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Editorial

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Editorial

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The three cases of adult xanthogranulomatous disease reported herein highlight the difficulty in recognizing and defining clearly these rare disorders. The adult xanthogranulomas fit into Class II histiocytic disorders, the so-called, non-Langerhans disorders (Table 1) (Zelger et al., 1994). We have recently undertaken a multi-institutional review of adult xanthogranulomatous disorders of the orbit that involved a total of 137 cases, which fell within four syndromes (Sivak-Callcott et al., 2006). These included, in order of frequency, necrobiotic xanthogranuloma (NBX), Erdheim-Chester disease (ECD), adult-onset asthma with periocular xanthogranuloma (AAPOX) and adult-onset xanthogranuloma (AOX). (Jakobiec et al., 1993; Bullock et al., 1986; Alper et al., 1983) These syndromes are often associated with other systemic manifestations, which serve as a basis for separating them. They have in common a histopathology including xanthoma cells, Touton giant cells, lymphocytic infiltration and varying degrees of fibrosis with occasional necrosis, particularly noted in NBX. They all occur in middle-aged individuals and usually have subcutaneous, subconjunctival or periocular xanthomatous-appearing (yellow) infiltrates.

AAO is isolated to the anterior orbit without systemic involvement. In contrast, AAPOX is characterized by an association with adult-onset asthma, lymphadenopathy and often an increased IgG level, suggesting an underlying stimulation of the B-cell population (Sivak-Callcott et al., 2006; Jakobiec et al., 1993) This is also evident in the systemic association of NBX, which tends to have ulcerated subcutaneous xanthomas and frequent paraproteinemia or multiple myeloma (Ugurlu et al., 2000; Mehregan and Winkelmann, 1992; Cornblath et al., 1992) Finally, ECD is associated with dense fibrosclerosis of the deeper orbit in contrast to the other three syndromes, which are more anterior in the orbit. ECD is also frequently associated with mediastinal, pericardial, pleural, perinephric and retroperitoneal fibrosis leading to death in two-thirds of the patients reported. In addition, characteristic bone involvement is seen (Murray et al., 2001; Scheer et al., 2000).

Early diagnosis and treatment remains a significant challenge in these syndromes. The best results having been obtained with a form of multi-agent chemotherapy with or without radiation or surgery.

Our histopathologic study of these cases suggested the histiocytes and fibroblasts might be stimulated by T-cells; thus several of our patients were treated successfully with T-cell as well as B-cell suppressors based on the local T-cell-rich environment and the systemic B-cell associations.

In order to avoid confusion and delay in diagnosis of these syndromes, it is important to recognize the underlying histopathology and to carefully assess the patients for systemic associations. As many of these associations may help to define the syndromes and may point to more appropriate treatment, it is important to contextualize within the systemic pattern of presentation.

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TABLE 1 **Non-Langerhans Disorders of Histiocytes**

Xanthogranulomatous disease of adults
Juvenile xanthogranuloma
Benign cephalic histiocytosis
Papular xanthoma
Multicentric reticulohistiocytosis
Xanthoma disseminatum
Reticulohistiocytoma
Generalized eruptive histiocytoma
Sinus histiocytosis with massive lymphadenopathy
Self-healing reticulohistiocytosis
Indeterminate cell histiocytosis

Data from Zelger B, Cerio R, Orchard G, Wilson-Jones E. Juvenile and adult xanthogranuloma. A histological and immunohistochemical comparison. Am J Surg Pathol 1994;18:126–135.

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