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Study Examines Relationship Between Autoimmune Skin Disease and Neurologic Disorders

Individuals with the autoimmune skin disease bullous pemphigoid appear more likely to have a diagnosis of neurologic disease, such as dementia and cerebrovascular disease, according to a report in the November issue of Archives of Dermatology, one of the JAMA/Archives journals.

"Bullous pemphigoid is a debilitating autoimmune skin disease that is characterized by large, tense blisters on the skin of the elderly," the authors write as background information in the article. The condition affects about 43 per million individuals per year in the United Kingdom and 7 to 13 per million in other parts of Europe. "Over the last two decades, various neurologic diseases have been described in association with bullous pemphigoid, including cerebrovascular disease, dementia, multiple sclerosis, epilepsy, Parkinson disease, Shy-Drager syndrome and amyotrophic lateral sclerosis."

Kathy Taghipour, M.D., M.R.C.P., of Oxford Radcliffe Hospitals, Oxford, England, and colleagues assessed 90 consecutive patients with bullous pemphigoid and 141 controls without the condition. Medical histories were reviewed for the presence of neurologic disease, defined as a confirmed diagnosis by a hospital physician; positive imaging findings; or considerable functional disability due to mental impairment.

Among patients with bullous pemphigoid, 42 (46 percent) had at least one neurologic disease, compared with 16 controls (11 percent). Four major neurologic diseases were observed among these patients: cerebrovascular disease, dementia, Parkinson's disease and epilepsy. However, only rates of cerebrovascular disease and dementia were significantly greater among patients than among controls.

Of the 36 patients with accurate information about the timing of their diagnoses, bullous pemphigoid was diagnosed after neurologic disease in most (26 of 36 or 72

percent), with a median (midpoint) time of 5.5 years between diagnoses.

"Neurologic symptoms may often be subtle, and the onset of disease may be insidious, leading to diagnostic delay," the authors write. "It is therefore likely that the interval between neurologic disease and bullous pemphigoid is longer than estimated, and it is tempting to speculate that certain neurologic conditions can predispose to bullous pemphigoid." The association could be explained by an immunologic cross-reactivity between the skin and the brain, in which neurologic disorders trigger an immune response that affects the skin, the authors note.

"The relationship between bullous pemphigoid and neurologic disease has been the subject of numerous case reports, but epidemiological data are scarce, and this subgroup of patients with bullous pemphigoid have not been further characterized," they conclude. "We have demonstrated that there is a significant association between bullous pemphigoid and neurologic disorders, in particular cerebrovascular disease and dementia, and that neurologic disease may be a predisposing factor for bullous pemphigoid. The mechanism by which neurologic disease may trigger bullous pemphigoid remains obscure and requires further investigation."

Source: Kathy Taghipour; Ching-Chi Chi; Angela Vincent; Richard W. Groves; Vanessa Venning; Fenella Wojnarowska. The Association of Bullous Pemphigoid With Cerebrovascular Disease and Dementia: A Case-Control Study. *Archives of Dermatology*, 2010; 146 (11): 1251-1254 DOI: 10.1001/archdermatol.2010.322