BESNIER-BÖECK'S DISEASE IN AN INFANT

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Besnier-Böeck's disease usually affects young adults, but may occur at any age. It is rare in children, and is met with only exceptionally in infancy. The case reported here is therefore of considerable interest.

Besnier (1889) described a syndrome characterized by bluish-red or violet infiltrations of the skin of the fingers, toes, cheeks, and nose. It bore some resemblance to the ordinary chilblain and was called by him *lupus pernio*. Böeck (1899) reported several cases having red or bluish coloured infiltrations of the skin, and enlargement of lymph glands. To this syndrome he gave the name of benign sarcoid of the skin. He came to believe that the lesions were inflammatory in nature and probably tuberculous. He later renamed the disease benign miliary lupoid. Schaumann (1924) put forward the view that Böeck's sarcoid was merely part of a general disease affecting many other organs besides the skin. He showed that Böeck's sarcoid and *lupus pernio* were identical histologically, the lesion being an area of cellular infiltration consisting of endothelial cells, some lymphocytes and a variable number of multinuclear giant cells. There was, however, never any caseation as in tuberculosis. Later a progressive fibrosis usually took place. He emphasized the visceral localization of the disease and described lesions in the lungs, and lymphatic tissues. He shared Böeck's opinion that the disease was a form of tuberculosis. Jungling (1921) described under the name *osteitis tuberculosa multiplex cystica* curious cystic spaces in the bones of the hands and feet of patients who suffered from sarcoid. Other bones may also be affected.

A voluminous literature has gathered round this interesting malady. Hunter (1936) has given an historical account of the earlier papers written about it, and Volk (1931) has recorded a full description of the skin lesions. A complete account of the syndrome together with a report of eight cases is to be found in the paper by Longcope and Pierson (1937). In Snapper's (1938) beautifully illustrated monograph, which was recently published, thirteen more cases are reported.

Case report *

Bernard, C., a male aged two years, was first seen in January, 1938, at the Hospital for Sick Children, Great Ormond Street. For three months he had had painless swellings of the fingers of both hands. He was a ten weeks premature infant, born of unrelated parents ; his birth weight was 4½ lb. His parents were healthy. There are four other children, all normal. There was no history of tuberculous contact.

On examination, the boy was pale. His weight was 24 lb. Both eyes were slightly prominent. The respiratory and cardio-vascular systems were normal. The blood pressure was 115/80 mm. Hg. The spleen and liver were not palpable. The lymph glands were not enlarged. There was a fusiform enlargement of all the fingers of both hands (fig. 1). The skin was shiny and slightly reddened.

Fig. 1.—The photograph shows the sausage-shaped swelling of the fingers and the shiny skin.

The toes were normal. There were no other skin lesions. The optic fundi were normal and the media were clear. The patient was afebrile while he was in hospital.

Investigations :

BLOOD COUNT : Red blood cells, 4,560,000 per c.mm. ; Haemoglobin, 62 per cent. ; Colour index, 0·7 ; White blood cells, 7,000 per c.mm. ; Polys., 44 per cent. ; Lymphos., 51 per cent. ; Monos., 2 per cent. ; Eosinos., 2 per cent. ; Basophils, 1 per cent.

WASSERMANN REACTION : Mother and Child—Negative.

MANTOUX REACTION : 1 in 1,000 to 1 in 10—Negative.

* This case was shown at the Children's Section of the Royal Society of Medicine on March 25, 1938, see *Proc. roy. Soc. Med.* 1938, 31, 1124. The Honorary Editors have kindly granted permission to publish it here.

PLASMA PROTEINS : Fibrin, 0.34 gm. per cent. ; Albumin, 4.0 gm. per cent. ; Globulin, 1.54 gm. per cent. ; Total proteins, 5.88 gm. per cent.

SERUM CALCIUM : 10.2 mgm. per cent.

INORGANIC BLOOD PHOSPHORUS : 4.6 mgm. per cent.

PLASMA PHOSPHATASE : 8 units.

ERYTHROCYTE SEDIMENTATION RATE : 15 mm. in 1 hour.

X-rays :

HANDS : There were multiple small cystic spaces in the phalanges of both hands. The changes were most extensive in the third and fourth fingers of the right hand and the second and third of the left (see fig. 2). The metacarpals were unaffected.

FEET : No changes in the phalanges and metatarsals.

CHEST : The lungs were normal. There was no increase in the mediastinal shadow.

LONG BONES AND SKULL : Normal.

Fig. 2.—The small cyst-like spaces are best seen in the phalanges of the second and third fingers of the right hand and the first and second of the left hand.

Course : In July, 1938, there was no change either locally in the hands or in the patient's general condition. No further lesions had appeared. A piece of soft tissue was removed from one of the affected phalanges and was examined histologically.

Histological examination (Dr. D. N. Nabarro) (see fig. 3 and 4).

The section is composed of fibrous tissue in which there are small cellular areas consisting of lymphocytes and endothelial cells. There are also several giant cells with many nuclei arranged roughly around the periphery of the cell. They resemble but are not identical with those met with in tuberculous gran-

ulation tissue. There is no caseation. No tubercle bacilli could be found in Ziehl-Nielsen stained preparations. In slides stained with methylene blue,

FIG. 3.—Photomicrograph showing the area of cellular infiltration amongst the mass of fibrous tissue. Several giant cells are discernible. $\times 40$.

clusters of small coccoid bodies were observed. They were situated both intra- and extra-cellularly. There was no inflammatory reaction around them. It is

FIG. 4.—High-power view ($\times 200$) of the same section showing several giant cells.

possible that they were granules extruded from mast cells. A piece of the tissue was inoculated into a guinea-pig. No evidence of tuberculosis was found when the animal was killed eleven weeks later.

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Discussion

The usual age of onset is in early adult life, between the ages of twenty and thirty years. An onset in childhood is rare.

Roos (1937) describes a case in a girl aged four years, who had lesions in the skin, liver, spleen, kidneys, lymph nodes and brain. He quotes from the literature nine other undoubted cases in children. All but two were over six years of age ; one was one and a half years old and another only four months of age. All of these patients had skin lesions ; five had cystic changes in the bones ; two had enlargement of lymph nodes, and in two others there was enlargement of the mediastinal lymph glands. Stalmann's patient (1933) was only one and a half years of age and had lesions in the hands which closely resembled those in the case described here. There were spindle-shaped swellings of the fingers and the skiagram showed multiple cystic spaces in the phalanges. Müller (1938) reports a case in a twelve-year-old girl. She had generalized swelling of the lymph glands, diffuse lesions in the lungs and early ones in the phalanges. There was a typical histological picture. Naumann (1938) describes two cases, one in an infant of three months and a second in a child aged fourteen years. In the latter there were diffuse lesions in the lung as shown by x-ray. In both the Mantoux reaction was negative.

It is now recognized that skin lesions are found in little more than half the cases of Böeck's sarcoid. There is a well-defined group in which the deposits are found only in the viscera and glands. In some the bones only are affected. The present case belongs to this latter group. Other manifestations of the disease may appear in the course of time.

The etiology of the disease has been the subject of much discussion. Böeck, Schaumann and others believed it to be a manifestation of tuberculosis. Many investigators, however, declare that tubercle bacilli cannot be demonstrated in the affected tissues either histologically or by guinea-pig inoculation. Moreover, it is a characteristic of the lesions that they do not, as in tuberculosis, proceed to caseation. The present case lends support to the non-tuberculous theory of origin. The Mantoux reaction was negative to low dilutions of tuberculin, no tubercle bacilli were seen in the sections, and inoculation into a guinea-pig did not give rise to any tuberculous lesions. Longcope and Pierson, Snapper, and other recent writers hold the view that Böeck's sarcoid is a specific reaction of the tissues of the body to an unknown virus.

Summary

A case of Böeck's sarcoid in a boy aged two years is described. There were characteristic cystic spaces in the x-ray picture of the phalanges and spindle-shaped swellings of the fingers. A piece of tissue removed from a finger showed a typical histological picture. The incidence of the disease in children is discussed.

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