The Possible Association Of Pyoderma Gangrenosum, Kartagener's Syndrome And Rheumatoid Arthritis

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Citation

Abstract
We describe the case of a 67-year-old male with Kartagener's syndrome and rheumatoid arthritis who presented with pyoderma gangrenosum. This is the second such report in the literature. The association between Kartagener's syndrome and pyoderma gangrenosum, which is probably due to neutrophilic dysfunction caused by the primary ciliary dyskinesia in Kartagener's syndrome, is discussed.

CASE REPORT
A 67-year-old male known to have Kartagener's syndrome (Fig. 1), rheumatoid arthritis for 20 years, severe steroid-induced osteoporosis and hypogonadotropic hypogonadism presented with bilateral ankle pyoderma gangrenosum (Fig. 2).

Figure 1
Figure 1: Chest x-ray showing dextrocardia, the typical radiographic sign of Kartagener's syndrome.

Figure 2
Figure 2: Multiple lesions of pyoderma gangrenosum.

Skin biopsy specimens from the margins of the ankle ulcers demonstrated mixed cellular inflammation with neutrophil...
predominance, compatible with the diagnosis of pyoderma
gangrenosum. The patient was treated systemically with 15
mg of prednisone and locally with normal saline wraps and
sodium cromoglycate solution, resulting in a decrease in the
size of the lesions and new granulation. The patient later
died due to pulmonary complications.

The pathogenesis of pyoderma gangrenosum is poorly
understood, although neutrophil dysfunction (i.e., defects in
chemotaxis or hyperreactivity) has been suggested.\textsuperscript{1,2}
Abnormal neutrophil trafficking and metabolic oscillations
were recently described in a patient.\textsuperscript{3} Furthermore,
interleukin-8 (IL-8), a potent leukocyte chemotactic agent,
was found to be over-expressed in pyoderma gangrenosum
ulcers and to induce similar ulceration in skin xenografts
transfected with recombinant human IL-8.\textsuperscript{4}

A review of the literature disclosed an association between
primary ciliary dyskinesia, Kartagener's syndrome and
cutaneous manifestations (including pyoderma gangrenosum):
Brenner et al.\textsuperscript{5} described a case of persistent deep-seated
folliculitis and impaired chemotaxis in a 24-year-old woman
with Kartagener's syndrome. Vazquez et al.\textsuperscript{6} reported three
types of cutaneous lesions — recurrent outbreaks of
nummular eczema, recurrent deep folliculitis, and two
episodes of pyoderma gangrenosum — in a 47-year-old man
with Kartagener's syndrome. This was the second report of a
case involving Kartagener's syndrome and pyoderma
gangrenosum.

The pathologies exhibited by our case can be tied together
and explained by neutrophil dysfunction, which has been
described in both Kartagener's syndrome and pyoderma
gangrenosum, as noted above. The connection between
rheumatoid arthritis, Kartagener's syndrome and pyoderma
gangrenosum, as seen in our case, is supported by the known
association between rheumatoid arthritis and pyoderma
gangrenosum, and the frequent occurrence of autoimmune
diseases like rheumatoid arthritis in Kartagener's syndrome
due to prolonged stimulation of the immune system by
recurrent infections from the bronchiectasis. While it is
possible that the association between the three entities is an
indirect one (connected through rheumatoid arthritis), we
suggest that the literature mentioned above,\textsuperscript{1,2,3,4} argues for a
direct connection between defective neutrophils in
Kartagener's syndrome and pyoderma gangrenosum. Unlike
the well-known ciliary dyskinesia underlying the
pathomechanism of neutrophil dysfunction in Kartagener's
syndrome, the cause of neutrophil dysfunction in pyoderma
gangrenosum has not yet been established.

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