Congenital Cervical Ependymal Cyst
Report of a Case with Symptoms Precipitated by Injury*

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Congenital intradural extramedullary ependymal cysts of the spinal canal are rare. In recording this additional instance of ependymal cyst of the cervical spinal canal we are focusing attention upon the diagnostic significance of associated bony defects of the cervical vertebrae as the stigmata of an embryonic developmental malformation. We are also emphasizing that the symptoms and signs of cervical spinal cord and root compression were precipitated by injury to the neck.

Case Report

The patient was a white 17-year-old man, who had been well until he suffered an injury of the neck in an automobile accident early in September, 1962. Immediately thereafter he complained of severe pain in the neck associated with radiation of pain down the right arm. There followed numbness of the right arm and leg. Roentgen studies on October 6, 1962, revealed fusion and narrowing of the bodies of the 3rd, 4th and 5th cervical vertebrae with an associated dilatation of the spinal canal in this region (Fig. 1). There was also a failure of fusion (spina bifida) of the spinous processes of the 7th cervical and 1st thoracic vertebrae (Fig. 2). Conservative treatment failed to alleviate the symptoms and the patient was admitted to the Montgomery Hospital, Norristown, Pennsylvania, on December 2, 1962.

Examination. At this time examination disclosed a stiff neck with limitation of movement, and insensitivity to pinprick on the right side of the body. The patient complained of pain in the neck and pain over the area of both trapezius muscles. The remainder of the neurological examination was normal. A myelogram (Figs. 2 and 3) showed a block in the region of the 3rd and 4th cervical vertebrae with widening of the canal in the same region.

Operation. Laminectomy was performed by Dr. Joseph Brady on December 14, 1962. The spinal canal was considerably widened and when the dura was opened the cord was found to be flattened by a gray smooth-suraced cyst, 2 cm. in maximum diameter. The cyst extended from the level of the inferior border of the 3rd cervical vertebra to the 4th cervical vertebra and lay ventral to the cord. It appeared to be attached to the "ventral aspect" of the cord between C3 and C4. A needle inserted into the cord proper yielded no fluid. Some of the fluid withdrawn from the cyst appeared turbid and contained numerous flecks of grayish material floating in it. The cyst was excised down to the pedicle which was attached ventrally to the cord; in order to avoid damage to the latter a small stump of cyst wall was permitted to remain. The patient recovered uneventfully from the operation and when last examined in December, 1964, he was working full time but still complained of some numbness of the right arm and leg.

Histological Study. The histological appearance of the cyst membrane showed it to consist mainly of connective tissue which contained a few lymphocytes and macrophages containing hemosiderin scattered through the tissues. The cyst wall was composed of an inner column of epithelial cells, the stroma of which was composed of a connective tissue matrix containing numerous macrophages and occasional fibrinoid material. A section of the proximal stump of the spinal cord showed an acute inflammatory leukemic meningitis with lymphocytes and macrophages in marked abundance. The spinal cord was atrophic and flattened; the gray matter was slightly reduced in thickness and there were a few gliomatous areas. A section through the cyst wall showed the cyst fluid to be a bloody turbid liquid containing numerous grayish flecks.

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Fig. 1. Note fusion of the bodies of C3, 4 and 5, with marked enlargement of the canal opposite C3 and 4.
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Among the fibers, and a lining of a single layer of low cuboidal and columnar epithelium (Figs. 4 and 5). The nuclei of these cells were centrally placed and ovoid in shape. The cytoplasm in some of the cells contained poorly-visualized small vacuoles. There was no distinct basement membrane and cilia could not be seen. The epithelium bore no resemblance to the tall columnar mucous-secreting lining of the intestinal tract but was characteristic of ependymal cells. A smear of the aspirated cyst fluid showed numerous polymorphonuclear leukocytes, a few mononuclear cells and some amorphous debris.

Discussion

At the time of operation the immediate question arose as to the origin of the removed cyst. Was it congenital in origin in view of the associated presence of bony abnormalities of the cervical vertebrae, or could it have been traumatic in view of the close relationship of the symptoms to the injury sustained in the auto accident? Austin stated that “trauma associated with hemorrhage probably has an invariable accompanying fibrosarachnitis. This is a conspicuous feature at the base of the brain in the presence of evidence of old bleeding. The arachnoid membrane appears particularly responsive to free blood with appearance of increased fibrous trabeculation. The same process undoubtedly operates in production of a chronic arachnitis in traumatized areas of the spinal cord.” We do not feel that trauma was responsible in the formation of the cyst reported here because of the immediate onset of symptoms following injury, the short duration of time between the injury and the surgical exposure of the cyst, and the size and histological appearance of the lesion. We do, however, suggest that the injury precipitated the onset of symptoms in a pre-existing intradural cyst either by causing some hemorrhage into the lesion, as is indicated by the histological appearance of hemosiderin-filled phagocytes in the wall of the cyst, or by disturbing the hydrodynamic forces in the subarachnoid space surrounding the lesion.

The presence of abnormalities of the cervical vertebrae of developmental origin contiguous to the cystic lesion indicate the probable congenital origin of the cyst itself. Hyman et al. in 1938,
The relationship of symptoms precipitated by injury in a pre-existing congenital intradural extramedullary cyst is illustrated by the case of an "intradural enterogenous cyst" reported by Harriman. This 10-year-old child fell and developed chest pains. Subsequent manipulation of the neck resulted in a complete paraplegia. X-ray studies and autopsy findings revealed no congenital anomalies of the vertebral column; however, the thickened leptomeninges were stained by hemosiderin and the cystic lesion was ventrally placed in relation to the spinal cord. The histological appearance of the lesion showed epithelial cells with goblet cells which the author described as an embryonic enterogenous cyst originating from a fistulous connection between neuro-ectoderm and yolk-sac.

Scoville et al. reported a case, which reflects the relationship of the development of symptoms following injury to the spine, of an intraspinal enterogenous cyst in a 28-year-old man. At first pain and numbness developed in the inner aspect of the left arm and subsequently he developed quadriplegia. At operation an intradural cyst was found ventral and lateral to the cord. Microscopic examination of the cyst showed it to be lined by a mucous-secreting single layer of columnar epithelium "which could come only from the endodermal germinallayer" and hence they considered it congenital in origin. The authors noted that their case was especially rare because of the "absence of the usual accompanying abnormali-

Fig. 4. Low power appearance of the cyst membrane. H. & E.; ×100.

described a ventrally placed extramedullary ependymal cyst of the cervicodorsal region of the spinal cord which also was associated with a defect in fusion of the lamina of the 7th cervical vertebra, flattening of the pedicles and dilatation of the bony canal, occurring in a 7-year-old child who had had symptoms from birth. Microscopically the cyst was lined with epithelial vacuolated columnar cells, cuboidal cells and occasional ciliated cells. The authors considered the ependymal cyst as embryonic in origin, derived from cells isolated from the floor plate of the neural tube at an early stage of embryonic development. Hoffmann's case of a 6-year-old boy was also characterized by the presence of symptoms from birth with the ultimate development of a flaccid quadriparesis. This is noteworthy because it indicates the probable congenital origin of the cyst in that case. This myelography disclosed a total block at the level of the 3rd cervical lamina and at operation a ventrally situated intradural extramedullary cyst was found lying between the upper border of the 2nd cervical vertebra and the lower border of the 4th cervical vertebra. The lesion was described as a "benign cyst lined along one margin by meningotheial cells of the low cuboidal type which are abutted upon a thin layer of fibrous tissue. In focal areas low cuboidal cells are noted lining projections of villus type. The general consensus of opinion is that this represents an arachnoidal cyst of developmental origin." In reviewing reports on the subject of intraspinal cysts, Hoffmann calls attention to the rarity of occurrence of arachnoidal cysts. His description and illustrations make it appear that this cyst was ependymal in cytological structure rather than arachnoidal.

Fig. 5. Cyst membrane showing lining cells, cuboidal and low columnar in shape, with oval to round nuclei and occasional vacuolated cytoplasm. H. & E.; ×400.
ties of spine, cord or body cavities,” and that “although congenital, it was asymptomatic for 28 years”; the symptoms and signs were precipitated by injury.

Intradural extramedullary cysts of the spinal canal may be lined by different types of cells designated as “arachnoidal cysts,” “ependymal cysts,” and “enterogenous cysts.” The cases reported in the literature of the so-called “cervical arachnoidal cysts”2,4,8 and the “ependymal cysts”5 have been in the main ventrally located. This was true of the cyst reported here which also was attached by a small pedicle to the ventral aspect of the spinal cord. The cellular structure of the epithelial lining in both forms of cysts is that of either primitive or adult ependymal cells. In all likelihood both types of these cysts are of ependymal origin. Bloom and Fawcett9 state that “in a few places, where the nervous wall is thin, as in the ventral fissure of the spinal cord, some ependymal cells span the entire distance between the ventricular and external surfaces. All of them do so in the early embryonic stages.”

We suggest that the following combination of findings may constitute a diagnostic formula useful in recognising these cysts:

1. Developmental defects of the vertebra consisting of incomplete fusion of the spinal processes, or fusion of the vertebral bodies.

2. Secondary enlargement of the bony canal due to encroachment by the cyst.

3. The onset of symptoms and signs of cord compression following an injury.

4. Myelographic evidence of a globoid lesion at the appropriate level.

Summary

We have reported the case of a 17-year-old man who developed neurological symptoms following an injury of the neck, in which bony abnormalities of the cervical vertebrae were observed; myelography revealed a complete block with a globoid shape of the medium at the level of the bony defects. At operation a ventrally placed intradural, extramedullary cyst was removed.

A combination of findings has been listed which should alert the clinician to the presence of this type of congenital cyst, whose symptomology seems to have been precipitated by an incident of trauma.

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References


