was seen one year post-operatively. One out of three nodules have reoccurred, while the other two have remained quiescent. She has resumed working and notes reduced pain with pinch grip. She states she would have the procedure again if needed and overall is satisfied with her result.

Discussion: This case demonstrates a rare case of multicentric reticulohistiocytosis. While not curative, excision of several of her hand nodules resulted in significant relief from mechanical irritation associated with daily activities. To our knowledge, selective excision of symptomatic hand nodules in patients with MRH has not been described in the medical literature. In our case, 1 of 3 nodules did reoccur. Despite this, with appropriate pre-operative counseling this procedure has the potential to produce significant pain relief and improve quality of life in a disease process where optimal medical regimen remains to be determined.
Figure 1: Patient’s left hand, depicting characteristic soft tissue nodules which in this patient produced discomfort with pinch and holding a writing instrument.

Figure 2: Representative histology from specimen, depicting histiocytic infiltrate including multinucleated cells (H&E; original mag 200x)