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Hypertrophic Interstitial Neuritis of Cauda Equina

—Case Report with Myelographic Findings—

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The author reports a case of surgery-proved hypertrophic interstitial neuritis of cauda equina. The pathogenesis of the unique myelographic findings is described and illustrated.

Key Words · hypertrophic interstitial neuritis · cauda equina · Déjerine Sottas syndrome · sheath of Schwann · linear radiolucency

Hypertrophic interstitial neuritis, otherwise known as Déjerine-Sottas syndrome, is a rare condition characterized by thickening of affected nerves due to hyperplasia of the sheath of Schwann[8)10)].

It may involve peripheral nerves, spinal nerve roots and the cauda equina, singly or in combination[3)6)9)]. In the absence of apparent peripheral nerve enlargement, diagnosis may depend on myelography[6)].

Although the condition starts in childhood and is progressive, the clinical manifestations are usually not apparent until later in life[9)]. Clinically, there is progressive motor and sensory neural involvement with periods of apparent remission[4)]. This report is made to stress the importance of characteristic myelographic findings in hypertrophic interstitial neuritis.

Case Report

This 41-year-old man was admitted on January 29, 1979 with the complaints of low back pain radiating to both lower extremities, hypesthesia below the inguinal region, and dysuria.

He first experienced acute onset of severe numbness and weakness of both legs and feet 20 years ago. He has since suffered several episodes, each of which has added residual numbness and weakness to a gradually increasing neurologic deficit. This has been slowly progressive and accompanied by atrophy of the legs, most marked peripherally. However, he was still able to walk with the aid of a cane. The day before this admission he lost control of bowel and bladder function.

Examination. The patient was chronically ill, wasted male. The cranial nerve functions were normal. There was generalized muscle atrophy and weakness, more marked peripherally. He was

unable to hop, walk on his right heel, and could not turn quickly. Except for knee jerk on the left which could be elicited with difficulty, deep tendon reflexes were absent in the lower limbs.

There was hypesthesia to pain and temperature below the level of T-1 dermatome, but posterior column sensaions were preserved. Median, ulnar and posterior tibial nerves were not palpated.

Routine laboratory tests were within normal limits. Lumbar puncture was accomplished with great difficulty and only 2ml of spinal fluid was obtained. The total CSF protein was 60mg/100ml. Radiographs of the lumbar spine showed mild scoliosis to the left, loss of normal lordosis, and narrowing of the interspace at L_3-L_4(Fig. 1 A and B). The interpedicular distances and the pedicles were normal.

Myelographic Findings. Lumbar puncture at L_2-L_3 resulted in a slow drip of spinal fluid. A 1cc. test dose of pantopaque was injected which collected behind lower portion of L_3 and could not move with tilt of the table. The possibility of subdural injection was considered but could not be excluded without an additional 2cc. of contrast substance. The dye column flowed reluctantly. For better visualization another 3cc. of pantopaque was injected. Tilt of the table again resulted in poor flow and the dye column encountered total obstruction with vertical, linear radiolucent streaks at the L_3-L_4 interspace(Fig. 2 A).

Repeat myelography via L_5-S_1 interspace demonstrated complete block with rapid tapering of dye column at the L_4-L_5 interspace(Fig. 2 B). Massively herniated lumbar disc was diagnosed, although a diffuse tumor or arachnoiditis were considered possible alternatives.

Operation. Exploratory complete laminectomy from L_3 to L_5 was performed. The dura was tense and non-pulsatile. On opening the dural sac, many large, swollen herniated nerve roots were seen. Each of the anterior and posterior nerve roots was approximately 4 times normal size, characteristic of hypertrophic interstitial neuritis(Fig. 3).

Postoperative Course. The patient's neurological examination was unchanged immediately after surgery. On the 7th postoperative day, he was able to walk with the aid of a cane. He gradually improved to the extent that 4 weeks after surgery he is free of pain in the lower limbs and has good sphincter control. Since his discharge he has been lost to follow-up review.

Discussion

Since the report of Déjerine and Sottas in 1893, numerous papers have appeared proposing modifications and extensions of the syndrome of hypertrophic interstitial neuritis which they described[1~3)6)7)11)12)]. The syndrome is characterized by the early onset, usually before age 20, of a slowly progressive distal motor and sensory neuropathy with enlargement of the peripheral nerves, diminution or absence of the tendon reflexes, and no signs of corticospinal tract involvement[5)]. Myelography in this condition has been previously described in 6 patients[1)6)7)11)].

The usual findings were slow flow of the pantopaque in the subarachnoid space, sometimes with a block to flow; transverse bar defects opposite the intervertebral discs; parallel linear radiolucent lines in the pantopaque column; and round filling defects in the pantopaque column, located at the level of the intervertebral foramen. The pathogenesis of the myelographic findings is directly related to the volume increase of the nerve root mass due to the sheath hypertrophy within

the limited confines of the spinal canal and intervertebral foramen[3]. As the nerve roots enlarge, they are visible as linear, parallel radiolucencies in the pantopaque column, either singly or aggregated to produce a composite wider band(Fig. 2 A). They displace the pantopaque column medially and create an increased distance between the medial aspect of the pedicle and the lateral margin of the pantopaque column(Fig. 2 B). Lewtas[7] and Hinck[6] suggest that the association of linear radiolucent defects together with the round filling defects in the pantopaque column is characteristic and should suggest the diagnosis of hypertrophic interstitial neuritis of cauda equina, even in the absence of palpable thickening of peripheral nerves. The myelographic findings in this case are interpreted as those of hypertrophic interstitial neuritis involving cauda equina which have been verified by surgery.

Summary

A case of surgery-proved hypertrophic interstitial neuritis of cauda equina is reported. The unique myelographic findings are illustrated. To the author's knowledge, no other condition produces these abnormalities.

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□ 國文抄錄 □

馬尾叢의 肥大性 間質神經炎 1例 : 脊髓造影術 所見

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池　　善　　豪

Déjerine-Sottas 症候群이라 불리우는 肥大性 間質神經炎은 Schwann 神經鞘의 增生(hyperplasia)으로 因한 末梢神經의 肥厚(thickening)를 招來하는 疾患이다.

著者는 41歲 男子의 馬尾叢(cauda equina)에 發生된 肥大性 間質神經炎 1例를 經驗한바 이를 手術에 依하여 確認하였고 그 特徵的인 脊髓造影術所見의 發現機轉을 論議하였다.

□ Explanation of Figures □

Fig. 1. Plain film roentgenograms show mild scoliosis to the left(A), loss of normal lordosis, and narrowing of the interspace at L_3-L_4(B).

Fig. 2. Myelograms reveal total obstruction with vertical, linear radiolucent streaks in the dye column at the L_3-L_4 insterspace(A) and complete block with rapid tapering of dye column at the L_4-L_5 interspace(B).

Fig. 3. Findings at operation. The greatly thickened roots filled the entire spinal canal. Each of the anterior and posterior nerve roots was approximately 4 times normal size, characteristic of hypertrophic interstitial neuritis.

□ 지 선호 논문 사진부도 □

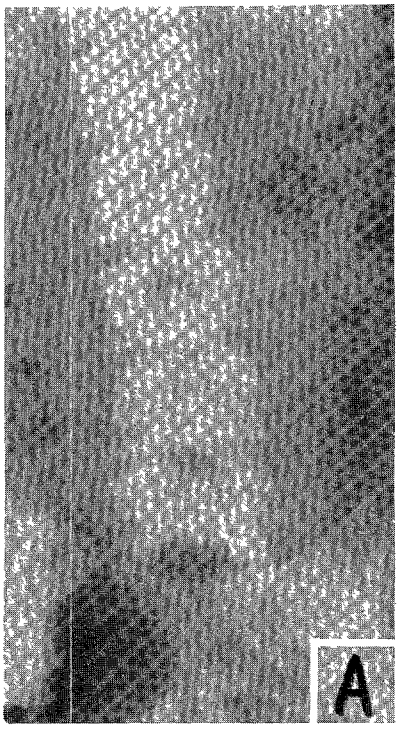

Fig. 1-A

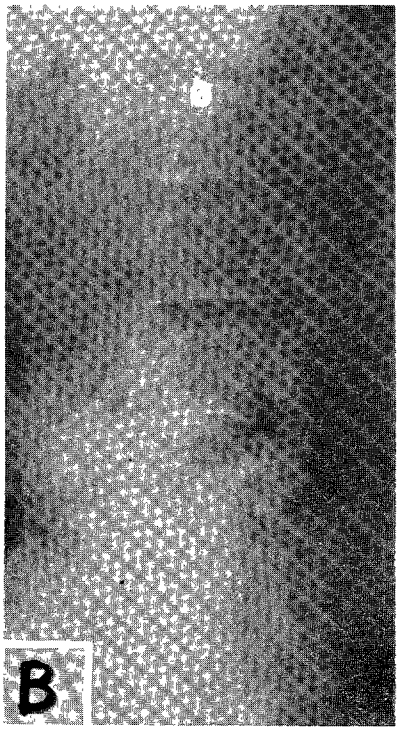

Fig. 1-B

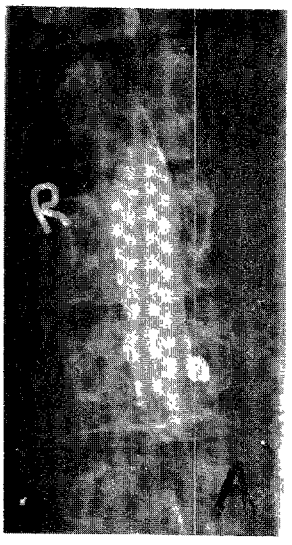

Fig. 2-A

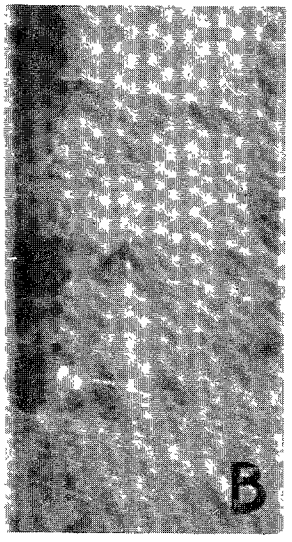

Fig. 2-B

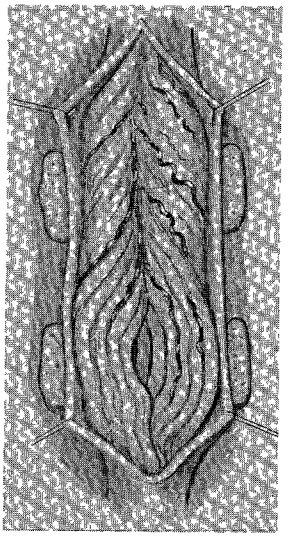

Fig 3.