Cervical intramedullary cyst due to Corynebacterium diphtheriae gravis

Case report

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An intramedullary cyst of the midcervical region, due to Corynebacterium diphtheriae gravis, is reported. The culture and identification of the organism are documented. The specific pathological findings at the initial cervical exposure and subsequent thoracic and lumbar exposures are described.

KEY WORDS - cervical intramedullary cyst - Corynebacterium diphtheriae gravis - rotoscoliosis - adhesive arachnoiditis - spinal cord constriction and cavitation

Cervical intramedullary cysts are quite frequently seen but one caused by the diphtheria bacillus is very uncommon. Such a case is recorded. We are reporting this case because of its rare incidence, the unusual and prolonged clinical course, and the interesting specific pathological changes noted at surgery.

Case Report

This 7-year-old-boy had had an upper respiratory infection and pneumonia for 5 weeks; he was treated with penicillin and chloromycetin for 10 days, with apparent recovery. Seven days before admission, he complained of a stiff neck and drawing of the head to the left. All the muscles of the cervical and thoracic region became spastic, with a marked deformity of the spinal column resulting.

Examination. The neurological examination revealed marked muscle contractures with cervical and thoracic rotoscoliosis. No sensory level was demonstrated. Spotty hyperesthesia over the upper thoracic and lower cervical regions was noted. There was weakness of all the muscles of both arms with absent reflexes. All of the reflexes of the legs were hyperactive. There were bilateral Babinski signs and sustained ankle clonus. Laboratory studies revealed a white count of 17,400, 62 polymorphonuclear cells, 35 lymphocytes, and a sedimentation rate of 60 mm. Spinal films were negative except for the rotoscoliosis. Spinal puncture disclosed a positive Queckenstedt, 4 white blood cells, a sugar content of 62 mg%, and a total protein content of 740 mg%. A cisternal puncture was performed and 2 cc of Pantopaque introduced into the cisterna magna. This revealed a complete block at C-3. The cisternal fluid had no cells, and a protein content of 16 mg%.

Operation. Laminectomy of C3-4,5,6 was performed. The dura was tense, and when opened the cord bulged through the incision. The cord was tightly adherent to the dura, with a dense adhesive arachnoiditis. Sharp dissection was necessary to free it. As the dissection was carried out, there was more and
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more ballooning of the spinal cord through the dural opening. By palpation, one could feel a cyst cavity at the most prominent portion of the cord (Fig. 1). A No. 23 gauge needle was introduced through the median raphe, and 6 cc of orange fluid aspirated and quickly coagulated. The cyst collapsed. The opening into the cyst was enlarged with a sharp instrument. It was then possible to dissect the rest of the cord free from the meninges and establish free communication of the spinal fluid above and below. The dura was easily closed.

Pathological Examination. Study of the fluid from the intramedullary cavity revealed 150 white blood cells, many red blood cells, and a total protein of 4400 mg%. The microscopic bacteriological studies and fluid culture demonstrated that the organism was Corynebacterium diphtheriae gravis. Sections from the arachnoid showed a dense fibrous tissue. Macrophages were embedded in fibrin and exudate, with evidence of hyaline degeneration. No tissue was taken from the spinal cord.

The immediate postoperative course was uneventful. Unsustained clonus, increased reflexes in the lower extremities, and tilting of the head to the left were still noted 6 months postoperatively.

Second Operation. Seven months later, because of progressing leg weakness, a myelogram was performed, and an incomplete block found at T-4. At surgery, an extensive, dense type of adhesive arachnoiditis was present. The arachnoid fitted the spinal cord like a thin rubber glove over a finger. Areas of contusion and cavitation of the cord were noted. A torsion and traction process of the spine, during twisting and bending, may possibly have contributed to the injury of the tightly bound spinal cord.

Approximately 15 months later, the patient fell off a bicycle. Acute bleeding occurred from an area of injury at the conus medullaris. Blood filled the lower lumbar sac. Again, the constrictive adhesive arachnoiditis contributed to a torsion tear on the conus medullaris. Sections of the arachnoid indicated inflammation, with a number of macrophages and lymphocytes in a relatively dense stromal structure, along with a coarse yellowish-brown pigment characteristic of hemosiderin.

Discussion

This apparently represents the first case reported of an intramedullary cyst of the spinal cord due to Corynebacterium diphtheriae gravis. A search of the literature does reveal reports of involvement of the spinal cord by the diphtheria toxin, but no report has been found of the invasion of the cord by the diphtheria organism itself.

We think the first illness, 5 weeks before we saw him, was due to a diphtheritic inflammation of the throat, with an associated bronchopneumonia. A culture from the throat during the second hospitalization grew the organism. Also, the sensitivity test of the organisms cultured from the intramedullary cyst showed a resistance to the two antibiotics used during the acute infection at the time of the first hospitalization. During surgery the dentate ligaments were sectioned to further release the cord. A small catheter was threaded above and below without obstruction. The spinal fluid flowed freely from the upper and lower limits of the dural opening.

Walshe has indicated that the soluble toxin of diphtheria disseminates in the lymph spaces of the pia arachnoid and in those of the substance of the cord. One would assume that the organism could invade the cord in a like manner. Organisms could also reach the
cord via the blood stream. Frosch\(^1\) was the first to find the organism in the heart's blood, liver, spleen, and kidney. Howard\(^4\) reported a case of ulcerative endocarditis caused by the diphtheria bacillus. Pearce\(^7\) found the organism in cases of endocarditis, empyemia, vulvovaginitis, and unhealed superficial cuts and abrasions. Hechst\(^3\) reports diphtheria causing quadriplegia, with absent reflexes. There was some demyelinization in the peripheral nerves, and within the spinal cord. The axones were intact. Hall\(^2\) noted encephalitis in cases of extreme toxicity. Kennedy\(^6\) reported complications of mono or hemiplegia that occurred from emboli originating from cardiac involvement.

References


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