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An isolated hand tumour as primary manifestation of sarcoidosis

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Abstract. *Background:* Sarcoidosis is a systemic disorder with unknown etiology, characterized by non-caseating granulomas in numerous organs and tissues. In 90% of patients lung and lymph nodes are involved. The incidence of sarcoidal granulomas in the upper extremities is low. Here we present the case of a primary hand manifestation of sarcoidosis without clinical systemic involvement. *Objectives:* A young woman presented with a painful swelling in her right hand. There were no signs of inflammation. Normal perfusion, mobility and sensibility were found. Magnetic resonance imaging (MRI) revealed a tumour infiltrating the muscles and flexor tendons of the third digit around the metacarpal bone and with pathological signal enhancement after administration of contrast medium. *Results:* Intraoperatively, nodular masses and fat tissue were seen. Histological examination after radical tumour resection showed sarcoidal granulomas. Postoperative staging diagnostics with computed tomography (CT) demonstrated multiple thoracic lymph node swellings in the mediastinum and bilateral hili. Follow-up after one year we saw normal scars in the palmar hand. There was no sign of local recurrence. The pulmological care is still going on. *Conclusions:* Sarcoidosis is a rare, often asymptomatic disease. Patients present with dyspnoe and cough caused by the inflammation of the lung. The first clinical manifestation of sarcoidosis as a tumor in the palmar hand is unusual. Extrapulmonary systemic or progressive sarcoidosis is regarded as an indication for therapy with glucocorticosteroids. This case demonstrates that surgical excision enabled complete local cure without necessity of systemic and/or local treatment with steroids. *(Sarcoidosis Vasc Diffuse Lung Dis 2011; 28: 72-74)*

Key words: tumors of the hand, sarcoidosis, noncaseating granuloma

Introduction

Sarcoidosis is a systemic granulomatous disease of unknown aetiology. It nearly always affects the intrathoracic lymph nodes (90%) and the lungs (70%) (3). Myoskeletal involvement is uncommon (1-15%) and often asymptomatic, but it can manifest as granulomatous synoviitis, acute and chronic arthritis, osseous cystic lesions, osteopathy or sclerosis. A primary manifestation of systemic sarcoidosis as an isolated tumour is extremely rare. It requires differentiation from malignancy and interdisciplinary treatment decision.

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Case report

A 36-year-old woman presented with a 2-month history of a progressive, tumorous swelling in

the palm of her right hand. 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 right palm 30 years prior. There had also been no occupational exposure to beryllium and the patient denied any bronchopulmonary symptoms such as dyspnoea or cough.

Physical examination revealed a painless tumour with solid consistency in the distal hand palmar to the second and third metacarpal bone. There were no signs of inflammation. Perfusion, mobility and sensibility of the hand were intact.

X-ray, ultrasound imaging and MRI showed an irregular (4 x 3 x 1.5 cm) soft tissue tumour on the proximal phalanx of the middle finger infiltrating the musculi lumbricales and interossei dorsales II and III and the flexor tendons of the third digit around the metacarpal bone without bone involvement or soft tissue calcification and without liquid components. MRI also showed infiltration of the palmar fascia, subcutaneous tissue and the cutis (Fig. 1).

Intraoperatively we saw a nodular tumour with firm consistency and fat tissue penetration, which macroscopically infiltrated the musculi lumbricalis and the flexor tendon sheath without involvement of the bones (Fig. 2). Intraoperative frozen section analysis revealed noncaseating granulomatous inflammation. Radical tumour resection was performed. Final histology revealed numerous noncaseating epitheloid cell granulomas of the sarcoidosis type with polynuclear giant cells, sparse lymphocytes and abundant collagenous fibres. There was no

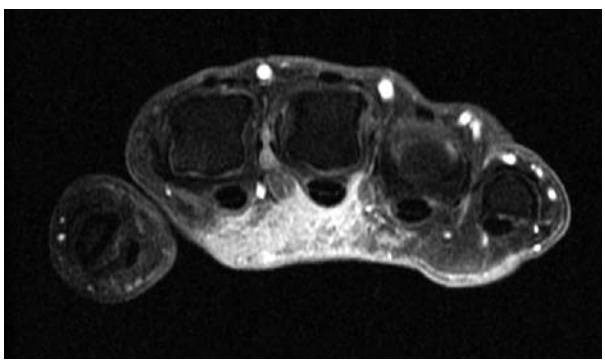

Fig. 1. Pre-operative MRI of the hand tumor infiltrating the musculi lumbricalis and musculi interossei dorsalis II and III and the flexor tendons of the third digit around the metacarpal bone

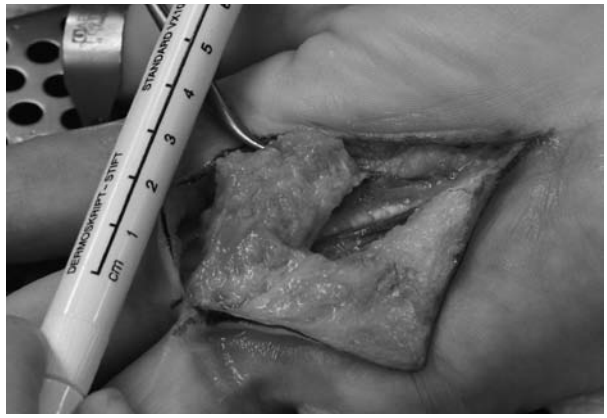

Fig. 2. Intra-operative findings nodular, compact tumor with fat tissue penetrated

evidence for acid fast bacilli and all investigations were negative for mycobacteria. Intradermal tuberculin skin test was also negative.

Chest X-ray and CT revealed mediastinal and bihilar lymph nodes without evidence for involvement of lung parenchyma. Pulmonary function tests were normal.

Fiberoptic bronchoscopy was performed. Bronchoalveolar lavage (BAL) fluid showed slightly elevated levels of lymphocytes (24%) and neutrophil granulocytes (3%), whilst the percentage of eosinophils was distinctly increased up to 13%. CD4/CD8 ratio was in the normal range (1.2). Biopsies of bronchial mucosa revealed slight signs of chronic bronchitis without evidence of sarcoidosis.

There was no indication for systemic antiinflammatory 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. It showed spontaneous regression of the mediastinal and hilar lymph node masses. MRI of the hand showed normal metacarpal and wrist bones and phalanges without signs of osteopathies. Tendons and palmar muscles appeared normal as well. There was no local recurrence.

Discussion

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. Hand sarcoidosis is observed in 0.2% (1), however the initial manifestation of sarcoidosis in the hand is described casuistically only (4, 6, 9). 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 (1). Tuberculosis and atypical mycobacterias, especially infection with mycobacterium 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 are investigations for mycobacteria. There is therefore convincing proof of sarcoidosis in our patient. Another important differential diagnosis, which warrants histology, is malignancy.

Papadavid et al (8) described a patient with a swollen and erythematous right forearm and hand in whom sarcoidosis was mimicking cellulitis. Contrarily in our case no signs of inflammation or infection were clinically apparent. The functional consequences of hand sarcoidosis are quite different. Indolent and functionally irrelevant nodules are frequent (5, 10). However involvement of tendons and tendon sheats may cause movement restrictions or ruptures of the tendons (5, 10, 11). Pain occurs when bones and joints are affected. Erosions, cystic bone lesions or pathologic bone fractures are rarely seen (2, 7). Our patient reported neither pain or restriction of mobility and there was no osseous involvement.

An early diagnosis with adequate therapy seems important. There is no report on a sole medical therapy with steroids – local or systemic - of 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 a radical resection with reconstruction of functional structures may enable immediate local control. Our case indicates that complete and long lasting local control can be reached by surgery alone without corticosteroids, however no data exist comparing surgery with local injections or systemic applications of steroids in this clinical situation

In our case the systemic, though asymptomatic disease with involvement of the mediastinal and hilar lymph nodes was detected after cognition of the histological diagnosis of sarcoidosis of the hand. The clinical and roentgenological appearance was quite typical and the BAL-pattern compatible with sarcoidosis. Treatment of pulmonary sarcoidosis is indicated in cases of progressive functional impairment or involvement of other organs like the heart, central nervous system or eyes as well as hypercalciemia. Therefore in our patient medical treatment with steroids was not justified. Follow-up showed spontaneous remission of the thoracic lymph node masses. It is impossible to conclude whether the local cure influenced the favourable course of the systemic disease.

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