The Role of Undiagnosed Primary Immunodeficiency in Orthopaedic Infection

LifeBridge Health
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Introduction

- Primary immunodeficiency diseases (PID) are heterogeneous group of genetic immune disorders, characterized by defects in one or more components of the immune system.
- In 34 percent, the age of onset is prior to age 10 and the remaining 66% present in the third decade; the average delay in diagnosis is 4-6 years.
- Delay in diagnosis can have serious consequences such as bacterial, fungal, or viral infections.
- PID should be suspected with recurrent, unusual or persistent infections.
- Host optimization for orthopedic infection is necessary and may require consideration beyond the obvious especially in patients without any of the apparent risk factors.
- The purpose of this study was to determine the incidence of laboratory immunological abnormalities observed in our infected orthopedic patient population.

Patients and Methods

- Retrospective chart review (2013–2016) performed to capture all orthopedic infection patients with available immunological laboratory results.
- Patients were excluded if they were undergoing immunosuppressive treatment.
- Medical history was recorded to stratify patient host status according to the Cierny-Mader classification system.
- Clinically significant results were defined as low IgG, low IgM, high IgE, low IgA, low C3, low C4, low total complement, positive ANCA, and positive rheumatoid factor.

Table 1. Patient host status characteristics

	Type A Host	Type B Host	Total Group
Host Status N(%)	18(34)	35(66)	53
Age Mean±SD	49.9±15.2	59.6±11.6	56.5±13.5
Male Gender N(%)	5(28)	16(46)	21
IGG Replacement N(%)	3(17)	2(6)	5(9)

Results

- In total, 53 patients with active infection from our institution were included in this analysis, 18 (34%) were type A, and 35 (66%) type B hosts.
- Thirteen (72%) of the type A hosts and 25 (71%) of the type B hosts had clinically significant abnormalities.
- Five (28%) of the type A hosts and 6 (17%) of the type B hosts had IgG deficiency.
- All patients with clinically significant abnormalities received a hematology/ immunology referral.
- Five (9%) subjects were prescribed subsequent IGG replacement therapy by a specialist.

Table 2. Laboratory assessments – clinically significant abnormalities

	Total Group N(%)	A Hosts N(%)	B Hosts N(%)
ANCA Atypical	1(2)	1(6)	0(0)
ANCA MPO	1(2)	0(0)	0(0)
Complement Total	3(6)	1(6)	2(6)
C-ANCA	0(0)	0(0)	0(0)
P-ANCA	1(2)	0(0)	1(3)
C3	5(17)	1(6)	4(11)
C4	3(6)	2(11)	1(3)
IGG	11(21)	5(28)	6(17)
IGM	12(23)	5(28)	7(20)
IGE	21(40)	5(28)	16(46)
IGA	4(8)	2(11)	2(6)
Rheumatoid Factor	3(6)	1(6)	2(6)
Any Significant Abnormalities	38(72)	13(72)	25(71)

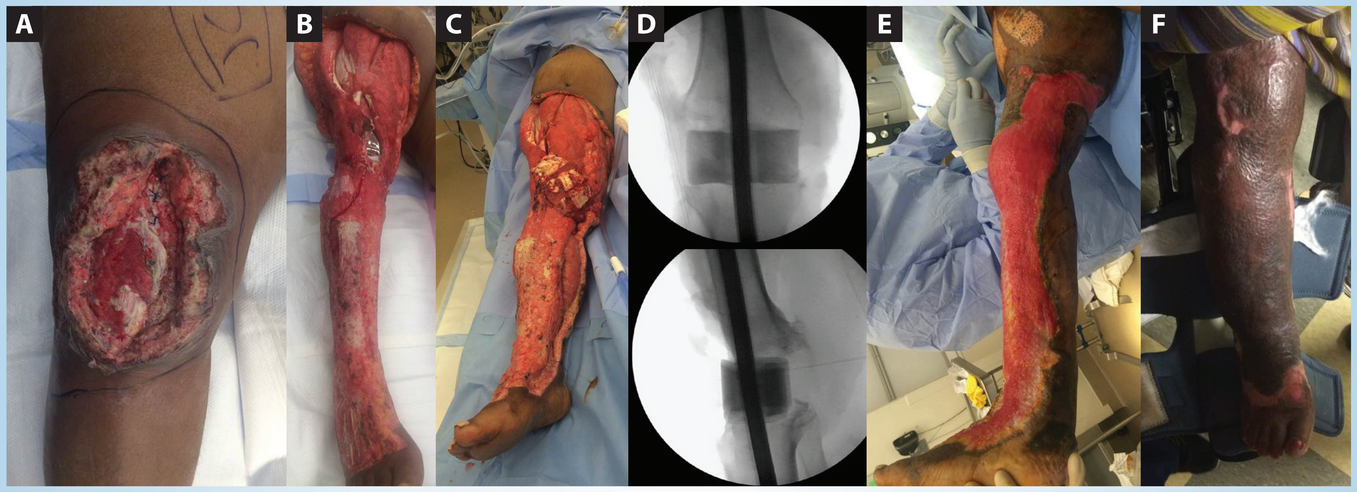

Figure 1. A, 67-year-old African American female with necrotizing fasciitis 15 days post primary total knee arthroplasty. **B**, Status post initial debridement. Immunological testing revealed IgM deficiency, increased IgE, and C4 deficiency. **C–E**, Patient underwent 17 surgeries including a temporary knee arthrodesis with antibiotic coated rod and spacer, gastrocnemius muscle transfer, and serial skin grafting. **F**, 7 months post initial total knee arthroplasty with complete skin growth.

Conclusion

- Maximizing the immunology system during an orthopedic infection is essential for successful management.
- Surgeons should consider primary immunodeficiency when treating patients with orthopedic infections.
- Host optimization is crucial to ensure proper eradication of infection.
- Patients with abnormal immunological values should be referred to an immunologist to monitor immune status and replenish IgG if indicated.
- This is the first orthopedic documentation to address the host immunological status during treatment.