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Perianal Manifestation of Cutaneous Sarcoidosis

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Background	A 53-year-old male patient with a history of pulmonary sarcoidosis presented with anal lesions. Examination under anesthesia and excisional biopsy confirmed the diagnosis of cutaneous sarcoidosis.
Summary	The patient initially noticed the anal lesions 2 to 3 years prior to presentation, experiencing symptoms of itching and occasional bleeding. Biopsies obtained during both colonoscopy and the exam under anesthesia demonstrated findings consistent with colonic and cutaneous sarcoidosis, respectively. Interestingly, the patient's history of pulmonary sarcoidosis was remote, with no other recent signs of systemic disease activity. Sarcoidosis, while known for its diverse cutaneous manifestations (commonly on the face, extremities, and trunk), rarely presents in the perianal region. This case underscores the importance of including sarcoidosis in the broad differential diagnosis of perianal lesions, particularly in patients with a known history of the disease.
Conclusion	Perianal sarcoidosis is a rare manifestation of cutaneous sarcoidosis. Here, we present a case of a 53-year-old male with a history of pulmonary sarcoidosis who developed perianal sarcoidosis. This case highlights the importance of considering a broad differential for lesions resembling condylomas. A thorough review of the patient's medical history is crucial, as it may reveal prior systemic diseases with potential cutaneous manifestations.
Key Words	cutaneous sarcoidosis; perianal lesion

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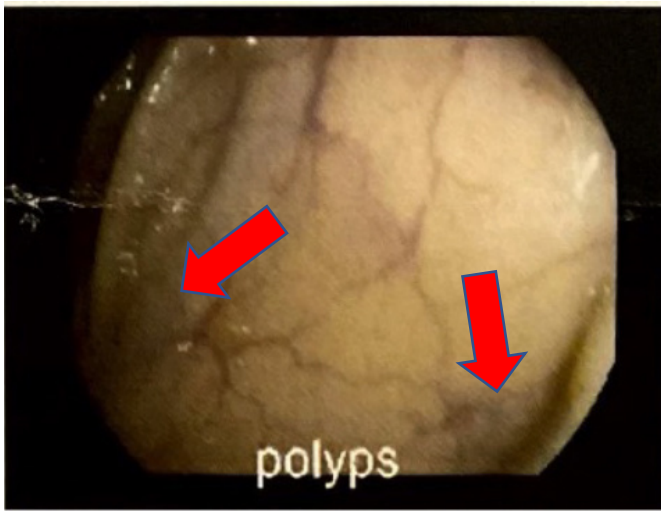
Case Description

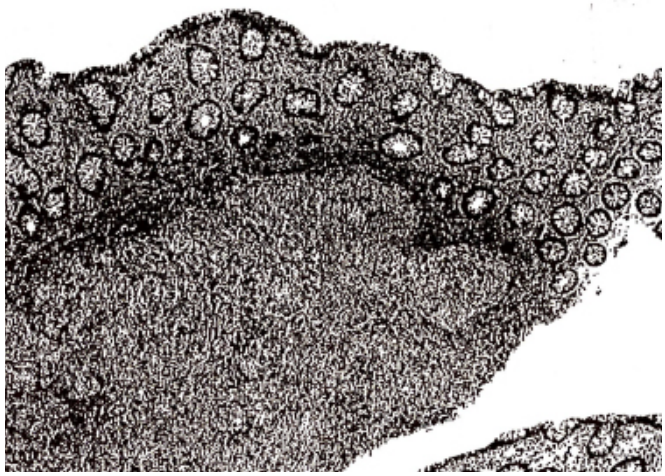
A 53-year-old male with a history of pulmonary sarcoidosis presented with a three-year history of pruritic perianal lesions. Relevant history included genital condylomas treated with cryotherapy two years prior, remote urethral chlamydia treated with antibiotics, and a negative HIV test two years prior. A routine colonoscopy six months before evaluation revealed multiple descending colon polyps (Figure 1), with biopsies demonstrating noncaseating granulomatosis consistent with colonic sarcoidosis (Figure 2). Anoscopic exam showed circumferential perianal thickening extending 2 cm from the anal verge, accompanied by ulceration. Due to poor tolerance of office-based anoscopy, the patient underwent an exam under anesthesia. External examination revealed thickened perianal skin with plaques and ulcerations, primarily anterior and posterior (Figure 3). Multiple biopsies were taken from ulcer edges and areas of perianal thickening.

Figure 1. Colonoscopic Images of Four Polyps in Descending Colon. Published with Permission



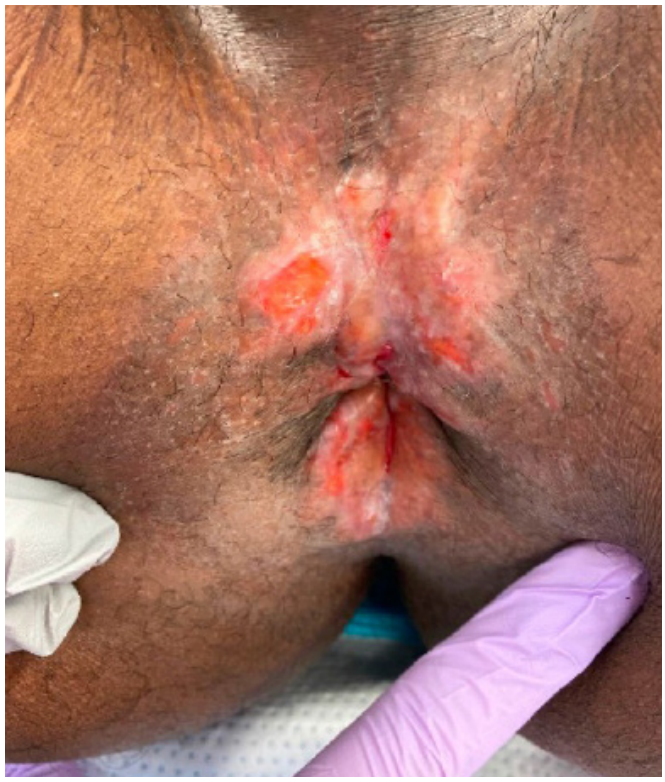
Sessile polyps of varying size in the descending colon, subsequently removed by polypectomy

Figure 2. Colonic Biopsy Demonstrating Non-necrotizing Granulomas. Published with Permission



Histologic sections of colonic mucosa and submucosa demonstrating extensive infiltration by confluent non-necrotizing granulomas. The extensive granulomatous inflammation is characteristic of sarcoidosis (AmeriPath GI Institute).

Figure 3. Cutaneous Sarcoidosis with Perianal Involvement. Published with Permission



Lesions observed during examination under anesthesia

Pathologic analysis demonstrated squamous mucosa with focal squamous epithelial hyperplasia, parakeratosis, and extensive granulomatous inflammation. Numerous noncaseating epithelioid granulomas in the stroma were consistent with sarcoidosis. Tests for infectious etiologies (HSV-1 and HSV-2, *Chlamydia trachomatis*, *Neisseria gonorrhoeae*, and Acid-Fast Bacilli) were negative. No evidence of dysplasia or malignancy was present. The patient was prescribed a topical steroid cream for perianal sarcoidosis.

At six-month follow-up, the patient reported limited pain at biopsy sites and tested positive for HSV IgG. Due to the synchronous discovery of colonic sarcoidosis during colonoscopy, a rheumatology referral was initiated. While the patient experiences infrequent perianal pruritis flares managed with topical steroids, preliminary pulmonary function tests indicate no current need for systemic treatment. The patient will continue symptom management of flares with topical agents.

Discussion

Sarcoidosis is a granulomatous disease primarily affecting the skin, lungs, and lymph nodes. Diagnosis relies on clinical and histopathologic findings, with the hallmark histologic feature being noncaseating granulomas. Cutaneous sarcoidosis manifests as papules, nodules, plaques, and infiltrating scars, often developing around the time of systemic diagnosis (or slightly preceding it) in approximately 80% of patients.[1] While elevated angiotensin-converting enzyme and sedimentation rate may support a sarcoidosis diagnosis, these laboratory findings alone are not definitive.[1]

Cutaneous sarcoid manifestations are often asymptomatic and diverse in presentation.[3] The most common are papular, nodular, and plaque lesions, differentiated by location, appearance, and tissue involvement. Papular sarcoid features 1 to 10 mm, non-scaly papules frequently found on the face near the eyelids and nasolabial folds.[2] Nodular sarcoidosis differs from papular by its involvement with dermal and subcutaneous tissue. Nodular sarcoidosis involves dermal and subcutaneous tissue and was found in roughly 36% of patients with cutaneous systemic disease in a 2008 study, highlighting its commonality.[4] Plaque sarcoidosis is one of the more difficult manifestations to identify and one that is often misdiagnosed. Plaque sarcoidosis poses diagnostic challenges due to its resemblance to conditions like Kaposi sarcoma, lichen planus, psoriasis, cutaneous T-cell lymphoma, and secondary syphilis.[5] Common non-pulmonary sites for sarcoidosis include the extremities, trunk, and buttocks. While the extremities, trunk, and buttocks are common sites, clinically significant gastrointestinal involvement is rare (<1% of systemic sarcoidosis cases).[6] Autopsies reveal it in 0% to 10% of such cases, with only three documented cases in medical literature highlighting the exceptional nature of this manifestation.[7,8]

When cutaneous manifestations raise suspicion of sarcoidosis in a patient without a prior diagnosis, a thorough evaluation for systemic involvement is warranted. Established guidelines include chest radiography, electrocardiogram, ophthalmic exam, comprehensive physical exam, detailed personal and medical history, and basic bloodwork (potentially including angiotensin-converting enzyme levels).[1] Treatment of cutaneous sarcoidosis depends on disease severity. For localized disease, recent recommendations favor intralesional or topical steroids, though further studies are needed to compare their efficacy.[9] In sensitive areas like the face and genitals, studies support the use of topical calcineurin inhibitors such as tacrolimus as an alternative to high-dose corticosteroids.[10]

Conclusion

While rare, cutaneous sarcoidosis can manifest in the perianal region. Our case, a 53-year-old male with a history of pulmonary sarcoidosis, highlights the importance of considering a broad differential diagnosis when presented with lesions resembling condylomas. A thorough review of a patient's medical history is crucial, as it may reveal prior systemic diseases with potential cutaneous manifestations.

Lessons Learned

Cutaneous lesions can signify underlying systemic disease, necessitating further evaluation and treatment. In patients with chronic conditions like sarcoidosis, routine screening and surveillance are essential for early detection and management of potential systemic manifestations.

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