Hydrodynamic Studies in Syringomyelia

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Although intramedullary cavitation of the spinal cord was recognized earlier, the term “syringomyelia” was first used by Oliver d’Angers in 1837 for cavities within the cord, regardless of etiology. Using the term in this manner, Schlesinger in 1902 noted the occurrence of intramedullary cavities in relation to trauma, inflammation, vascular disorders, tumors, malformations of the nervous system, pachymeningitis, and leptomeningitis, while in other cases he could identify no associated pathology. Holmes, Collier, and Schneider have studied the role of trauma in causing central cord cavitation. Tauber and Langworthy reported cavities associated with arterial insufficiency. McLaurin, et al., have experimentally produced cavitation secondary to adhesive arachnoiditis. Netsky has suggested arterial pathology as a basic cause of idiopathic syringomyelia, although, in general, the thickening and hyalinization of the blood vessels in the walls of syringes has been regarded as a secondary reaction, possibly due to hydrostatic pressure within the cyst.

There is a high incidence of syringomyelia in association with intraspinal neoplasms. Kernohan, et al., and Poser reported an incidence of 53.5% and 31% respectively. This association has been ascribed to necrosis within the tumor, obstruction of vital arterial and venous channels by the tumor, or transudation of fluid from such neoplasms as hemangioblastomas. Poser in his recent exhaustive review concluded that both the syrinx and tumor result from abnormal glial and mesodermal elements included in the cord as a result of faulty closure of the dorsal raphé of the neuraxis during embryonic development.

Syringomyelia is often present in cases of dysraphic and dysplastic phenomena within the central nervous system, such as meningomyelocele, duplication of the spinal cord, and the Arnold-Chiari malformation, and in the body as a whole in cases of skeletal abnormality, asymmetry of the two sides of the body, genitourinary abnormality, and intestinal duplication. Accordingly, syringomyelia is often regarded as part of the syndrome of “status dysraphicus” of Bremer.

Mackay and Favilli considered abnormal glial proliferation followed by degeneration and cavitation to be the primary pathogenesis of spinal cord cavitation. Hassin suggested that the process was primarily one of degeneration, defective glial undergoing atrophy. Taylor, et al., concluded that disintegration of congenitally unstable glia at the union of the embryonic basal and alar plates of the primitive neuraxis explained the distribution of the cavities in the cord and brain stem.

Recognizing the high incidence of other malformations in association with syringomyelia, many investigators have questioned whether spinal cord cavitation is a part of the dysplastic process or whether it is due, partially or completely, to cerebrospinal fluid hydrodynamic changes imposed by the other malformations.

Taylor, et al., quoted Grund (1908) as being the first to consider cerebrospinal fluid pressure as an important factor in the formation of syringomyelia. Taylor, et al., (1922) suggested that pressure might be a contributing factor. They described the clinical course and pathological findings of two patients. One had a progressive course and evidence of hydrocephalus at postmortem examination. The second patient, 4½ years after the onset of his symptoms, subsequently had no further deterioration during 24 years of further observation. The patient committed suicide by hanging. Postmortem examination demonstrated a rupture of the syrinx into the subarachnoid space, presumably at
the time of the arrest of symptoms. The authors felt that the specimen showed evi-
dences of previous hydrocephalus and previous increased pressure which were also
presumably relieved at the time of the sponta-
neous rupture of the syrinx.

It has been established that, in 71%\textsuperscript{10} to
80%\textsuperscript{26} of normal subjects, the central canal of
the spinal cord closes and becomes a vestigial
structure represented by a core or nests of
ependymal cells. Cameron\textsuperscript{5} regarded the
majority of the spinal cord cavities occurring in
dysraphic states as due to the delayed
closure or persistent opening of the lower
neuraxis (meningomyelocele). He believed
that, during embryogeny, the cerebrospinal
fluid ran down the persisting central canal
and out the defect, thus dilating the canal
(hydromyelia).

Syringomyelia has frequently been de-
scribed in association with hydrocephalus due
to blockage of the outlets of the fourth ven-
tricle, with or without the Arnold-Chiari
malformation.\textsuperscript{1,2,7,15,18,25,35} In other reports,
the cause of hydrocephalus has not been
apparent.\textsuperscript{36,47}

Gardner\textsuperscript{15–19} considered the Arnold-Chiari
malformation, diverticula and cysts of the
outlets of the fourth ventricle, and the
Dandy-Walker syndrome to be varying ex-
pressions of embryonal atresia of the fourth
ventricle. He reported that such malforma-
tions were found in each of his 74 cases of
syringomyelia in which the posterior fossa
was explored.\textsuperscript{19} Some cases had hydrocepha-
lus due to blockage of all outlets of the
fourth ventricles, but others had normal
ventricular volume and were felt to have only
the midline foramen of Magendie embar-
rassed.\textsuperscript{17} Gardner hypothesized that blockage
of this foramen prevented free egress into
the subarachnoid space for the fluid pulse
waves initiated by the pulsating choroid
plexus. Thus, the pulsation was diverted down
the central canal of the spinal cord, leading
to cavitation of the cord. This produced
hydromyelia if the cavity was contained by
ependyma, or syringomyelia if it broke
through the ependymal lining and dis-
sected into surrounding nerve tissue. Gard-
ner stated that this entity accounted for
the majority of cavities within the spinal
cord. He recommended the surgical treat-
ment of opening the foramina of the fourth
ventricle and blocking the connection be-
tween the fourth ventricle and the spinal
cord cavity.

There is no unanimity of thought as to
either the etiology or the best therapy for
spontaneous cavitation within the spinal
cord. Radiation therapy is used by those who
regard the basic pathology to be glial prolif-
eration.\textsuperscript{13,27,37,40} Others drain the cyst
directly, presumably regarding the syrinx to
be isolated and under tension.\textsuperscript{15,27,29,36,50,53}
Both methods have met with some success.

In cases where the cavity is connected with
the ventricles, Gardner’s method has merit.
The purpose of our study was to find criteria
by which to diagnose cases of this type pre-
operatively.

Surgical therapy to correct the proposed
hydrodynamic problem had been undertaken
in over half the cases. We have used the term
“syringomyelia” to indicate a proven or
presumed cavity in the spinal cord regardless
of etiology, with the exception of an acute
central cord syndrome secondary to trauma,
which we have excluded. Proven cervical
intramedullary tumors will be discussed
separately.

\textbf{Material}

Between 1958 and 1962, 17 cases with a
spontaneous progressive central syndrome
of the cervical cord were seen on the neuro-
surgical service at the University of Cali-
ifornia at Los Angeles. During the years
1963 to 1966, five additional cases were seen
at the Medical College of Virginia.

Of this total of 22 patients, two were
initially diagnosed as having intramedullary
cervical cord tumors which subsequently
were proven to be Grade 1 astrocytomas,
one cystic and one probably solid. Of the
remaining 20 patients, it was possible to
study 12 in the manner listed below (Table
1). Six of these underwent posterior fossa
exploration and cervical laminectomy of
C-1 through C-4 and were found to have
cysts of the cervical cord in connection with
the fourth ventricle. A seventh patient had
had a cervical laminectomy 10 years pre-
viously; at that time, clear fluid was aspi-
rated from a cyst in the cord but the pos-
terior fossa was not visualized. The remain-
ing five had similar radiological findings and
appear to represent similar pathology. The