Treatment of Calcinosis Universalis with Aluminium Hydroxide

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A 9-year-old boy with calcinosis universalis due to dermatomyositis was treated with oral aluminium hydroxide with excellent response.

Calcinosis universalis is a rare condition of generalized, predominantly subcutaneous calcification, which in about one-third of cases is secondary to scleroderma or dermatomyositis (Atkinson and Weber, 1938). Treatment so far has been unsatisfactory. A case is reported in which a good response to oral aluminium hydroxide was obtained.

Case Report

This male child was born in February 1958. In early infancy he had an operation of the left kidney, but otherwise he was well until the age of 3½ years when he developed a skin rash and muscle contractures. Dermatomyositis was diagnosed, and the result of a muscle biopsy was as follows. "The muscle shows areas of oedema, a patchy but diffuse inflammatory cell infiltrate composed of lymphocytes, large mononuclears, some segmented neutrophils, an occasional eosinophil, and some plasma cells. Muscle fibres appear to be completely gone, with collapse of the sarcolemmal sheath, and in many areas there is a light to dark blue change in staining reaction of the muscle fibre. Other fibres still contain pink-staining material but have completely lost striations."

Extensive ectopic calcification with ulceration through the skin developed over the next 5 years. 2 years before admission he had deteriorated sufficiently to become chair-bound, but there had been little progress immediately before he was seen. In June 1965, he was treated with corticosteroids, but these were discontinued shortly afterwards because of cellulitis of the right arm. In January 1967, at the age of nearly 9 years he was admitted to The Royal National Orthopaedic Hospital. Examination showed a pale child, small for his age. There was extensive muscle wasting, with contractures and subcutaneous tissue calcification resulting in ulceration. Movement of all joints was restricted (Table I).

Blood. Hb 12.6 g./100 ml.; ESR 43 mm./hr. (Westergren); white cells 6000/cu. mm.; blood urea 20 mg./100 ml.; serum calcium 10.4 mg./100 ml.; inorganic phosphate 4.4 mg./100 ml.; alkaline phosphatase 8.4 King Armstrong units; serum creatine kinase 0.5 ml.; serum proteins—total 6.9 g./100 ml.; albumin 4.3; globulin 2.6; (strip shows increase in α₂-globulin), electrolytes: sodium 138 mEq/l., potassium 4.9 mEq/l., chloride 102 mEq/l., and CO₂ combining power 23.1 mEq/l.

Urine. Mid-stream urine normal; 24-hour urine calcium 33 mg./410 ml.; no myoglobin detected.

X-rays. Very extensive calcification in the soft tissues of the thoracic wall and abdomen. All limbs were similarly affected and the calcification in these areas tended to be of a more linear disposition. Large confluent areas were present in both axillae and around the left hip joint. The hands and feet were only slightly involved. The musculature in all limbs was extremely wasted (Dr. E. H. Allen) (Fig. 1).

TABLE I

Range of Movement of Joints at First Examination

<table>
<thead>
<tr>
<th></th>
<th>Right</th>
<th>Left</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shoulders</td>
<td>−5 to 80°</td>
<td>−5 to 35°</td>
</tr>
<tr>
<td>Elbows</td>
<td>100 to 130°</td>
<td>Fixed in neutral position</td>
</tr>
<tr>
<td>Forearms</td>
<td>Fixed in neutral position</td>
<td>Fixed in pronation</td>
</tr>
<tr>
<td>Wrists</td>
<td>0 to 5°</td>
<td>Fixed 90°</td>
</tr>
<tr>
<td>Hands</td>
<td>Fixed 15°</td>
<td>0 to 15°</td>
</tr>
<tr>
<td>Metacarpophalangeal joint:</td>
<td>Limited movement</td>
<td>Fixed adduction</td>
</tr>
<tr>
<td>Thumbs</td>
<td>45 to 50°</td>
<td>45 to 50°</td>
</tr>
<tr>
<td>Hips</td>
<td>110 to 140°</td>
<td>90 to 130°</td>
</tr>
<tr>
<td>Knees</td>
<td>Fixed in equinovarus</td>
<td>In equinovarus</td>
</tr>
<tr>
<td>Feet</td>
<td>Muscle power poor throughout</td>
<td></td>
</tr>
<tr>
<td></td>
<td>No sensory changes</td>
<td></td>
</tr>
<tr>
<td></td>
<td>No abnormalities</td>
<td></td>
</tr>
<tr>
<td></td>
<td>in other systems</td>
<td></td>
</tr>
</tbody>
</table>

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**Fig. 1a**

**Fig. 2a**

**Fig. 1b**

**Fig. 2b**
Fig. 1 a, b, c, d.—Before treatment with aluminium hydroxide—January 1967.

Fig. 2 a, b, c, d.—After 22 months treatment with aluminium hydroxide—January 1969.

Films taken in January 1969, show the remarkable clearance after therapy. Considerable regeneration of muscle is shown (Fig. 2).

Progress. He was treated with aluminium hydroxide (Aludrox) 15 ml. × 4 a day (2.4 g. aluminium oxide per day). Within 6 months there was considerable clearing of the calcium deposits, and further improvement took place over the next 18 months. His disease continues clinically in remission, and at the beginning of 1968 his ESR had fallen to 6 mm./hr. The reduction in his calcium deposits continued, and by the end of 1968 being aged 10½ years, he had only one lesion which was slightly ulcerated. Aluminium hydroxide treatment had by then been given for 1½ years. The power of his muscles had increased though they were still very wasted; function was good considering the degree of deformity due to contractures. However, even his fixed deformities had become less, and range of movement is given in Table II. X-rays (Fig. 2) confirmed reduction in calcification, and also considerable regeneration of muscle. Blood urea 19 mg./100 ml., urine calcium 70 mg./24 hr., and urine phosphate 568 mg./24 hr.

Discussion

The best approach to the treatment of this child seemed to be reduction in the intake of the consti-
aluminium phosphate, decreases the intestinal absorption of phosphate. In this case the deficiency of phosphate resulted in satisfactory reduction of his ectopic calcification. There was also a possibility that it might lead to reduction in bone mineralization. This has not been seen yet; but the bones of the patient will require close observation while treatment is continued.

As response to treatment with steroids is generally disappointing (Transactions of the St. John's Hospital Dermatological Society, 1967), despite the report of Schmidt-Peter and Pfeil (1966), oral aluminium hydroxide may prove to be the treatment of choice in calcinosis universalis.

REFERENCES


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