

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

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## 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.<sup>1,2</sup> Abnormal neutrophil trafficking and metabolic oscillations were recently described in a patient.<sup>3</sup> 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.<sup>4</sup>

A review of the literature disclosed an association between primary ciliary dyskinesia, Kartagener's syndrome and cutaneous manifestations (including pyoderma gangrenosum): Brenner et al.<sup>5</sup> described a case of persistent deep-seated folliculitis and impaired chemotaxis in a 24-year-old woman with Kartagener's syndrome. Vazquez et al.<sup>6</sup> 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,<sup>1,2,3,4</sup> 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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