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Rare Case Report: A Cutaneous Swelling Present as A Primary Manifestation of Sarcoidosis and Can be an Early Solved Mystery for Sarcoidosis

KEYWORDS

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ABSTRACT *Introduction: Sarcoidosis is a systemic disease that can involve almost any organ system. Infiltration with non-caseating granulomas is the hallmark of the disease.*
Case report: A 36-year-old woman presented with a 2- month history of a progressive, tumorous swelling in left index finger. Excision done and Final histology revealed numerous non-caseating epitheloid cell granulomas of the sarcoidosis.
Discussion: Hand sarcoidosis is observed in 0.2%. The functional consequences of hand sarcoidosis are quite different. Indolent and functionally irrelevant nodules are frequent.
Conclusion: Solitary soft tissue sarcoidosis may albeit the physician for an early diagnosis of systemic sarcoidosis.

INTRODUCTION:
Sarcoidosis is a systemic disease that can involve almost any organ system. Infiltration with non-caseating granulomas is the hallmark of the disease, and it may result in various clinical manifestations[1]. The underlying cause of sarcoidosis remains unknown. Myoskeletal involvement is uncommon (1-15%) and often asymptomatic, but it can manifest as granulomatous synovitis, acute and chronic arthritis, osseous cystic lesions, osteopathy or sclerosis. A primary manifestation of systemic sarcoidosis as an isolated tumour is extremely rare. Incision biopsy is needed to provide a diagnosis[3]. In case of cutaneous sarcoidosis of finger further investigation on systemic involvement have to be performed. We present the case of sarcoidosis of the finger.

CASE REPORT:
A 36-year-old woman presented with a 2- month history of a progressive, tumorous swelling in left index finger. There was no history of recent traumatisation or previous or current systemic disorder. A mature scar was the only residue of a minor superficial cut of her left index finger 30 years prior. The patient denied any bronchopulmonary symptoms such as dyspnoea or cough.

Physical examination revealed a painless tumour with solid consistency in the distal extensor aspect of left index finger. There were no signs of inflammation. Perfusion, mobility and sensibility of the finger were intact.

X-ray and ultrasound imaging revealed a soft tissue tumour on the middle phalanx of the left index finger without bone involvement or soft tissue calcification.

Intra-operatively we saw a nodular tumour with firm consistency and fat tissue penetration which macroscopically infiltrated the extensor tendon sheath without involvement of the bone. Excision was done.

Final histology revealed numerous non-caseating epitheloid cell granulomas of the sarcoidosis type with polynuclear giant cells, sparse lymphocytes and abundant collagenous fibres. There was no evidence for acid fast bacilli and all investigations were negative for mycobacteria. Intradermal tuberculin skin test was also negative.

The patient was referred to the department of internal medicine for further diagnostic investigation and therapy. Chest X-ray and CT scan was done and shows no evidence of involvement of lung parenchyma. Pulmonary function tests were normal. CD4/CD8 ratio was in the normal range. The initial angiotensin converting enzyme (ACE) level was normal (reference range 8–52 U/l). There was no indication for systemic anti-inflammatory treatment of the asymptomatic patient with normal pulmonary function tests and without evidence of further systemic involvement. Follow-up was performed three, six and twelve months later

On Further follow up at 3rd months CT scan chest revealed mediastinal and hilar lymph nodes. Angiotensin converting enzyme (ACE) level was only slightly increased (59 units per litre, reference range 8–52 U/l). Oral corticosteroid therapy with an initial daily dose of 40 mg prednisone was started. Hilar lymphadenopathy resolved After 12 month's therapy with a stepwise decrease of the corticosteroid dosage. ACE-levels decreased to 26 U/l. At present, 36 months after first presentation, a maintenance dosage of 7.5 mg prednisone per day is needed

DISCUSSION:
Reportedly, cutaneous manifestations in sarcoidosis occur in up to one third of patients who are known to suffer from systemic sarcoidosis. This is often classified as "specific" type with granulomatous reaction demonstrated in skin biopsy and alternatively "nonspecific" type is characterized mainly by reactive changes but no traces of granulomas. In about 33% of cases, skin lesions precede the systemic disease while in the remaining the skin manifestations appear simultaneously[2,3].

Sarcoidosis is a systemic disorder with primary involvement of the lungs and lymph nodes, which often has an asymptomatic course. In rare cases of advanced sarcoidosis, disease manifestations in bones, joints and tenosynovial tissues may occur[4]. Hand sarcoidosis is observed in 0.2%[3]

The symptoms are not specific for sarcoidosis. Differential diagnosis comprises rheumatism, gout, pseudogout, xanthoma, giant cell tumours, foreign body granulomas as well as syphilis or borrelia infections[5]. Tuberculosis and atypical mycobacterias, especially infection with mycobac-

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terium marinum (swimming pool granuloma) may have a similar macroscopic appearance. In TB however histology mostly shows caseating granulomas. Tuberculin skin testing is negative in patients with sarcoidosis, as they are investigations for mycobacteria. Another important differential diagnosis, which warrants histology, is malignancy[6].

With plaques and subcutaneous nodules affecting the face, particularly around the nose, cheeks and eyes, is termed lupus pernio. Lupus pernio is more common in patients of African descent; it is rare in Caucasians. Less commonly, sarcoidosis may manifest with extensive, deep subcutaneous nodules with oedema, particularly of the lower extremities; the term Darier-Roussy sarcoidosis was earlier used to describe this problem[4,5].

Granulomatous nodules often develop at sites of previous scars. Rarely, nodules in the skin may develop at sites of tattoos, body piercings or previously punctured sites of the skin or oral mucosa. Other reports document granulomas at sites of silica injections, although the nodules were probably due to sarcoid-like foreign body reaction[7]

The functional consequences of hand sarcoidosis are quite different. Indolent and functionally irrelevant nodules are frequent. However involvement of tendons and tendon sheaths may cause movement restrictions or ruptures of the tendons. Pain occurs when bones and joints are affected. Erosions, cystic bone lesions or pathologic bone fractures are rarely seen [6,7].

Our patient reported neither to have pain nor restriction of mobility and there was no osseous involvement.

An early diagnosis with adequate therapy seems important. There is no report on a role of medical therapy with steroids local or systemic for sarcoidosis of the hands in the literature. This may be partly due to the fact that most cases are reported from surgical Departments. In our opinion excision of tumour may enable immediate local control. Our case indicates that complete and long lasting local control can be reached by surgery, however no data exist comparing surgery with local injections or systemic applications of steroids in this clinical situation. Therefore patients presenting with disease confined to skin alone, should be followed up regularly for the probable risk of developing systemic manifestations at a later date[8]. We asked her to attend chest, eye and skin clinics every three months.

CONCLUSION:

Every suspicious or progressive swelling needs a complete work-up with consideration of differential diagnoses and radiological examinations. In case of cutaneous or subcutaneous swelling possible differential diagnosis of sarcoidosis should keep in mind. For histological proof and exclusion of a malignant process a biopsy is to be performed. The biopsy offers the right diagnosis and thus leads to an appropriate, reasonable and successful treatment of sarcoidosis. In most cases satisfactory clinical results can be achieved by systemic or topic application of corticosteroids. Sometimes a long term therapy is needed, even if the local manifestation seems to be gone. Further local surgical procedures are not indicated. Solitary soft tissue sarcoidosis may albeit the physician for an early diagnosis of systemic sarcoidosis.

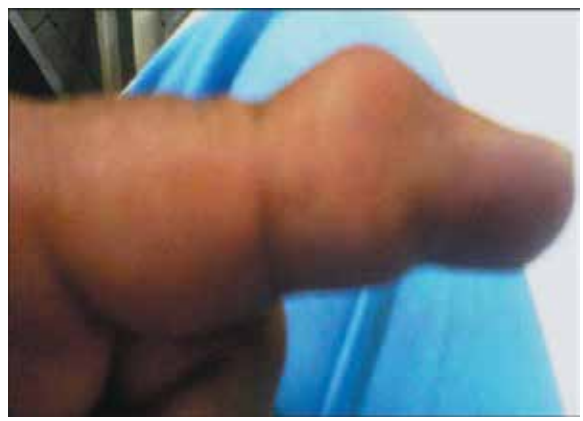

Fig1: showing tumour on finger.

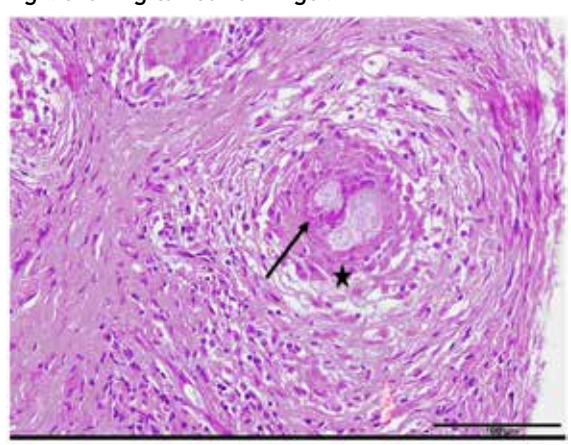

Fig2: HPR showing a granulomatous lesion (sarcoidosis)

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