# CASE REPORT

# Shoulder Mycetoma with 'Dot-in-Circle' Sign

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### ABSTRACT

Mycetoma is a chronic granulomatous disease that can be debilitating, increasing the healthcare burden of patients and their families. It is caused by fungal or filamentous bacterial infection. Mycetoma usually develops in farmers after a traumatic inoculation of the organism into the subcutaneous tissue, muscle or tendon. The foot is the most commonly affected site. The leg and upper extremities including the neck, chest and shoulder can also be involved with decreasing incidence. The 'dot-in-circle' sign is a classical feature on magnetic resonance imaging (MRI). We presented a case of shoulder mycetoma, a rare site of infection that showed the 'dot-in-circle' sign on MRI. Microscopically, it exhibited multiple granulomatous inflammations surrounded by abundant foamy macrophages. The fungal elements were highlighted using Gomori methenamine silver stain.

Keywords: Shoulder mass, Mycetoma, Dot-in-circle sign

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#### **INTRODUCTION**

Mycetoma is an infection that involves subcutaneous tissues affecting all parts of the body, predominantly foot. It can also progress to deeper structures. The worldwide burden of this disease is still unclear, but it is primarily endemic in subtropical and tropical regions. Until now, there is no definite global prevalence available for mycetoma due to limited data. The highest prevalence was noted in Sudan with 1.8 cases per 100,000 population. Most cases were reported in areas with hot and dry climates, and there was no reported person-to-person disease transmission (1). In a region where mycetoma rarely occurs, the attending physicians often misdiagnosed the disease. Instead, it is erroneously diagnosed as neoplasm or other forms of granulomatous condition (2). While no age preference has been reported, it rarely affects children and usually develops in men aged 20 to 40 years (1).

To date, mycetoma predominantly affects men compared to women in both children and adults with a

ratio of 1.6–6.6:1. This can be due to the involvement of a majority of men in the agricultural field (1). Clinically, eumycetoma and actinomycetoma exhibit similar presentations. Nevertheless, the latter is shown to be more aggressive with early bone invasion and causes more damage. As a result of the slow progression of the disease and its painless symptom in most cases, the disease usually presents in adults with voluminous tissue and bone damage after many years of getting infected (3).

#### **CASE REPORT**

A 65-year-old diabetic female farmer presented to our hospital with a complaint of right shoulder swelling for one year duration. Its initial size was similar to a 50cent coin, which later increased to about the size of a softball. It was associated with minimal pain, without affecting shoulder movements. She had a history of loss of appetite and weight for the past 3 months. However, she denied any history of trauma, discharging sinuses, pain, fever or presence of any swelling elsewhere on the body. There was no lymphadenopathy detected. She had no family history of malignancy.

Physical examination showed an 8 x 10 cm, non-tender fluctuant/soft mass on the right shoulder. The mass was

mobile and not attached to the underlying muscle or skin. Dilated superficial veins were observed over the mass with a small superficial ulcer and minimal contact bleeding. Routine full blood count was normal. Plain radiography (Fig. 1) showed a homogenous appearance of a soft tissue lesion at the lateral region of the right shoulder. No area of lucency or calcification was noted. The adjacent right humeral bone was normal. The findings from plain radiograph were consistent with soft tissue lesion, suggestive of lipoma. In view of its huge size, liposarcoma was established as a differential diagnosis.



Figure 1: Plain radiography of the right shoulder showing homogenous soft tissue mass over the lateral region of the right shoulder (\*). No calcification was noted within the mass. Adjacent humeral bone appeared normal.

Magnetic resonance imaging (MRI) showed a well encapsulated mass at the subcutaneous region of the right shoulder, which did not involve the adjacent deltoid muscle. The mass appeared heterogeneous in intensity, and there was an area that suppressed on fat suppression sequence, suggestive of an adipocytic component. The mass showed moderate heterogeneous enhancement with an area of hyperintense foci on T1weighted (Fig. 2a), T2-weighted (Fig. 2b) and short tau inversion recovery (STIR) images exhibiting the 'dot-incircle' signs, which were more prominent on the T1weighted fat-saturated gadolinium-enhanced images (Fig. 2c). Based on the 'dot-in-circle' sign in MRI, mycetoma was established as a provisional diagnosis. The differential diagnosis was liposarcoma as there was an area that suppressed on fat sequence.

The patient was subjected to trucut biopsy, and the result was reported as granulomatous inflammation due to fungal infection. Histologically, the mass composed of a collection of epithelioid histiocytes and some multinucleated giant cells surrounding the necrotic area with peripheral rimming of lymphoplasmacytic cells (Fig. 3a). Gomori methenamine silver (GMS) stain



Figure 2: Coronal view of MRI conducted on the patient, which showed a well-encapsulated lesion (arrows) in the subcutaneous region of the lateral side of the right shoulder. There were multiple areas of well-defined hyperintensities of varying sizes on T1-weighted (2a) and T2-weighted (2b) images exhibiting 'dot-in-circle' signs (\*) that were more prominent on the T1-weighted post-gadolinium image (2c).



Figure 3: The granulomatous lesion exhibited collection of epithelioid histiocytes mixed with multinucleated giant cells surrounded by small mature lymphocytes (haematoxylin and eosin staining [H&E] 200x) (3a). GMS stain highlighted the cell wall of the 45°-branching fungal hyphae (GMS staining 400x) (3b).

highlighted the cell walls of the 45° branching fungal hyphae (Fig. 3b). GMS is a special stain to detect fungi.

The patient initially received intravenous voriconazole 300mg for 2 doses then 200mg twice daily for 2 weeks, followed by surgical excision of the mass. The excised specimen (Fig. 4) showed a fairly circumscribed lesion with yellowish solid cut surface with multiple dot-like appearances. The surrounding tissue was uninvolved. There was no area of necrosis or area haemorrhage observed. The histopathological features from the excised specimen showed a similar appearance with biopsy. Fungal culture was not performed as the lesion was solid and there was no discharge noted from it. The final diagnosis of granulomatous inflammation due to fungal infection (mycetoma) was made after the histopathological examination. The patient was discharged well and provided with oral itraconazole 200 mg once a day for 6 weeks. She was given an appointment for a review but defaulted on the followup.

# DISCUSSION

Mycetoma is an infective disease that involved barefooted farmers, commonly in tropical and subtropical regions, especially in rural agricultural communities. The initial



Figure 4: Excised shoulder mass exhibits a fairly circumscribed lesion with yellowish solid cut surface with multiple dot-like appearance.

trauma site provides an entry route for the organism to enter the body. If left untreated, the infection can spread to adjacent structures, which can lead to destruction and significant deformities. Clinical suspicion of eumycetoma can be made based on a typical history of trauma, being barefooted during gardening and swelling with discharging sinuses. A classic triad of painless subcutaneous tissue swelling with sinus tract and grain extrusion is suggestive of mycetoma (1). Eumycetoma caused by fungi usually requires a combination of surgical and medical therapy like in this patient. Fifty-six different microorganisms, such as bacteria and fungi, are the causative agents of mycetoma. Some organisms are found in the soil or the gut and casts of earthworms. The etiological agents implicated are Curvularia geniculata, Curvularia lunata, Exophiala jeanselmei, Phialophora verrucosa, Pseudallescheria boydii, Acremonium spp., Aspergillus spp. and some Fusarium spp. Most of them can be cultured on potato dextrose agar. The characteristic features of the colonies and hyphal and conidial appearances can be observed microscopically by performing the lactophenol cotton blue test.

Without a typical history, mycetoma may be difficult to diagnose. Histopathological examination will give the definitive diagnosis of mycetoma in this type of cases. Moreover, mycetoma can sometimes be misdiagnosed as neoplasm, chronic infection or neuropathic foot. The World Health Organization in 2014 had included mycetoma in the list of neglected diseases and it warrants attention because it is preventable. MRI with 'dot-in-circle' sign will help in diagnosis.

Radiologically, soft tissue swelling is the most common finding in plain radiography of mycetoma. Bone sclerosis, cavitation formation and periosteal reaction are the associated features. The best choice for soft tissue assessment is MRI (3). Czechowski et al. reported the MRI appearance of mycetoma as small low-signal intensity lesions on T1-weighted and T2-weighted MRI in 16 of 20 patients (3). These changes were suggested to be due to susceptibility of the metabolic products of the fungal grains. Since then, there were a few cases reported on MRI as showing the 'dot-in-circle' sign in cases of foot and of thigh mycetoma (2). The 'dot-incircle' sign could be observed in T2-weighted, STIR and T1-weighted fat-saturated gadolinium-enhanced images as a highly specific sign for mycetoma. It was hypothesized that the 'dot-in-circle' sign was due to the presence of central fungal grain being surrounded by inflammatory granuloma. The diffusion-weighted imaging sequence could be used to detect any sinus tract occurrence.

However, it is still not possible to differentiate between actinomycetoma and eumycetoma based on MRI examination alone because of the overlapping features.

Although the 'dot-in-circle' sign is a typical sign for myocetoma, it may also can be seen in soft tissue hemangioma, thus it's important to correlate clinically. The synovial fluid in rheumatoid arthritis and tuberculosis infection can also shows foci of low-signal intensity, mimicking the 'dots' (4). The 'dot-in-circle' sign also can be seen in ultrasound examination of myecetoma patient and is comparable to the MRI examination. The fungal grain will produce hyperechoic are giving rise the dot appearance and is surrounded by a circle of hypoechoic tissue (4).

In Malaysia, there are no data on the exact prevalence of mycetoma. However, there were a few reported cases of mycetoma, which mainly involved the lower extremities with Phialophora jeanselmei, Madurella mycetomatis and Streptomyces somaliensis detected as the primary causes (5). Mycetoma typically involved the lower extremities, particularly the foot. Shoulder involvement in mycetoma is rare, with some authors reporting an incidence of < 4%. To the best of our knowledge, this is the first case of shoulder mycetoma showing the 'dot-in-circle' sign being reported in Malaysia. This patient who worked as a farmer probably had a cut at the right shoulder earlier on before she had swelling.

#### CONCLUSION

The 'dot-in-circle' sign in MRI is quite specific for mycetoma and can help in the diagnosis of patients who had atypical presentations and unique locations for soft tissue mycetoma. However, HPE and GMS stain are two critical diagnostic techniques that must be incorporated to characterize the histological features of fungi-infected tissues. Nevertheless, fungus species identification using the culture test is a gold standard for diagnosis.

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