Small Fiber Neuropathy causing chronic generalized pruritus: Should Dermatologist screen for it?

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CASE REPORT

History of Present Illness: A 65 y/o Caucasian female presented to clinic with a two-year history of generalized chronic pruritus. Previous treatment includes Triamcinolone .1% cream and Pramoxine cream for several months without improvement. She also reported electric “shooting” pains and burning in bilateral upper and lower extremities, most severe in the evening.

Past Medical History:
Hypertension, Cholelithiasis, uncontrolled Type II Diabetes, Bladder incontinence

FINDINGS

Physical Exam: Excoriated erythematous urticarial papules and plaques on back, upper arms, and upper thighs with sparing of central back (Figures 1,2)

Diagnosis:
• Punch biopsy specimen submitted for H&E (Figure 3) revealed a superficial and deep perivascular and interstitial infiltrate with eosinophils
• Neurology performed two punch biopsies which were submitted for epidermal nerve fiber density studies. Frozen sections of the tissue was stained with PGP 9.5, an immunohistochemical stain targeting peripheral nerve fibers. The study showed significantly reduced epidermal nerve fiber density (Figures 4,5) consistent with the diagnosis of Small Fiber Neuropathy. Figure 6 demonstrates normal density of intraepidermal nerve fibers for comparison.

DISCUSSION
• Small fiber neuropathy (SFN) is caused by damage to small, unmyelinated and myelinated fibers in peripheral nerves
• These nerves convey pain and temperature sensations from the skin and mediate autonomic functions. Dysfunctions in these nerve fibers can cause symptoms such as pruritus, numbness, burning, paresthesias, and dysesthesias
• Diagnosis requires examination of the small nerve fibers in the skin using staining with PGP9.5 axonal marker. The standard biopsy sites for diagnosis include the thigh, distal leg, and dorsal foot.
• Epidermal Nerve Fiber Density (ENFD) is a standardized test that measures density of small sensory nerve fibers in the skin after staining with PGP9.5.
• Underlying causes include systemic diseases, the most common being diabetes mellitus or glucose intolerance. Other causes include metabolic syndrome, hypothyroidism, autoimmune rheumatologic disorders such as lupus, vasculitis, sarcoidosis, inflammatory bowel disease, nutritional deficiency (B12, B6, B1 vitamins), celiac disease, Lyme disease, HIV/HCV infection, alcohol abuse, amyloidosis, and drug/toxin exposure. 40% of cases are idiopathic.
• Treatment is aimed at symptom control (gabapentin, topical lidocaine or capsaicin) and treating the underlying cause. Acute onset cases have responded to prednisone.
• Chronication processes may lead to refractory pruritus, thus treatment should be started as soon as possible.

REFERENCES