Short Reports

Spinal Cord Compression as a Complication of Haemophilia

Neurological complications of haemophilia are more common than once thought. Because of the greater life expectancy with modern treatment methods, functional deficits of these complications assume increasing importance. Most previous reports have been concerned with intracerebral complications. Silverstein (1960) reviewed 174 cases of haemophilia and found 6 proven cases of intracranial bleeding and 5 probable cases. He found 28 peripheral nerve lesions in 206 haemophiliacs (Silverstein, 1964). There have been few reports of spinal cord bleed as a complication of haemophilia. Aggeler and Lucia (1944) reviewed the literature up to 1944 regarding neurological complications of haemophilia. They found 45 cases reported with bleeding involving the central and peripheral nervous system. 11 patients with spinal cord involvement, including 3 quadriplegics, were reviewed. 6 cases had spinal cord bleed with involvement of the spinal meninges, including epidural, subdural, and subarachnoid bleeds. 5 cases had bleeds into the spinal cord. 4 died within several weeks of onset, 2 had partial cauda equina lesions with improvement or recovery, while the others had residual neurological deficits, or the course was unknown.

Three cases of spinal cord compression in haemophilia unassociated with cervical fracture or trauma have been reported in more detail. Schiller, Neligan, and Budtz-Olsen (1948) reported a case of subdural haematoma causing paraplegia in a 16-month-old haemophiliac. Laminatectomy with clot removal was performed. Haemostasis was a problem postoperatively, but the neurological status improved significantly. Schenk (1963) studied the pathological findings in a 21-year-old haemophilic paraplegic who died from wound bleeding after laminatectomy. Syringomyelia with glial proliferation was present from cervical to lumbar region, and the author speculated that this might have been a reaction to repeated bleeds. Fessey and Meynell (1966) reported complete C-6 quadriplegia in a patient with mild factor IX deficiency. The patient improved immediately after administration of fresh frozen plasma and went on to complete recovery within one week.

Case Report

A 6-year-old white boy with haemophilia (factor VIII less than 1%), relatively resistant to cryoprecipitate, had factor VIII levels of 11 to 14% after receiving 10 units of cryoprecipitate (normally this would approach 50% level). He was seen on numerous occasions for treatment of haemarthrosis and soft tissue bleeds. In August 1970, he fell, hitting the lumbosacral region of his back without immediate apparent injury. During the next several days he was febrile, lethargic, and remained in bed. He presented to the hospital with abdominal distension and obstipation. On examination on admission to the hospital, the patient had no response to pin below T2-3 area, but touch was appreciated. Abdominal breathing was present. No voluntary motion was observed in the lower extremities. Knee-jerks were absent and ankle-jerks hypoactive. Plantar stimulation caused flexor withdrawal of the lower extremities. There was a question of weakness of the distal upper extremities. Admission haematocrit was 15, BUN 138, arterial Po2 46, Pco2 38. Spine and skull series were negative. Catheterization of the bladder

References


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produced 800 ml urine and a Foley catheter was left in place. The patient was treated with cryoprecipitate and blood transfusion. He required ventilatory assistance for the first week but subsequently did not have any respiratory problems. A lumbar puncture on the fourth day of admission showed some xanthochromia of CSF with protein of 60 mg/100 ml. There was no evidence of subarachnoid block. Myelogram was not done. An operation was not considered because of his resistance to cryoprecipitate. The clinical diagnosis was spinal cord bleed and the patient was treated, with three weeks of bed rest with the usual attention to skin, bowel, and bladder care, and range of motion. Neurological examination two weeks after admission revealed decreased sensation to pin below T2-3, but position and touch sensation were intact. No voluntary motion was present in the lower extremities, and hyperactive reflexes were present with ankle clonus and Babinski sign bilaterally. In the upper extremities there was weakness of triceps and wrist groups bilaterally and of the right hand intrinsics and no voluntary motion of the left hand groups. The patient subsequently has shown little change in neurological status except for some voluntary motion of the toes bilaterally.

Three weeks after admission, the patient was transferred to the rehabilitation service and was discharged three months later. The following problem areas common to rehabilitation in quadriplegia were complicated by haemophilia.

**Neurogenic bladder.** An indwelling catheter was in place for 7 weeks with development of bacteriuria and two episodes of haematuria. By the seventh week strong spastic contractions were seen on cystometrogram, and IVP and cystogram were normal. The catheter was removed and residual urines were low, 35 to 50 ml. Bacteriuria cleared on a course of antibiotics. The residual urines increased to 75 to 90 ml but because of the imminent complication of bleeding with an indwelling catheter, it was decided to leave the catheter out and follow urine cultures. Residual urines were checked on several occasions without bleeding occurring. The urine remained sterile without antibiotics and the patient has remained catheter free. At present, he is dry by day but not at night.

**Skin.** The patient developed a small sacral decubitus sore which healed well with avoidance of pressure to the area. The danger of bleeding from an open wound was anticipated but did not develop.

**Range of motion.** The patient maintained good range of motion of the paretic extremities with passive exercises. However, soft tissue bleeds of the right upper arm and the gastrocnemius groups bilaterally led to some decreased range of movement of the elbow and heel cords, respectively. Manual stretching was done with caution. Because of the patient's relative resistance to cryoprecipitate a more vigorous exercise or stretching programme was not undertaken. Ankle dorsiflexion was maintained at zero degrees.

**Mobility.** Ten weeks after admission the patient was able to crawl on the mat, do a wheelchair transfer with minimal assistance, and propel his wheelchair. His age and the weakness in his extremities with abnormal body mechanics made him more susceptible to trauma, and bruises were always present on the extremities. These conditions did lead to further functional deficits. He developed a large tense haematoma of the soft tissues of the right upper arm with subsequent median and ulnar palsies. Since this was his more functional extremity, he was no longer able to do transfers. Four months after onset he was able to crawl on the mat and propel his wheelchair but required much assistance with transfers. The patient started school after discharge and at present remains dependent on his wheelchair.

**Discussion**

Diagnosis and management of spinal cord compression presents certain problems in haemophilia. Lumbar puncture was found to be a safe procedure in haemophiliacs, most of whom had not received substitute therapy. Silverstein reported 100 lumbar punctures performed in haemophiliacs with the only complication of bleeding externally at the site of puncture in two cases (Silverstein, 1960). In patients who respond to cryoprecipitate with an increase in factor levels to greater than 30%, more vigorous workup with myelogram can be done followed by surgical decompression if indicated. The early administration of cryoprecipitate and surgical decompression might prevent progressive or permanent neurological deficit, but further clinical observations must be made to evaluate the most appropriate management of the acute bleed. Since our patient was resistant, myelogram and operation were not done. The site of bleeding was thus not proven, but clinically the mild CSF xanthochromia and normal CSF dynamics suggest haematomyelia. As in the three cases reviewed, there was no fracture of the spine in this case. The bleed was apparently precipitated by minor trauma in this patient.

For patients with inhibition of factor VIII, such as in the case presented, Biggs (1970) has suggested a method of cryoprecipitate administration. High potency concentrate in high doses should be given over a short period.

Catheterization for residual urines did not result in bleeding in our patient, which suggests that intermittent catheterization might be preferable to an indwelling catheter in the initial phase of bladder management. Achievement of the catheter free state is the ultimate goal.

Bleeds of the soft tissues and joints of the functional and paralysed extremities should be treated immediately at the first sign of pain or
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


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.](http://adc.bmj.com/adc/47/255/826/fig1.jpg)