INTRASPINAL NEUROBLASTOMA IN A NEWBORN BABY

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(RECEIVED FOR PUBLICATION NOVEMBER 4, 1957)

Intraspinal and spinal cord tumours are quite rare in childhood. Most authors reporting cases of tumours found within the spinal canal use the term 'spinal cord tumour' in a broad sense without regard to whether they are spinal cord tumours proper or of extradural origin. In this paper we refer to an 'intraspinal' neuroblastoma in a newborn infant, and wish to emphasize that though it is intraspinal, yet it is not a tumour of the spinal cord proper, but is situated extradurally.

Anderson (1953) described 21 cases, of which the youngest was 5 months old. Dandy (1925) reported on such tumours in 36 patients of whom five were under 15 years of age. Ford (1944), amongst 70,000 neurological cases in children, found only three spinal cord tumours, the youngest child with this type of tumour being 9 years old. Stookey (1928) analysed 160 cases of spinal cord tumour of which only eight were under 12 years of age. In Elsberg's series (Elsberg, 1925) the youngest patient was 3 years old and Buchanan's youngest patient was aged 3 months (Buchanan, 1950). In the entire world literature, according to Mosberg (1951) and Elefant, Jeklerová and Lesný (1955), altogether 25 cases diagnosed during the first year of life have so far been described. Mosberg's studies have also shown that 75% of all spinal tumours diagnosed during the first year of life consist of lipomas, dermoids, teratoids or teratomas.

Ingraham and Matson (1954) divide intraspinal tumours in children into three groups: (1) congenital tumours, ranging from simple dermoid cysts to highly variable tridermal teratomasi; (2) intramedullary gliomas, including astrocytomas, ependymomas, medulloblastomas and multiform glioblastomas; (3) extradural extension of paraspinal lesions, including neuroblastoma, reticulum cell sarcoma and lymphosarcoma.

Omitting cases of teratoma, dermoid and tumours associated with developmental anomalies such as spina bifida, meningocele or sinus pilonides, the present authors were unable to find in accessible

literature more than two cases of pure spinal cord tumour diagnosed in the newborn (Katcher, 1952; Parkinson, Medovy and Mitchell, 1954). They therefore consider it appropriate to record the following case.

Case Report

M.L. is the second son of a mother who during the second month of her pregnancy suffered from renal colic and from the eighth month with sciatica-like pains. Normal delivery took place at a maternity hospital on June 23, 1955, of a lively baby weighing 3.580 Kg. and measuring 52 cm. in length. On the 5th day the hospital paediatrician (Dr. R. Nosková) discovered considerable flabbiness of the lower limbs, especially the left, and laxity of the joints. On the following day the condition was worse: the legs were cool, the tonus was lowered, more so on the left side where no reflexes could be elicited. The baby was transferred on the same day to the Third Paediatric Clinic for investigation.

On admission the lower limbs were cool and the soles of the feet cyanotic; Moro's reflex showed no brisk response in the lower limbs. Spontaneous movement of the right leg was weak, and dorsiflexion of the left foot absent; the calf muscles were hypotonic on both sides, more so on the left, and possibly even hypotrophic. The anus was gaping and the perineum around it formed a pouch-like bulge, thought to be due to paresis of the pelvic floor muscles, which when compressed produced a stream of urine. A barium enema and cystoradiography revealed no abnormality. Lumbar puncture on the 15th day produced only a small amount of xanthochromic cerebrospinal fluid with a light admixture of blood. Plain radiographs of the spine showed sinistroscoliosis with maximal curving of the upper lumbar portion. At this level there was also a suspected dilatation of the spinal canal which was particularly apparent in a lateral view. The arches of the 1st and 2nd lumbar vertebrae were conspicuously thin, and formed an angular kyphosis (Figs. 1 and 2). On the basis of these findings an expansive process within the upper lumbar portion of the spinal canal was suspected (neuroradiologist, J. Jirout).

On July 13, when the infant was 3 weeks old, pneumoperimyelography was carried out (Figs. 3 and 4). Following insufflation of 20 ml. of air by the lumbar route in a

FIG. 1.

FIG. 2.

FIG. 1.—Plain radiograph of spine (anteroposterior).

FIG. 2.—Plain radiograph of spine (lateral).

FIG. 3.—Pneumoperimyelogram. The air filling extends up to arch of L1 (marked by arrow).

FIG. 4.—Pneumoperimyelogram. On slightly raising pelvis the air filling reaches upper border of arch of L1 (marked by arrow).

FIG. 3.

FIG. 4.

lying-down position, the lateral view radiographs revealed an air filling in the sacral and lumbar portions extending cranially up to L2. This is in keeping with pressure changes above L1 (J. Jirout).

At the same time a sample of cerebrospinal fluid was withdrawn and examined: the fluid was blood-stained and, after centrifuging, mildly xanthochromic, with 103 lymphocytes and numerous erythrocytes. Pandy's reaction was + + + +, Ross Jones + + +, Nonne Appelt + + +, and the protein 0.86 g. per 100 ml. A radiograph of the lungs showed a light shadow over the right cardiophrenic angle. The baby was afebrile throughout.

At a neurological examination on July 15 it was found that since the previous examination the movement of the left leg as well as the tone of the calf muscles of both sides had improved. Spontaneous mobility of both lower limbs was symmetrical, though in suspension the right lower limb was more feeble. The reflexes of L5-S2 were absent on both sides. The perineal bulging continued on both sides while the anus was somewhat more gaping than a week earlier. It was decided that the findings were consistent with compression of the cauda.

Improvement in the mobility of the lower limbs was observed after the first lumbar puncture. Since, during a second lumbar puncture, a quantity of dark blood had escaped, a haemangioma was believed to be present, which diminished in size after puncture and so exerted less pressure on the cauda equina. Laminectomy of L1-L2 was recommended.

The baby was operated on at the age of 28 days on July 22, 1955, at the neurosurgical department of the Střešovice Hospital in Prague, and laminectomy of L1-L2 was performed under local anaesthesia. Beneath the arches a bluish violet tissue with a delicate capsule was found, which at first gave the impression that a haematoma beneath the dura mater was present. On closer examination, however, it was found to be neoplastic tissue which it was possible to remove gradually with comparatively little haemorrhage. It had grown practically across the entire antero-posterior dimension of the spinal canal. Not until the greater part of the tumour had been removed could it be established that the pouch of the dura mater was pressed forward and to the left, whereas the remainder of the canal was filled by a tumour which extended even under the arch of T12. The latter arch was also removed but the tumour reached even higher. In the lower part of the tumour there was a cyst the size of a pea filled with a greyish yellow caseous substance. The upper part of the tumour was penetrated through and through by one of the nerve roots, as was confirmed by the response to traction. In view of the extent of the laminectomy and the fact that the baby began to breathe more quickly and the pulse rate to rise, it was decided to discontinue the operation even though the entire tumour had not been removed.

Histological examination of the excised mass was carried out by M. Vorreith who found portions of strongly haemorrhagic tumorous tissue and nerve ganglion. The tumour cells were comparatively small, mostly rounded in shape with a dark nucleus and a

well-defined basophilic cytoplasm. In some parts between the islets of cells there was a fibrillary structure, pointing to the neurogenic nature of the tumour. In numerous places the tumour cells constituted rosette-shaped formations. In the portions broken up by haemorrhagic areas there were occasional leucocytic infiltrations and foci of siderophages. Mitoses were not found. He concluded that the findings were consistent with neuroblastoma.

Following the operation the baby was in good condition, drank well and gained in weight. From early in July to early August, 1955, the baby was given radiotherapy: 100r were administered in each of 26 sittings by V. Stašek. During this treatment only once was there a change in the blood count, the erythrocytes falling to 3,760,000, the haemoglobin decreasing to 60% and immature white cells making their appearance.

Before discharge the baby was re-examined neurologically. Signs of affection of the cauda were still present, more so on the left side, but compared with the findings before operation there was pronounced improvement on both sides, especially at the right. Before discharge the baby was thriving satisfactorily and at the age of 3½ months weighed 5.31 Kg.

Of the laboratory and other investigations we record the following results: the blood group was A rh; the W.R. negative; an electrocardiogram revealed tachycardia, but was otherwise normal; a radiograph of the lungs on August 5, 1955, showed elevation of the right diaphragm.

The baby was readmitted for six days on January 22, 1956. The ocular fundi were normal. The blood picture showed 4,490,000 erythrocytes, 74% haemoglobin, 8,300 leucocytes; polymorphs with rod-shaped nuclei 4%, with segmented nuclei 36%; lymphocytes 59%; and monocytes 1%.

At 7 months of age the flaccid peripheral paresis of the lower limbs was still present but the sphincter signs were much improved. As against the earlier picture it was now the right lower limb that was the more affected of the two. A radiograph of the skeleton showed no destructive changes, nor evidence of metastases, and there was no scoliosis; only at the sites of the laminectomy the vertebral bodies were more translucent. At home exercises were being carried out with the baby according to instruction.

The third stay in hospital at the age of 11 months took place between May 25 and June 16, 1956. The baby weighed 10.3 Kg. and measured 76 cm. The circumference of the head was 47 cm., and of the chest 49 cm. The baby had sat up without support from the age of 10 months, was standing and could already walk holding on to the cot. Dentition began at 8 months, speech at 10 months. By this time the anterior fontanelle was closed, the occiput firm, and the operation scar in the thoracolumbar region of the back well healed. The blood picture continued to be normal. A radiographic examination of the chest showed elevation of the central portion of the right diaphragm, while one of the skeleton showed no metastases, and normal ossification of the long bones. The ocular fundi were normal.

At the age of 12 months a further neurological examina-

tion showed peripheral hypotonia of the left lower limb. There was a slight pes valgus on the left. When the left lower limbs was moved spontaneously extension of the foot was carried out incoordinately by means of the extensor muscles of the toes; tibialis anterior responded well to stimulation and showed normal tonus. Patellar reflexes were increased on both sides. The plantar reflexes were of the extensor type. The reflexes of L5-S2 were bilaterally decreased. There was a physiological lumbar lordosis. With support the baby stood comparatively firmly, the centre of gravity being transmitted to the right leg. Micturition was streamlike, the flow of urine was promptly halted and did not dribble away. The anal sphincter was well-contoured though slightly enfeebled; the anal reflex could not be elicited. As far as sensation was concerned, there was a loss of pain sensibility on the right side distally from L4, on the left from L5.

Since the abnormal neurological findings were receding, no further radiotherapy was recommended for the time being. The myelogram showed no tumour cells and the marrow was normal.

The baby began walking without support at 18 months. It was last examined at the age of 20 months, when it weighed 12.75 Kg. It had one regular daily stool and micturition was normal. Its mental development was in keeping with its age. The sensory disturbance in the region of S2-S3 persisted as well as mild hypotonia of the right S1-S2 segments but otherwise the neurological findings had almost returned to normal.

Discussion

It is not intended to deal here with the clinical entity of spinal cord compression as such, which in childhood presents numerous features in common with compression in the adult. It should, however, be recalled that, just as in intracranial tumours, intraspinal expansive processes have their syndrome of hypertension. Apart from changes in the composition of the cerebrospinal fluid (increased protein, xanthochromia), changes in its pressure and disorders in its circulation may help in diagnosis. They are an equally valuable diagnostic help in children even though in the younger ones their evaluation is more difficult because of lack of co-operation. In the younger subjects sensory and root manifestations have a greatly restricted value in determining the levels of localization since here it is a matter of estimating the approximate level of disordered sensation on the basis of the child's general reaction, elicited as painful stimuli passing from a zone of diminished or absent sensitivity, hypoaesthetic or anaesthetic, to a normal or hyperaesthetic one.

On careful examination of plain radiographs of the spine in cases of cord compression, dilatation of the spinal canal, occurring especially in children and confined to a few segments, cannot be missed.

Note is taken of the interpedicular distance, flattening of the inner surfaces of the pedicles or their possible disappearance. The changes are greatest at the level of the tumour, below which they decline in magnitude. Another important feature is the change in the normal shape of the spine. At the site of the expansive process the spine is altered in shape over a small area, being straightened as if it were stiff, or there may be a scoliosis confined to a brief segment of the spine (Jirout, 1956). Impressions of the posterior wall of the vertebral body and dilatation of the intervertebral foramina are diagnostically as valuable in childhood as they are in adults. An accurate spatial picture of the boundaries of the expansive process may be obtained with the aid of pneumoperimyelography, which is kinder to the patient than iodine perimyelography.

The authors' own experience as well as that recorded in the literature (Ingraham and Matson, 1954) shows that the most frequent erroneous diagnoses in cases of spinal cord compression are poliomyelitis or amyotonia congenita.

The symptomatology of our present case included a feature which we have not so far come across either in our experience or in the literature. This was paresis of the pelvic floor manifesting itself by a pouch-like bulge, in the differential diagnosis of which a cystic formation within the pelvis had to be excluded.

In view of the clinical signs of compression of the cauda equina and the corresponding changes in the radiographs of the spine, together with the results of pneumoperimyelography, the upper boundaries of the compression were not searched for. Prompt improvement of mobility following operation, even though the entire tumour had not been removed, justifies the belief that it did not extend higher than one segment above that reached at operation. There is reason to believe that early operative intervention on the 28th day of life—the patient being perhaps the youngest ever subjected to such an operation—was responsible for the remarkable disappearance of the clinical signs and the fact that the child is now free from gross motor disability.

The effect of the radiotherapy cannot be unequivocally evaluated in the light of the latest findings on the spontaneous remissions of the malignancy in neuroblastoma (Cushing and Wohlbach, 1927; Lehmann, 1932; Farber, 1940). Gross (1953) as well as other workers have found that neuroblastoma treated before the age of 2 years has a more favourable prognosis than those diagnosed later.

Neuroblastoma, one of the most frequent malignant tumours in childhood, originates primarily in paraganglionic areas but often penetrates into the

spinal canal via the intervertebral foramina. It is a highly vascular, fairly soft, cellular tumour; it never, however, penetrates the dura mater or nerve sheaths. Its aetiology and pathogenesis are not clearly understood. If signs of rapid progression of the cord compression are present it would be wrong to await the effect of irradiation or other conservative treatment. In such cases laminectomy with complete excision of the intraspinal tumour is called for as soon as possible. Such treatment with subsequent radiotherapy leads to functional restoration. Extensive laminectomies in small children may lead to spinal deformities (lordosis, kyphoscoliosis). By combining surgical measures with radiotherapy, Wittenborg (1950) found that 30% of patients with neuroblastoma survived over three years. In general, however, and with rare exceptions—a survival of 15 years (Lehman, 1932) and one of 17 years (Oberkircher, 1953)—the disease ends fatally within one year. It is noteworthy that Lehman's case was not treated radiologically at all.

Summary

The literature is surveyed on spinal cord compression in children in general and in infants and the newborn in particular.

A case of intraspinal neuroblastoma is described, which was diagnosed in a newborn baby on the eighth day after birth, and treated on the 28th day by laminectomy with partial extirpation of the tumour and post-operative radiotherapy.

A follow-up at the age of 20 months showed very good results from the surgical treatment and possibly also from the post-operative radiotherapy. The child walked safely without support and there were no signs of incontinence or metastases.

The baby was re-examined at the age of 34 months and its neurological condition was found to be unchanged.

To our knowledge it is the third case of pure intraspinal tumour in the newborn recorded in world literature.

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