CASE REPORTS

SCLEREMA ADIPOSUM: A CASE WITH UNUSUAL FEATURES IN AN INFANT

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This condition is not common and there is much confusion in the literature regarding the exact nomenclature of the various types of subcutaneous fat induration occurring in young infants.

Sclerema adiposum was originally described by Usembenzius (1722), and some years later Underwood (1784) reported the condition in some detail. A classical description of the types of induration occurring in the subcutaneous tissues of newborn infants was given by Ballantyne (1890). Since then, however, a number of distinct clinical and pathological conditions have been grouped under the one heading of 'sclerema neonatorum.' It has become especially difficult to differentiate between cases of sclerema adiposum and those of subcutaneous fat necrosis. Harrison and McNee (1926) described a number of cases of sclerema adiposum but did not separate cases of subcutaneous fat necrosis from those of true sclerema. Channon and Harrison (1926) investigated the chemical changes occurring in the fat in both conditions. Fox (1933) describes his cases under the name 'subcutaneous fat necrosis,' a term introduced by Fabyan (1907) and revived by Bernheim-Karrer (1922). Gray (1933), on the other hand, prefers the term sclerema adiposum when describing cases with similar clinical features to those of Fox (1933).

More recently Hughes and Hammond (1948) have reviewed the cases of sclerema adiposum occurring in the literature from Ballantyne (1890) onwards, and have added three cases of their own. They rejected all cases of subcutaneous fat necrosis and confined their survey to cases which fitted their definition of sclerema neonatorum.

The following case exhibited the clinical features and pathological changes of sclerema adiposum.

Case Report

Case history. The patient was a male infant aged 6 months at the time of admission on Dec. 24, 1947. He was the first child of healthy middle-aged parents. Born by Caesarean birth at full term, he

weighed 8½ lb. Fed on National Dried Milk with adequate addition of vitamins, he thrived normally. He had not had any previous illness.

Two weeks before admission he appeared a little listless and 'off colour,' and did not use his arms or kick his legs about normally. A day or two later the mother noted some swelling of the legs, affecting first the left leg, but within two days both legs were swollen and brawny. She noticed that the skin over the swollen parts was shiny, especially over the calves, and she thought that the legs were a little painful when touched. A week before admission the arms began to show similar swelling and the swelling became generally more marked. The baby had remained fairly well and was taking his feeds normally. The bowels had opened normally. There had been no pyrexia.

Clinical examination. The patient was a fat, healthy looking, well nourished but rather pale infant of 18 lb. 6 oz., who lay quietly in bed taking an interest in his surroundings, but who appeared to resent handling. The general impression was of a healthy skin, but the legs, especially the calves, were hard, swollen, and brawny. There were also 'nobbly' hard swellings on the thighs just above the knees, and on the buttocks and arms. These swellings were well defined and felt like fat masses. The arms were swollen from the shoulder to the wrist. This swelling was very hard, and had a well defined margin above at the shoulder, and a more noticeable one at the wrists, the latter giving the impression of a cuff. The hands and fingers were not swollen. These swellings were not tender and did not pit on firm pressure. The skin was firmly adherent to the masses and could not be wrinkled. The limbs were held stiffly and had a deep purplish colour. The hard swelling affected the outside of the limbs much more intensely, the inner aspects appeared much more normal on handling. There were firm, hard patches on both cheeks, which were due to involvement of the sucking pads; this gave the face a stiff look. The buttocks were hard and tense and beginning to ulcerate. The apparently unaffected areas were the scrotum, hands, fingers, toes, soles of the feet, back of the trunk, abdomen,

the flexures and internal aspects of the limbs and mucous membranes. There was no adenitis; the mouth and throat were normal. No abnormality was detected on examination of the chest, abdomen, or nervous system. The temperature was 99° F. on admission but otherwise remained between 98·4° and 97·6° F. until death seven days later. The white blood count was 26,000 per c.mm. of blood; polymorphs 62 per cent., lymphocytes 36 per cent., red blood count 2,560,000 per c.mm., haemoglobin 55 per cent. (Sahli). The blood chemistry showed the following figures: serum phosphatase 8 units, serum phosphorus 3·2 per cent., serum calcium 9·4 per cent., serum protein, total 5·38 per cent., albumin 3·69 per cent., globulin 1·69 per cent., blood urea-nitrogen 18 mg. per 100 c.cm., chlorides 420 mg. per 100 c.cm., cholesterol 138 mg. per 100 c.cm.

On the sixth day after admission the infant became more listless, and developed slight diarrhoea. He lost weight rapidly, and suddenly died on the eighth day of our observation.

Post-mortem report. The body was that of a fairly well developed male infant. A curious change was present in the subcutaneous fat. Large firm swellings were present over the four limbs but avoided the flexures. The cheeks were the site of a brawny swelling. In these areas the skin was tense and the underlying fatty tissue was very firm and hard. The subcutaneous tissue was freely movable on the underlying musculature. A small bed-sore was present over the buttocks. Post-mortem lividity was present over the upper trunk. No congenital abnormalities were found on external examination.

The pharynx and oesophagus were healthy. The thyroid and thymus glands showed no abnormality. The trachea and bronchi were clear and covered with a thin film of mucus. The pleura was clear and glistening and there was no free fluid in the pleural sacs. The lungs were well expanded and reddish-pink in colour. On section there was considerable congestion of both lower lobes but no inflammatory reaction had occurred and there was no pneumonia. The pericardium was normal. The heart was of normal size and shape, and the endocardium was healthy. No congenital abnormalities were present.

The peritoneal sac was healthy and the alimentary tract showed no pathological change. The liver, spleen, and kidney were normal. The suprarenal glands had a normal appearance and the pancreas appeared to be healthy. The dura mater and pia-arachnoid were healthy. The brain was well developed and rather soft, but on section no intracerebral lesion was found.

Histological report. The cutis was quite normal in appearance. The subcutaneous fatty layer was grossly thicker than normal and the fat cells were larger than usual. Their walls were not sharply defined, and they stained deeply with eosin. Running throughout the adipose tissue were intersecting bands of

fibrous tissue. These trabeculae were much larger than usual and were densely infiltrated with inflammatory cells. These cells were mainly polymorphonuclear leucocytes, but lymphocytes and mononuclear macrophages were also present. Foreign-body giant-cells were not present. There was considerable endarteritis obliterans in the small vessels in the trabeculae, and these arterioles were surrounded by a dense polymorphonuclear infiltration. Special staining of the skin sections was carried out. Scharlach R stained the fat a bright red but failed to reveal any fat crystals. Nile blue sulphate failed to reveal the presence of free fatty acids, and Benda's stain showed that no necrosed fat was present. Thus only neutral fat could be demonstrated in these sections.

The skeletal muscle was completely unaffected. In the kidney the capsule, glomerular epithelium, and Bowman's capsule were normal. The convoluted tubules showed moderate cloudy swelling. The collecting tubules were healthy.

In the myocardium the fibres were pale and showed cloudy swelling. Some inflammatory cell infiltration had occurred. The cells were mainly polymorphonuclear leucocytes. The appearance was that of toxic myocarditis.

The liver capsule was normal. The liver cells were healthy and the normal lobular architecture was preserved. There was dense infiltration of polymorphonuclear leucocytes. These were present in the sinusoids and in the portal tracts. The bile-duct epithelium was healthy.

Examination of fat from the chest and abdomen failed to reveal changes similar to those found in the subcutaneous adipose tissue.

This was a case of sclerema adiposum with well marked lesions in the subcutaneous adipose tissue. There was probably a terminal septicaemia.

Comment

As Sternbach and Robinson (1947) have pointed out, the diagnosis of sclerema has not always been applied to conditions of similar etiology. As Eichenlaub and Sandler (1937) remarked, five separate clinical entities tend to be grouped under the heading 'sclerema neonatorum.' These are scleroedema, oedema, scleroderma, subcutaneous fat necrosis, and sclerema adiposum. Oedema and scleroedema show pitting on pressure and are more generalized in the early stages. Scleroderma is a condition involving the skin; the subcutaneous fatty layer is unaltered. In the present case there is clearly no involvement of the skin (fig. 2).

Subcutaneous fat necrosis may be differentiated both by the clinical and by the pathological findings. It is a less fatal disease and occurs more often over the shoulders and posterior aspect of the trunk. It exhibits less tendency to become generalized than is the case in sclerema adiposum. Calcification or cyst formation may be expected. Histologically the cellular picture is also dissimilar. The predominant cell is the lymphocyte, and a foreign-body giant-cell

reaction is not uncommon. As has been stated, however, all authors do not necessarily make the same distinction between the two diseases. Harrison and McNee (1926) described several cases of sclerema adiposum whose histological changes more closely resembled those of subcutaneous fat necrosis described by Fox (1933). More recently Zeek and Madden (1946) have described a case of sclerema adiposum where the histological appearances are of a chronic nature.

Our case presents several unusual features from both the clinical and pathological points of view. Clinically, the manifestations are strikingly similar to the majority of the cases reported as sclerema adiposum neonatorum, especially in the typical distribution of the areas in which the indurated fat lesions occur in the subcutaneous tissues, and in their firm non-pitting and non-tender character. In all previous reports the onset of the fat changes was noted early in life and usually a few days after birth, but always within the first twenty-one days of life; death followed after several weeks or months of illness. In our patient the onset was at 5½ months of age and death occurred within three weeks.

The pathological features also do not completely conform with those of other recorders. Harrison and McNee (1926), Zeek and Madden (1946), and others have described cases with a chronic inflammatory reaction in the subcutaneous fatty layers. In these cases giant-cell formation of the foreign-body type has been a prominent feature and crystals of neutral fat have been found in the affected tissue. Hughes and Hammond (1948), on the other hand, classify all such cases under the heading of subcutaneous fat necrosis. They confine their series to cases which show thickening of the connective tissue bands in the subcutaneous fatty layers as the only constant feature. In their cases cellular infiltration is absent.

In the present case, however, the findings were not completely typical of either group. Considerable thickening of the connective tissue bands had occurred, but dense cellular infiltration was also present (figs. 3 and 4). This cellular reaction was not chronic in character but was very acute. The

predominant cell was the polymorphonuclear leucocyte. There were only a few lymphocytes, and no giant-cells were found. Thus the appearance was that of a lesion of an acute character, and this is in complete accord with the clinical findings. Harrison and McNee (1926) in their review postulate an infective process as the underlying causal agent in this disease. This hypothesis is an attractive one and would seem to meet this case, since there is reason to believe that there was terminal septicaemia.

A thorough bacteriological examination of an acute case of sclerema adiposum should help to clear up the obscurity surrounding the etiology of the condition.

Summary

A case of sclerema adiposum is reported. The case showed several unusual features, namely the age of onset and the acuteness of the disease process revealed by histological examination. The histological appearances would appear to indicate that an acute infective process is at work.

REFERENCES

- Ballantyne, J. W. (1890). *Brit. med. J.*, **1**, 403.
- Bernheim-Karrer, J. (1922). *Schweiz. med. Wschr.*, **52**, 12.
- Channon, H. J., and Harrison, G. A. (1926). *Biochem. J.*, **20**, 84.
- Eichenlaub, F. J., and Sandler, I. L. (1937). *South. med. J.*, **30**, 527.
- Fabyan (1907). *Bull. Johns Hopk. Hosp.*, **18**, 348.
- Fox, H. (1933). *Arch. Derm. Syph. Chicago*, **27**, 237.
- Gray, A. M. H. (1933). *Brit. J. Derm. Syph.*, **45**, 498.
- Harrison, G. A., and McNee, J. W. (1926). *Arch. Dis. Childh.*, **1**, 63.
- Hughes, W. E., and Hammond, M. L. (1948). *J. Pediat.*, **32**, 676.
- Sternbach, A., and Robinson, J. (1947). *Ibid.*, **31**, 465.
- Underwood, M. (1784). *A Treatise on the Diseases of Children*. London. p. 76.
- Usembenzius, J. A. (1722). *Acad. Nat. Curios. Ephem.*, **30**, 62.
- Zeek, P., and Madden, E. M. (1946). *Arch. Path.*, **41**, 166.

FIG. 1.

FIG. 1.—Infant with sclerema adiposum.

FIG. 2.—Section of skin overlying sclerematous tissue. The skin is quite normal in appearance. There is no oedema or inflammatory reaction. $\times 100$.

FIG. 3.—Section showing large fat cells and a broad central trabeculum. A small vessel is present and shows endarteritis obliterans. There is considerable perivascular cellular infiltration. $\times 100$.

FIG. 4.—Section showing a broad trabeculum in which there is heavy polymorphonuclear cell infiltration. $\times 400$.

FIG. 2.