A CASE OF CALCINOSIS CIRCUMSCRIPTA

BY

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Calcinosis, or chalk gout as it is commonly named owing to the tophus-like swellings it presents, is a slowly progressing condition of unknown aetiology and pathogenesis. Two types are described, *circumscripta* and *universalis*. The female sex is the more commonly affected. The *circumscripta* variety usually occurs in those in middle or late life.

Discussion

Atkinson and Parkes Weber (1938) from whose paper most of the references given below are quoted, state, in a complete survey of the condition, that the first case of calcinosis was described in 1878 by H. Weber, and that Leissier in 1877 recorded the case of a woman aged 21 years who had had since childhood hard nodules in the skin and subcutaneous tissues: there was a discharge of chalky particles, and he believed his case to be one of gout associated with scleroderma. In 1844 Virchow described metastatic calcification in nephritis. In 1899 Derville published a case, and in the same year Durett suggested a familial incidence, but this was not proved. In 1900 Profichet wrote a thesis on calcinosis, with a résumé of all cases published up to date. It was afterwards called by foreign writers the syndrome of Profichet. Riche reported a sulphuric-acid worker who formed nodules of calcium carbonate and phosphate. In 1902 Hutchinson reported a case of a woman aged 58 years who probably suffered from scleroderma; the fingers felt wooden, and calcareous plaques were found in the deep layers of the skin of the forearms. This seemed to be a case of scleroderma with calcinosis, and was the first published in England.

Lehrenbecker in 1927 described a patient who had formerly undergone bilateral sympathectomy for Raynaud's disease and in whom calcification later supervened. The author wondered whether the operation had been the exciting cause of the calcinosis.

Steinitz (1930) reported a case in a woman aged 82, the oldest patient on record. Brauer believed ovarian function was diminished and that this fact had something to do with calcinosis. He also believed that circulatory disturbances might have favoured a chalky deposition. Wilson (1935) thought the deposits of calcium phosphate found in the fingers of one of his cases might have been due to an endogenous hypervitaminosis D connected with an unusually high blood cholesterol, superimposed on old Raynaud's disease which had remitted. The same authors state that sclerodactylia and scleroderma are frequently associated, the joints remaining free. They state that calcinosis *universalis* may be fatal.

Comroe (1944) states that the *circumscripta* variety is usually restricted in

distribution to the articular region of the phalanges and the extensor aspects of the knees and elbows. Calcium metabolism reveals no abnormality, and no demineralization of bone or arthritic changes are seen.

Pedersen (1943) says that the French call the condition "concretions calcaires des atrophies cutanées". He describes a woman at the menopause who had typical calcinosis circumscripta with a tendency to blue and cold hands. He excludes correlation between calcinosis and changes in the endocrine system, as in three cases post-mortem changes showed no abnormality in the latter system. He also thinks that the marked correlation between calcinosis and acrocyanotic conditions and scleroderma suggests some abnormality in the blood stream.

Pedersen (1943) gives the chemical composition of calcium deposits from a patient with calcinosis. There were moist brown tough lumps with a hard central core. The tough part was muscle fibre, the remainder, stony core. Analysis showed the presence of calcium, phosphate, a little carbonate, and a faint trace of magnesium, but no oxalate or uric acid. He concludes that the core was very similar to bone. Wigley and Hunter (1945) describe a case of calcinosis in chronic nephritis with secondary hyperparathyroidism. The patient died, and necropsy revealed chronic nephritis with shrinkage of the kidneys. There was hyperplasia of all four parathyroid bodies.

Of treatment, Comroe (1944) states that hemithyroidectomy and parathyroidectomy have been tried with little result. Pedersen (1943) states that one case responded to rest, colchicum, cinchophen, and mud packs. Atkinson and Weber (1938) state that drugs, physiotherapy, dietetic, and glandular and surgical treatment have been tried with no certain success. Hunter experimented with fibrolysin, Hoffman with hydrochloric acid, Weissenbach used thiosinamine, iodide, and thyroid gland. Chopra gave sodium phosphate for long periods, finding that the formation of nodules was thus prevented. Thyroid and ovarian extract were of no benefit when tried by Weil, Weissmann-Netter, and Brauer, though Werley claimed improvement from thyroid extract. Chopra found that anterior pituitary gland with small doses of thyroid reduced the size of the nodules. The use of x rays removed the nodes in a case described by Borak. Drucker thought radium useless, but Ducasse found that the nodules became liquefied with this treatment. Ramsdel performed unilateral thyroidectomy in one case. The parathyroids appeared normal, and great improvement followed; 50% of the nodules disappeared, and the crippled girl became active and resumed her duties. Brody and Bellin considered that the benefit of parathyroidectomy lay in its causing less saturation of the blood-serum with calcium, and in a tendency for the deposits to be decreased. Sheldon at the Royal Society of Medicine on March 25, 1938 (Atkinson and Weber, 1938), showed a child from whom the calcareous deposits had disappeared without any special treatment since the case was originally demonstrated in 1936.

The following case is reported owing to the widespread calcinosis and its response to local surgical treatment.

Case History

A man aged 40 years was first seen at the British Red Cross Rheumatism Clinic in December, 1942. He gave a history of the presence of small hard swellings for the past twenty to twenty-five years, the first one being on the left shin; both knees and elbows next became affected, and more recently firm nodules had developed on the left hand. If injured, the nodules became painful, whilst at other times they were inflamed and swollen. The patient's chief complaint was of the unsightly appearance of the nodules. There was a history of the nodules breaking down and discharging: they did not reappear at these sites.

He had no other symptoms, and there were no symptoms of circulatory disturbances. There was no family history of deposits. An elbow nodule had been excised, but a swelling recurred some three years after operation at the same site. His occupation had always been sedentary.

EXAMINATION.—Nothing abnormal was found in any system. There were multiple small nodules around the right elbow, and a larger firm area over the triceps muscle on the right arm and to a lesser extent over the left biceps muscle (Fig. 2, A). The nodules were marked over the left wrist and elbow. They were also present between the left tibia and fibula. Evidence of old ulceration and discharge was seen at this site. The patient showed no response to colchicum therapy.

INVESTIGATIONS.—At this stage the blood sedimentation rate (Westergren) was 7 mm. in the first hour, and 14 mm. in the second. Blood uric acid was 3 mg. per 100 c.cm. of blood; haemoglobin 89%; red blood cells 5,500,000 per c.mm. and white blood cells 7,100; the colour index 0.81%; and the blood calcium 10 mg. per 100 c.cm. A radiograph of the legs showed multiple disseminated calcinosis.

The patient's condition remained much the same until he reported again on July 4, 1946. He then complained of an increase in the size of the swellings, especially of the one over the posterior surface of the right arm. He also complained of limited movement of the right ankle joint.

Radiographs taken at this time of ankles, tibiae, elbows, wrists, and left hand, again showed multiple disseminated calcinosis (Figs. 2 and 3). There was an area of calcification over nearly the whole of the right triceps muscle (Fig. 2, B); subcutaneous deposits were also seen around both elbows and the left shoulder. Calcification involved nearly the whole of the left tibialis anticus muscle.

The sedimentation rate, blood uric acid, blood count, blood calcium and acid and alkaline phosphatase, and blood cholesterol were all within normal limits.

The patient was seen by the orthopaedic surgeon, and, in view of the unsightly appearance and discomfort caused by the large size of the triceps swelling, it was decided to explore and if possible remove the calcareous mass.

OPERATION NOTE (by Mr. W. D. Coltart).—The swelling was explored by an incision on the posterior surface of the arm. The posterior wall of the swelling was formed by the fascia of the arm, its anterior wall by the fascia on the posterior surface of the triceps muscle. On incision of the swelling about one pint of fluid with the consistency of white paint was evacuated.

The walls of the swelling were heavily encrusted with calcareous fragments, and it was possible to dissect away the whole of it, leaving the clean surface of the triceps muscle with its tendon below, although some difficulty was experienced in removing the whole swelling at its distal extremity, where it was apparently continuous with an olecranon bursa.

The wound was closed by suture of subcutaneous tissue, and the skin healed by first intention. The patient rapidly regained full range of movements and muscle power of the arm. (For post-operative radiograph see Fig. 2, C.)

HISTOLOGICAL EXAMINATION

The inner surface of the fibrous material contained calcareous material, and section showed a fibrous wall with haemorrhage on one side, and extensive deposits of calcium (Fig. 1). Inflammatory reactions were slight, as is usual with calcinosis.

A post-operative radiograph showed small residual areas of calcinosis. Further nodules are to be excised.

Summary

A case is described of calcinosis circumscripta in a man aged 40 years, with a history of twenty-five years' onset of nodules increasing in size; otherwise he was perfectly healthy. Laboratory investigations were normal. Radiographs showed multiple disseminated calcinosis. A mass over the posterior aspect of the right arm was removed and a layer of calcified fibrous tissue excised. A short survey is given of cases described in the literature and treatments used.

Conclusion

The case is of interest owing to the normal pathological findings and the inability to connect the condition with any circulatory disturbance or family history. The surgical treatment of the arm has so far proved satisfactory from a cosmetic point of view, and providing all the calcified material is removed there should be no recurrence.

I wish to thank the Medical Staff of the British Red Cross Clinic for permission to publish this case, and Mr. W. D. Coltart and Dr. Oswald Savage for their kind assistance with the preparation of this paper.

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FIG. 1.—Photograph to show the walls of the swelling heavily encrusted with calcareous fragments.

FIG. 2.—(A) Calcified mass on posterior aspect of right arm, left elbow, and wrist. (B) Radiograph of right arm before operation, showing calcified mass; and (C) postoperative radiograph.

FIG. 3.—Radiographs of (A) left leg, showing extensive calcinosis; (B) left hand; and (C) left arm, showing calcified mass, but smaller than on the right arm.

FIG. 1.—Illustrates hystercal bent back (camptocormia), the patient being unable (A) to straighten his back while in the standing position, although when recumbent (B) he can do so (note the lumbar lordosis), and although he can accomplish complete anterior flexion (C) without discomfort.

FIG. 2.—Another example of hystercal bent back. Postural correction resulted following narcosis studies and suggestion.

FIG. 3.—A hystercal bent knee. The patient could straighten the knee fully while lying but not while standing. He walked with a bizarre gait, throwing the foot outward and semi-squatting with each step.

FIG. 4.—Illustrating (A) hystercal deformity of the right hand of eleven months' duration. Under sodium amytal narcosis the patient was able to move the fingers normally. Radiographs (B) of the involved hand show marked diffuse secondary bone atrophy from disuse.

FIG. 1.

FIG. 1 (left).—Radiograph of hands showing punched-out areas in the first left interphalangeal and the second left metacarpo-phalangeal joints, originally diagnosed as being gouty in origin, but later shown to be subacute abscesses.

FIG. 2 (opposite page).—(a) Lymph gland, $\times 70$, showing four necrotic germinal centres. (b) Lymph gland, $\times 375$, showing detail of marked degenerative changes in one of these germinal centres, including small necrotic particles of pyknotic nuclear material, lymphocytes, histiocytes, and degenerated background.

FIG. 3 (opposite page).—(a) Metacarpo-phalangeal joint, $\times 4\frac{1}{2}$, showing base of proximal phalanx. Note proliferated synovial membrane directly covering a broad zone of normal bone. The articular cartilage is destroyed. (B) A subacute abscess, $\times 600$, showing polymorphonuclear leucocytes and lymphocytes.

FIG. 2 (b)

FIG. 2 (a)

FIG. 3 (b)

FIG. 3 (a)

FIG. 1.—Subcutaneous nodules on the ulnar edge when arthritis was of 49 years' duration (terminal stage). Good general health.

FIG. 2.—Same case as Fig. 1. Subcutaneous nodule on right clavicle.

FIG. 3.—Subcutaneous nodules on the inner side of thumbs in a patient discharged as markedly improved by treatment. Rheumatoid arthritis of 8 years' duration (terminal stage).

FIG. 4.—Subcutaneous nodules (more than 10 years old) below the elbow joints. Rheumatoid arthritis at the terminal stage, of 15 years' duration. Good general health.