Case reports

Association of ganglioneuroblastoma with syringomyelia

Case report

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Summary

The association of a syndrome of a degenerative nature such as syringomyelia and a neuroblastoma can be of clinical interest. We will describe the case of a young female suffering from a retroperitoneal neuroblastoma and secondary development of syringomyelia.
The possible pathogenetic link between these two pathologies will be discussed. Firstly, the dysontogenetic interpretation will be underlined. Other hypotheses will concentrate on the presence of tumors within the cord, which tend to cavitate, and furthermore, on the association between the edema and some diseases such as neoplasma, traumas, and on arachnoiditis as a major pathogenetic factor in syringomyelia. The existence of a possible link between arachnoiditis and the radiotherapy received by the patient after the surgical excision of the retroperitoneal neuroblastoma, will be discussed.
Lastly a further pathogenetic hypothesis will be pointed out: an intramedullary softening due to disturbed blood supply, caused by the extramedullary neoplasm.

Key words: Syringomyelia, Neuroblastoma, Arnold Chiari Malformation, Computed Tomography, Nuclear Magnetic Resonance

Introduction

The association of a syndrome of a degenerative nature such as syringomyelia and a neuroblastoma was only once described in literature, in an old description of a thirty-six-year-old woman with von Recklinghausen's disease, a neuroblastoma in the fourth ventricle and syringomyelia[1].

The syringomyelic syndrome consists in a central cavitation of the spinal cord owing to undetermined causes. A similar clinical syndrome is sometimes observed in other pathological conditions such as intramedullary cord tumors, traumatic and post-radiation myelopathy, spinal arachnoiditis, infarction (myelomalacia) and bleeding (hematomyelia) and yet rarely with extramedullary tumors, cervical spondylosis and cervical necrotising myelitis[2].

Among the existing classifications of syringomyelia the most accepted is that recently proposed by Barnett *et al.*[3].

Neuroblastoma is the second most frequent tumor in children. It is the only childhood tumor which has such a wide range of malignancy: from quite mild to very severe. The prognosis varies from excellent, for patients with localized tumors, to very poor for those with bone metastasis. In some patients the tumor regresses spontaneously[4].

We will describe a young female patient suffering from a retroperitoneal neuroblastoma and secondary development of syringomyelia. The possible link between these two pathologies will be discussed.

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Fig. 1. Centrally located liquordense cavity in the upper cervical spinal marrow and medulla oblongata together with a slight ectopia of the cerebellar tonsils.

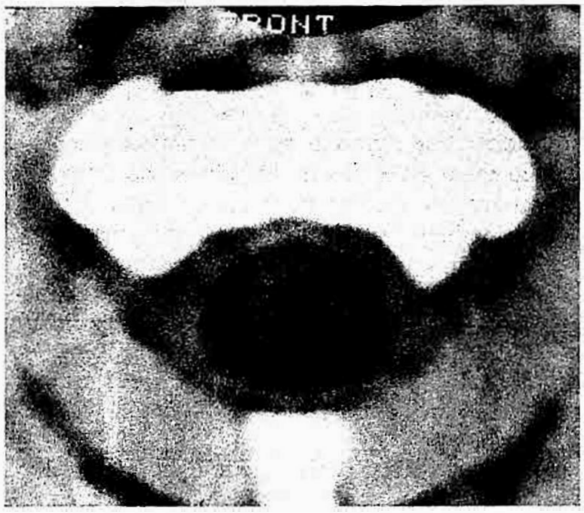

Fig. 2. Computer assisted myelography: syrinx from the upper cervical spinal marrow and medulla oblongata without connection to the 4th ventricle.

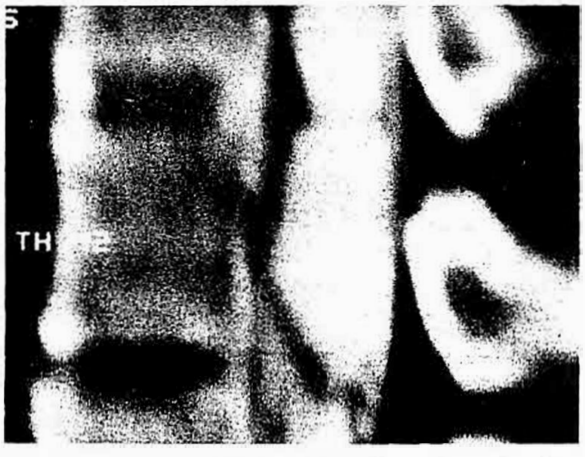

Fig. 3. Computer assisted myelography: syrinx extending

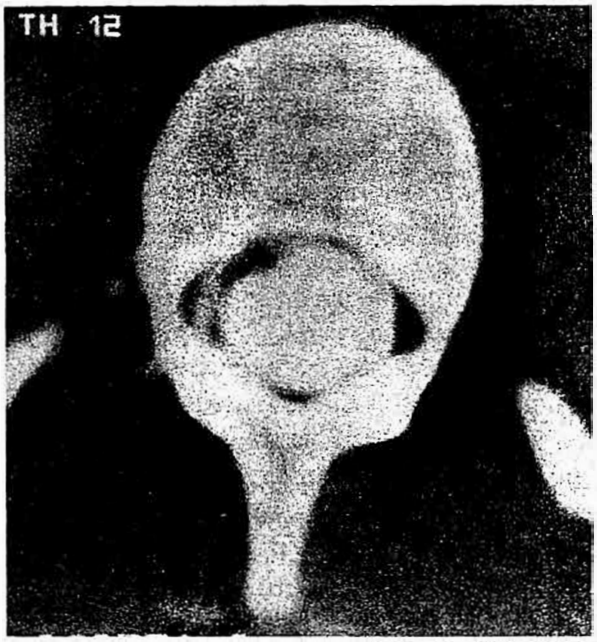

Fig. 4. Computer assisted myelography: communication with the subarachnoidal space at the level of Th 11/12.

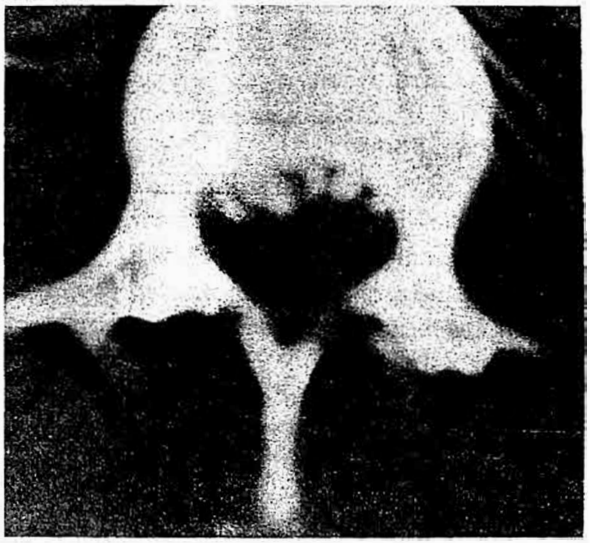

Fig. 5. Computer assisted myelography: extradural tumor at level L1/L2/L3/L4.

Case report

A 14-year-old girl was admitted with a history of acute hemiparesis on the right side, preponderant in the lower limb area and with hypoesthesia on the same side. She had been complaining of paresthesia of the right hand for one month.

The mother recalled a normal pregnancy, but delivery by forceps was followed by cyanosis.

with the exception of gait disturbances since infancy.

At the age of 5 a silent abdominal mass was diagnosed as being a retroperitoneal non-metastatic ganglioneuroblastoma, located at the level of T 12-L 4 and displacing the surrounding structures. The tumor measured 9.9 cm and was partially calcified and solitary. A dolichocolon represented an accidental finding in the diagnostic work-out.

Neurological examination showed an areflexia in the lower limb area and a bilateral peroneal weakness.

Neuroradiological examination demonstrated an erosion of the vertebral bodies at levels L2 and L3, an enlargement of the foramina intervertebralia at the levels L1/L2, L2/L3, L3/L4, and a blockage of contrastproduct at level T12 and L4.

Surgical excision of the ganglioneuroblastoma was incomplete due to invasion of the intervertebral foramen L2/L3 and was therefore followed by radio- and chemotherapy. the former consisted of telecobalt with a total dose of 3000 rad. directed at the upper right abdomen and the latter of a combination of vincristine and cyclophosphamide. A control myelographic examination revealed a free passage of the contrastproduct.

At the age of 10, the patient underwent surgical correction of a claw foot on the left, of which there was no relevant family history. On admission, the physical examination, which was performed paying particular attention to dysplastic features, was normal. The neurological examination evidenced a segmental weakness (without atrophy) of the upper right limb, concomitant hemisensory loss on the upper limit C3 and cerebellar signs on the right side. The myotatic reflexes were more vivid on the right side and were accompanied by pyramidal signs. In the lower limb area a peroneal weakness, hypotrophy and a slight weakness of the M. triceps surae were found bilaterally. Position sense was disturbed and the myotatic reflexes were absent bilaterally. The plantar reflexes were found to be in flexion.

Routine laboratory examination results were normal including the presence of catecholamines in the urine after 24 hours. Examination of CSF showed normal values. The EEG was

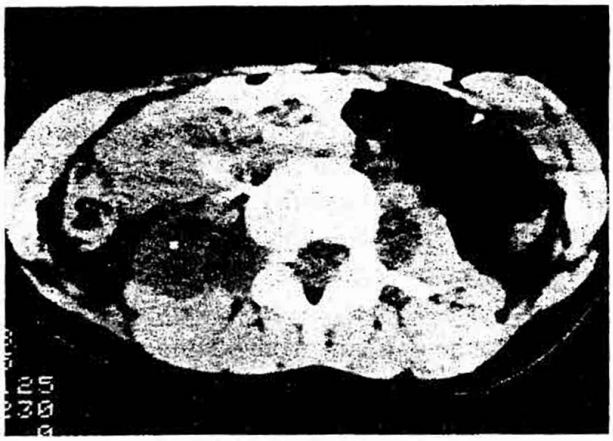

Fig. 6. CT scan of the pelvis: at the level of the foramina intervertebralia L2-3 bilaterally and L3/L4 on the right the tumor extended in a lateral direction into the paretebral space.

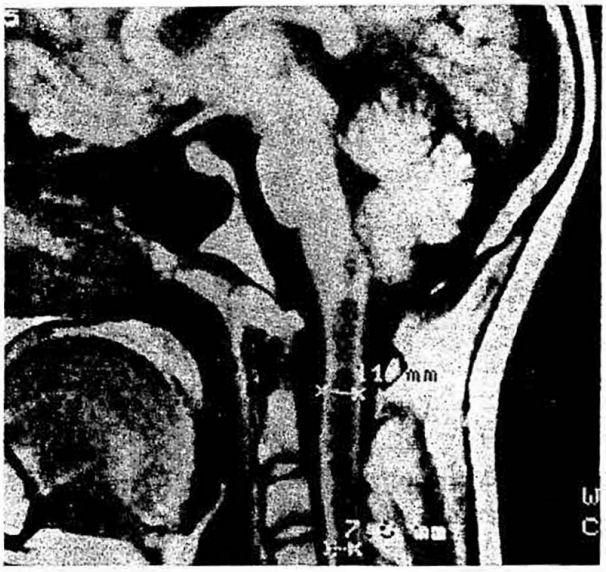

Fig. 7. Nuclear Magnetic Resonance scan of the spinal column: extensive syringohydromyelia with sagittal diameter of 7.5 mm to 11 mm.

normal: the somatosensorial evoked potentials showed a decrease in conduction time on the right side. X-rays of the cervical and thoracal spinal column and the skull disclosed right cervical skoliosis. A CT scan of the brain and craniocervical transition without contrast-enhancement revealed a centrally located liquordense cavity in the upper cervical spinal marrow and medulla oblongata, together with a slight ectopia of the cerebellar tonsils reaching the inner border of the foramen magnum (Chiari malformation type 1) (Fig. 1). A computer assisted myelography was performed after administration of 8 ml jopamiro 200 intrathecally and 100

ml of jopamiro 300 intravenously.

The latter examination disclosed a syrinx from the upper cervical spinal marrow and medulla oblongata without connection with the 4th ventricle (Fig. 2), which extended until the level Th 9/10 and communicated with the subarachnoidal space at the level of Th 11/12 (Fig. 3, 4). A homogenous contrast-enhancing extradural tumor began at level L1, narrowing the spinal canal from the right and from behind, producing a subtotal obstruction at level L2 and extending downwards to end above the disc at level L3/L4. (Fig. 5). At the level of the foramina intervertebralia L2/L3 bilaterally and L3/L4 on the right the tumor extended in a lateral direction into the paravertebral space. Here it reached a sagittal diameter of 4 cm and a craniocaudal length of 5 cm, as can be observed in the CT scan of the pelvis (Fig. 6). A Nuclear Magnetic Resonance (NMR) scan of the spinal column showed extensive syringo-hydromyelia, without communication between the latter and the 4th ventricle. (Fig. 7, 8).

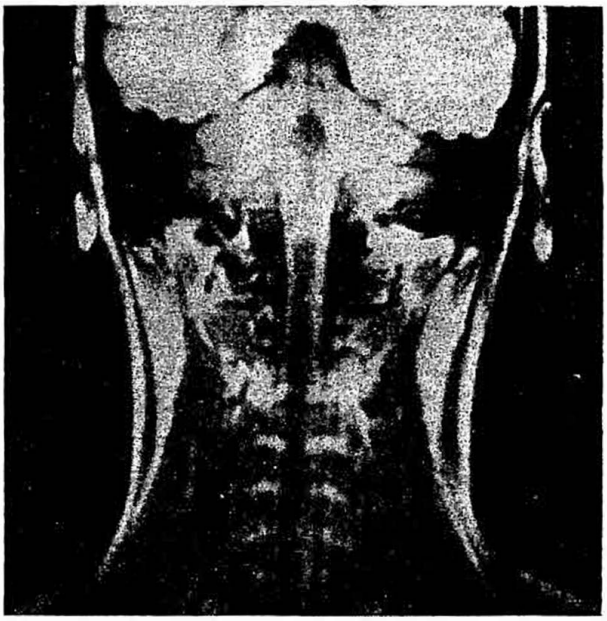

Fig. 8. Nuclear Magnetic Resonance scan of the spinal column: extensive syringohydromyelia, without communication between the syrinx and the 4th ventricle.

Discussion

As regards the diagnostic possibilities offered by the NMR and CT scans, the former allowed better visualization of the syringomyelic cavity, whereas the latter gave a better picture of the non-communication between this cavity and the ventricle by means of the stop of the contrast medium (Fig. 2). As compared to the NMR scan, the CT scan with contrast-enhancement was more successful in demonstrating the site, the extension and the root compression of the extramedullary tumor. Furthermore the CT examination showed the communication between the syringomyelic cavity and the subarachnoidal space, not revealed by the NMR scan.

As far as a dysontogenetic interpretation is concerned, it is interesting to emphasize that a dolichocolon was accidentally discovered in our patient, who already manifestated an Arnold Chiari malformation. One basic developmental abnormality has already been proposed as a possible pathogenetic factor of the association of syringomyelia with intramedullary neoplasms[5,6,7]. Moreover, the etiological relation between the neuroblastoma, being an embryon-

with von Recklinghausen disease, together with the syringomyelia in our case, could be emphasized. It is also well known that the neuroblastoma can be associated with congenital anomalies, such as spina bifida, coarctation of the aorta, hydrocephalus, polydactylia and various malformations.

The fact that our patient had suffered a traumatic birth is also of some importance, since in 24% of patients with syringomyelia, delivery had been traumatic[8].

With regard to the pathogenesis of syringomyelia, Gardner's theory suggests that a congenital failure of the outlets of the 4th ventricle leads to an abnormal flow of CSF, with the consequent dilation of the central canal and secondary development of ramifying diverticulum and of a syringomyelic cavity[9,10].

In our case the evidence of a non-communicating syringomyelia does not contradict this pathogenetic interpretation, since the cavity could initially have been connected to the central canal and then have become blocked at a later stage[11].

Ball and Dayan suggested that in the presence of a subarachnoid obstruction at the craniocervical junction, the cavitation of syringomyelia

sure, into the spinal cord from the subarachnoid space along perivascular spaces (Virchow-Robin). This fluid slowly coalesces to form a unique cavity, which might at first not be connected to the central canal, but eventually become connected to it[12].

Another hypothesis for syrinx development is the presence of a blastomatous formation in the spinal cord, such as tuberous sclerosis or central von Recklinghausen disease, in which there is a tendency for the abnormal tissue to cavitate. Spontaneous autolysis of the tumor or hemorrage into the cord from the tumor has been suggested by some authors[13]. Particularly in our case, the association of the neuroblastoma with a central von Recklinghausen disease, could be hypothesized.

Finally, Feigin *et al.* have pointed out that edema could be a major pathogenetic factor in syringomyelia in cases of angiomatous malformations, neoplasms, traumas, arachnoiditis and other diseases that cause edema[14].

Moreover in our case, it should not be forgotten that the young patient had received radiotherapy (3000 rad.) after the surgical excision of the retroperitoneal neuroblastoma.

Vascular alterations and phlogistic manifestations such as arachnoiditis processes consequent to radiation treatment might be of particular interest in our case in order to explain the association of these two diseases.

Although the association of a neuroblastoma with syringomyelia was only once described in literature, a case report of the association of cutaneous neuromelanosis with syringomyelia has already been described by Leany[15].

More recently a group of authors reported a case of syringomyelia and syringobulbia associated with an ependymoma of the cauda equina involving the conus medullaris[16]. The authors reviewed several possible explanations for this association, the most interesting of which appears to be intramedullary softening due to disturbed blood supply, secondary to the associated neoplasm[17].

As already suggested by Ball and Dayan[12], the Arnold Chiari malformation could represent, in our case, a functional obstruction, due to the 'tethering' of the cord by dense adhesions at the site of the tumor.

We believe that the report of this case could offer some suggestions as to the pathogenesis of syringomyelia.

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