decreased range of motion to avoid further functional deficits and contractures. Methods to manage acute bleeds of the extremities and the complication of contractures have been evaluated by DePalma (1967) and Ahlberg (1967).

The prime importance of the preventive aspects in the physical management of chronic disease was illustrated in this case. The psychological, social, and vocation aspects of management have not been included in this paper but are of great significance.

Summary

Spinal cord bleeds with compression are a relatively rare complication of haemophilia. Review of the literature indicates that most cases are not associated with significant trauma or spinal fracture and the course of recovery is variable. A case of quadriplegia is presented emphasizing problems in skin care, bladder management, and mobilization during rehabilitation. Prophylaxis and control of continuous bleeds, catheter-free status, and early management of extremity bleeds are stressed in the light of preventing complications resulting in further disability.

REFERENCES


MARJORIE L. KEELY,* NEAL TAYLOR,† and RONALD L. CHARD, JR.
Department of Rehabilitation Medicine, University of Washington School of Medicine, and the Children's Orthopedic Hospital and Medical Center, Seattle, Washington, U.S.A.

*Present address: St. Joseph Hospital, Albuquerque, New Mexico, U.S.A.
†Correspondence to Dr. Neal Taylor, Department of Rehabilitation Medicine, Children's Orthopedic Hospital & Medical Center, 4800 Sandpoint Way, N.E., Seattle, Washington 98105, U.S.A.

Histiocytoma Presenting as Swelling on the Toe

Histiocytomata are common in adults and well known to dermatologists, but these tumours are rare in children. The tumour usually presents as a painless single nodule, slowly growing, sharply circumscribed, and varies in colour depending on the vascularity, lipid, and haemosiderin content. Two cases seen during the past five years presented as swellings of the toes in children below the age of 1 year.

Case Reports

Case 1. Mother noticed a swelling on the left second toe 10 days after normal delivery of a twin. This gradually increased in size during the following 6 months at the same rate as the growth of the toes. On examination, there was a hard tumour approximately 1 cm long on the dorsum of the left second toe, extending from the nail bed to the skin crease at the distal interphalangeal joint (Fig. 1). This was observed at monthly intervals in the outpatients, but as the tumour appeared to grow and become softer, excision was advised. At operation, the distal phalanx appeared to be involved in the tumour, and disarticulation was performed at the terminal interphalangeal joint. The postoperative course was satisfactory, and at follow-up in 1970, there was no recurrence of the tumour.

Histology. The overlying epidermis was normal. The underlying dermis contained a non-encapsulated tumour composed of interwoven bands of fibrous tissue surrounding some blood vessels (Fig. 2). Very few inflammatory cells or histiocytes were present but the appearances were those of a histiocytoma.

Case 2. At the age of 5 months, mother noticed a small, reddish swelling appearing on the dorsal surface

Fig. 1.—Histiocytoma on second left toe of Case 1.
of the left third toe, just proximal to the nail. This gradually increased in size until seen 6 months later.

Examination showed a swelling approximately 1 cm in diameter present over the dorsomedial aspect of the terminal phalanx of the left third toe, which was bluish-red and nontender. An excision biopsy was performed through the terminal interphalangeal joint using a posterior skin flap. The postoperative course was satisfactory and the patient was discharged from hospital with the wound healed.

Six months later, the mother noticed that the swelling had gradually recurred over the dorsum of the toe, and when seen there was a recurrence approximately 1 cm by 0.25 cm, fixed to the underlying phalanx. A disarticulation of the stump of the left third toe through the metatarsophalangeal joint was performed.

Histology of the original tumour showed a very similar appearance to that described in Case 1, with interwoven bands of fibrous tissue encompassing blood vessels. The recurrent tumour showed a similar appearance, but the fibrous tissue was more sclerotic, possibly due to a greater age of the lesion or because of the previous biopsy. No evidence of malignant change was seen in any of the tissue examined.

Discussion

Histiocytoma is a common dermal tumour composed of blood vessels, fibrous tissue, histiocytes, and inflammatory cells of varying proportions. The histology of histiocytoma reveals three basic patterns. The first is that of a preponderance of whirled fibrous tissues (dermatofibroma); secondly a vascular tumour with variable degree of reactive fibrous obliteration of the vessels (a sclerosing angioma); and finally, the type containing many active histiocytes.

This variability of pattern has given rise to dispute over the pathogenesis of the lesion. Gross and Wolbach (1943) laid great stress on the basically vascular nature of the tumour. Dawson (1948) was of the opinion that histiocytoma was the result of haemorrhage in a capillary haemangioma with ensuing fibrosis. Many histiocytomata contain large numbers of haemosiderin-laden macrophages indicating previous haemorrhage which gives some support to this hypothesis.

However, more recent writers, notably Kauffmann and Stout (1961), feel that the tumour is not essentially vasoformative in nature, suggesting that the collagen is produced by histiocytes which have assumed a facultative fibroblastic function.

The appearances in the two cases here presented are those of a rather quiescent, later dermatofibroma. They show no evidence to support the theory of an angiomatous aetiology as no haemosiderin-containing macrophages could be seen. There may be recurrences if excision is incomplete, as reported by Kauffman and Stout (1961).

However, the main interest in these two cases lies in the age of the patients and the site of the lesions. Neither of these cases was diagnosed

**FIG. 2.**—Histology showing interwoven bands of fibrous tissue surrounding some blood vessels. (H. and E. × 30.)
Glycogen Storage Disease,
Type VIII*

Glycogenolysis occurs in a sequential series of reactions involving hydrolysis of \( a_1 \rightarrow 4 \) and \( a_1 \rightarrow 6 \) glycosidic linkages, resulting in the liberation of phosphorylated and free glucose. One enzymatic component of this system, phosphorylase, is activated by a kinetic cascade mechanism mediated by cyclic AMP as a consequence of hormonal action. Since 1966 (Hug, Schubert, and Chuck, 1966b), disease states characterized by phosphorylase deficiency and glycogen accumulation have been shown to occur from defects in phosphorylase kinase, in the 3',5'-AMP dependant kinase system (Hug et al., 1970a), and from absence of phosphorylase.

The diagnosis of these genetic enzymopathies is dependent upon the delineation of the biochemical defect, the accumulation of glycogen of normal structure and the occurrence of certain clinical symptoms. Recent investigations (Huijing and Fernandes, 1969) have employed leucocyte phosphorylase assays to establish the diagnosis. This report presents studies of carbohydrate homoeostasis in a young girl with a defective hepatic phosphorylase system and suggests the unreliability of the leucocyte assay as an indicator of hepatic phosphorylase activity.

Case Report

A 17½-month-old black girl was seen with a referral diagnosis of hepatomegaly secondary to glycogen storage disease. She was the product of a term gestation with an uncomplicated neonatal period. The patient was admitted to hospital at 3 months of age because of diarrhea and dehydration. At 7 months of age, the mother noticed the onset of abdominal enlargement which progressed to the present time. She was admitted to Tampa General Hospital at age 16 months for diagnostic evaluation; fasting blood sugars ranged from 20–40 mg/100 ml, with normal postprandial blood sugar values, bone-marrow examination was within normal limits, and X-rays of the chest and skull were normal. A liver biopsy showed enlarged cells laden with glycogen and without signs of perportal fibrosis. There was no family history of liver disease, disorders of carbohydrate metabolism (including diabetes), or other significant family illnesses.

Physical examination revealed a small girl with a very protuberant abdomen. The significant physical finding was a massively enlarged liver which extended to the iliac crest; it was diffusely enlarged, non-nodular, and firm to palpation. Haematological evaluation revealed a markedly prolonged clotting time. Blood sugar determinations throughout 24-hour periods indicated on some occasions hypoglycaemia after fast periods of up to 10 hours. Liver function tests indicated increased levels of transaminase enzymes and a marked rise in alkaline phosphatase; uric acid level was high normal for our laboratory. A modest rise of blood lipids was also noted (570 mg/100 ml). Glucose tolerance tests were normal, showing a normal insulin response and a concordant depression of blood lactate (Table I). A normal rise in blood glucose occurred after glucagon stimulation (1 mg).

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