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Surgical Treatment of Extensive Tumoral Calcinosis Associated with Systemic Sclerosis

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Extensive tumoral calcinosis affecting a large joint is uncommon in patients with systemic sclerosis. We report the case of a 52-year-old female patient referred for a growing calcified mass in the shoulder. She was diagnosed with interstitial lung disease and progressive systemic sclerosis. Although the pain and disability associated with the affected joint was not severe, the patient underwent surgical excision because the mass continued to grow and was likely to produce shoulder dysfunction and skin ulceration. The patient appeared well 10 months after surgery with no signs of recurrence. This report highlights the timing and indication of surgical excision in similar cases.

Key words: 1. Tumor, benign
2. Calcinosis
3. Scleroderma, systemic
4. Surgery
5. Quality of life

CASE REPORT

A 52-year-old woman was referred to our department with a growing calcified mass in the left shoulder. She had been previously diagnosed with interstitial lung disease and progressive systemic sclerosis eight and five years prior, respectively. Four years before admission, chest computed tomography first revealed a small calcified lesion in the subscapular area (Fig. 1A). During regular follow-ups, the calcified lesion increased in size, eventually becoming a large tumor-like mass (Fig. 1B).

At presentation, the patient exhibited slightly limited motion in the affected joint and complained of mild pain and tenderness when leaning against a wall. The patient had no family history of tumoral calcinosis and no history of trauma, with the exception of a previous thoracoscopic lung biopsy of the right lower lobe. On physical examination, no skin lesions were noted on the body surface, but the mass was palpable on the neck and back. Laboratory test results revealed that serum calcium, phosphorus, and vitamin D values were within normal limits. Autoantibody test results were positive for antinuclear antibodies and negative for anticentromere antibodies. X-ray imaging of the chest revealed a large, lobulated, and trapezoidal calcific plaque extending from the tip of the left scapula to the lower neck (Fig. 1C).

When the patient was referred to us, she was being treated with a steroid (methylprednisolone, 2 mg) and an immunosuppressive drug (azathioprine, 100 mg) for interstitial lung disease. However, the patient received no specific therapy to control the progression of the calcification in the

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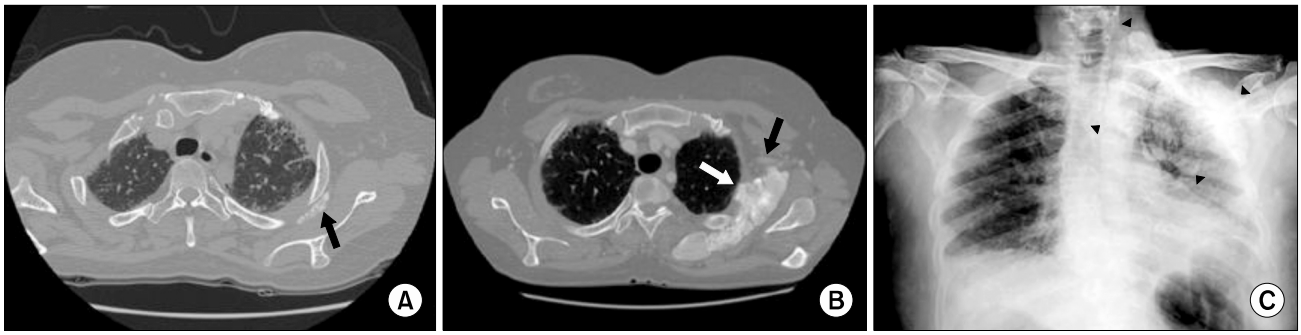

Fig. 1. (A) An initial chest computed tomography (CT) scan revealed a small calcified lesion (black arrow) on the subscapular area. (B) A chest CT scan obtained before surgery demonstrated that the calcified lesion had increased in size to a large tumor-like mass. The mass almost penetrated the pleural cavity (white arrow) and nearly compressed an axillary artery (black arrow). (C) A chest X-ray revealed a huge lobulated and trapezoidal calcific plaque (an arrow at each corner) extending from the tip of the left scapula to the lower neck.

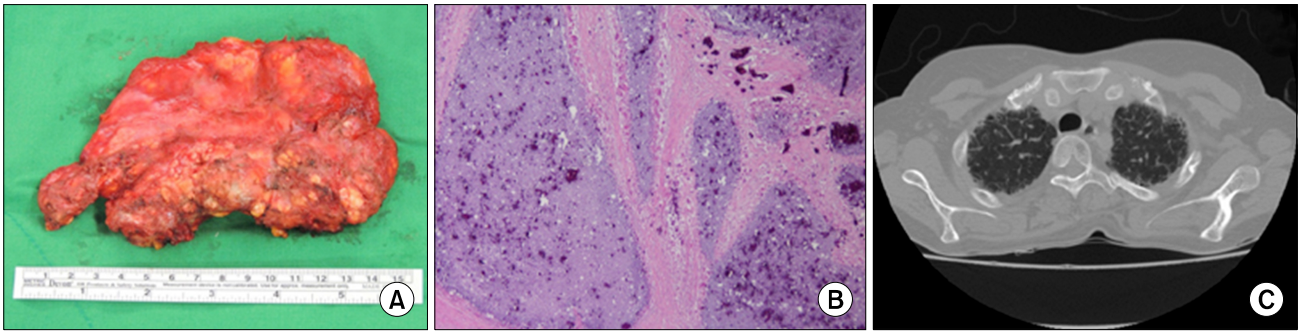

Fig. 2. (A) Macroscopic examination revealed a yellowish 14×13×3.5-cm mass containing multinodular fibrosis and marked necrosis. (B) Microscopic findings indicated non-specific calcification deposits (H&E, ×400). (C) A chest computed tomography scan performed 10 months after surgery revealed no recurrence of the calcification.

shoulder. Although the pain and disability associated with the affected joint were not severe, we surgically excised the chest wall mass because the mass continued to grow for several years, showing a tendency for more rapid growth in the years immediately before the decision to perform a surgical excision. This was particularly problematic because the patient was completely dependent on a wheelchair due to polio-induced paralysis, and therefore, the preservation of arm function was essential for maintaining her quality of life. Moreover, we were concerned that further growth of the tumor was likely to produce skin ulceration or compression of the brachial plexus and axillary vessels, which could potentially lead to psychological trauma to the patient and the ultimate loss of shoulder function.

In the operating room, the patient was positioned in a semi-prone position, and a skin incision approximately 15 cm in length was made along the medial border of the left scapula. The mass was located between the rib cage and the scapula, and was tightly attached to the first and second intercostal muscles. The mass was carefully dissected without damaging the neurovascular structures and completely excised (Fig. 2A). Pathological examination of the resected tumor revealed calcifications consistent with tumoral calcinosis (Fig. 2B).

The patient was discharged on the sixth postoperative day without any complications. In outpatient follow-up appointments, she appeared well, and no recurrence was observed on a computed tomography scan 10 months after the surgery (Fig. 2C).

DISCUSSION

Calcinosis, which refers to the deposition of calcium phosphate crystals in the soft tissues, is a reasonably frequent condition in patients with rheumatic disease or CREST (calcinosis, Raynaud's phenomenon, esophageal motility disorders, sclerodactyly, telangiectasia) syndrome [1]. The hands and fingers are affected in approximately 80% of patients with systemic sclerosis who experience calcinosis. The superficial soft tissues overlying bony prominences are most commonly involved; however, periarticular, intra-articular, or muscular involvement is also possible [2]. The calcinosis in systemic sclerosis is typically small and limited, although less common cases involving generalized or tumor-like calcifications have recently been reported [3-5].

The term tumoral calcinosis was originally used to refer to a rare hereditary disease involving phosphate metabolic dysfunction with a pattern of autosomal recessive transmission. This disease is characterized by multilobulated, densely calcified masses that are generally located on the extensor surfaces of large joints [6]. However, tumoral calcinosis gradually came to include huge mass-like calcifications that are secondary to various medical conditions such as renal failure, connective tissue disease, and hormonal imbalances.

Although chronic tissue inflammation and macrophage activation causing tissue devitalization seem to play an important role in the pathogenesis of calcinosis in rheumatic disease, the precise mechanism has not been fully elucidated [1]. Therefore, no standard treatment to prevent or eliminate calcinosis has been established. A few pharmacological agents such as warfarin, colchicine, diltiazem, and minocycline are occasionally beneficial in specific clinical circumstances; however, their success rates are variable, and no large series of controlled studies have investigated the effects of these drugs [1].

In general, surgical excision may be required when the lesion is large and causes severe nerve pain, deformity, disability, or ulceration of the affected joints. Carbon dioxide laser therapy can be an alternative treatment for small and superficial lesions. The present case has some limitations in terms of its ability to provide general guidance for the surgical treatment of tumoral calcinosis. First, attention was initially paid to the dominant pulmonary pathology, whereas the growing calcification in the shoulder was ignored as a typical epiphenomenon of systemic sclerosis. Second, medical treatment was thus not utilized to treat the early stages of the calcified mass. Finally, our patient was paraplegic, stemming from a prior polio infection, and preservation of the function of the affected arm was critical for the patient's quality of life.

Despite these limitations, it is imperative to treat tumoral calcinosis before the mass becomes ulcerative and disabling. Moreover, no pharmacological agent to date has demonstrated proven efficacy in the reduction or elimination of very large calcified masses, such as the mass observed in the present case. In light of the specific details of our case, we determined that a delay in surgery to allow for a possible response to medical treatment was potentially more risky than immediate surgical excision. In a previously reported case, a patient who did not undergo surgery eventually lost the functionality of his hand and the ability to engage in his occupation [4]. In another case, a patient presented with a huge ulcerated mass that had not been treated for several years; the painful mass limited his daily life and even resulted in depression [7]. When determining the optimal timing for surgery, it is important to consider the size of the calcified mass, the time lapse between the onset of calcification and the start of treatment, the patient's symptoms and general condition, and the presence of impending complications. Although there is a risk of recurrence and further calcification due to surgical trauma, it may be beneficial to perform surgery before the condition progresses to become a very large, painful, disabling, and ulcerated mass.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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