TECHNICAL REPORT
29-67
業 績 報 告 書

CASE REPORTS - 1967
症例報告 — 1967年

DIPLOMYELIA
脊髄二分症

RUPTURE OF FALLOPIAN TUBE IN ECTOPIC PREGNANCY
子宮外妊娠における卵管破裂

ABCC

ATOMIC BOMB CASUALTY COMMISSION

国立予防衛生研究所－原爆傷害調査委員会

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DIPLOMYELIA, REPORT OF AN AUTOPSIED CASE
脊髄二分症の1剖検例

YOHEI II, M.D.（ 井 洋平 ）; DRAKE W. WILL, M.D.; AKIRA SANDA, M.D.（ 三田 明 ）

RUPTURE OF FALLOPIAN TUBE IN ECTOPIC PREGNANCY COMPLICATING USE OF INTRAUTERINE CONTRACEPTIVE DEVICE, REPORT OF TWO AUTOPSIED CASES
子宮内避妊器具使用の婦人に起こった子宮外妊娠における卵管破裂，2症例の報告

YOSHIAKI TSUKADA, M.D.（塚田義明）

ABCC
ATOMIC BOMB CASUALTY COMMISSION
HIROSHIMA AND NAGASAKI, JAPAN

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との日米共同調査研究機関

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CONTENTS
目　次

DIPLOMYELIA, REPORT OF AN AUTOPSIED CASE
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CASE REPORT

The present instance of diplomyelia (doubling of the spinal cord) is the first autopsied case of this rare anomaly to be reported from Japan.

A 65-year-old woman, who had suffered from diabetes mellitus and hypertension for approximately 10 years, was hospitalized at the Hiroshima Railway Hospital on 4 April 1962 with complaints of oppressive sensation in the left front portion of the chest, palpitation, shortness of breath, and dizziness. At the time of hospitalization, she presented the following findings: average build and nutrition; blood pressure 124/76 mmHg; funduscopic findings Keith-Wagner class II; edema of the face; widening of the area of cardiac dullness on both sides; splitting of first apical sound; accentuation of second pulmonic sound; depression of liver edge approximately two fingerbreadths below the right costal margin; and negligible edema of the legs. Laboratory findings: urinary protein 0.1 g/100 ml; sugar trace; and white blood cells (+++) in sediment; glucose tolerance test blood levels/100 ml, fasting 139 mg, 2 hours 302 mg, 3 hours 286 mg; glucose present in urine at 2 and 3 hours; catalase present in urine; serum protein 8.5 g/100 ml; A/G ratio 0.9; icteric index 4.2; cholesterol 306 mg; NPN 25 mg; erythrocytic sedimentation rate 70 mm in 1 hour; erythrocyte count 3,890,000; leukocyte count 13,000; Hb 79% (12.8 g/100 ml); and Wassermann negative. Electrocardiogram exhibited evidence of myocardial damage. X-ray films of the chest disclosed bilateral widening of the cardiac shadow, and increased pulmonary markings.

症　例

ここに報告するdiplomyelia（脊髄二分症）はきわめてまれな奇形であって本邦では第1報告例と考えられる．

65歳の女性で約10年来糖尿病と高血圧症があるといわれている．1962年4月4日左胸部の圧迫感，動悸，呼吸促拍，目まいなどを訴えて広島鉄道病院へ入院した．入院時の所見としては体格，栄養ともに中等度で，血圧は124／76mm Hg，眼底所見はKeith-Wagner 2度，顔面浮腫，両側心濁音界の拡大，第1心尖音の分裂，第2肺動脈音の亢進を認めた．肝は肋弓下約2横指触れた．脚の浮腫は認めなかった．検査所見としては尿蛋白0.1 g／100 ml，糖－痕跡程度，白血球(卅)，耐糖試験は空腹時血糖139 mg／100 ml，2時間後302 mg／100 ml，3時間後286 mg／100 mlであった．ブドウ糖は尿中に2時間および3時間後にもみられた．またカタラーゼも尿中に認めた．血清蛋白は8.5 g／100 ml，A／G 0.9，黄疸指数4.2，コレステロール306 mg，NPN 25mg，赤沈1時間70mm，赤血球数389 ×10⁴，白血球数13,000，Hb 79％（12.8 g／100 ml），ワッセルマン反応陰性．心電図では心筋障害の所見を示した．胸部X線写真では心臓の陰影の左右への拡大と肺紋理の増強を認めた．

ABCC Department of Pathology[1] and Hiroshima Railway Hospital[2]
ABCC病理部[1]および広島鉄道病院[2]

†Hiroshima Branch Laboratory, Japanese National Institute of Health
国立予防衛生研究所広島支所

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Clinical Course Digitoxin and Theocolin were administered. In November 1962, the patient presented symptoms of a myocardial infarct; and in November 1963, she complained of dyspnea. Examinations revealed bilateral cardiac enlargement, right hydrothorax, severe ascites, edema of the lower extremities, and decreased urinary output. Under treatment with Digosin, injection of Igrosin, and oral administration of Diamox, the hydrothorax and ascites markedly decreased. Eight months later, on 2 July 1964, she suddenly died. The clinical diagnoses were diabetes mellitus, coronary atherosclerosis, myocardial infarction, chronic pyelonephritis, and Kimmelstiel-Wilson syndrome.

At autopsy ([redacted]), the principal gross findings were: myocardial infarcts (old scars in anterior and posterior walls of left ventricle, ventricular septum, and papillary muscles of right ventricle); bilateral ventricular hypertrophy (heart weight 450 g); moderate to severe atherosclerosis, particularly of aorta, coronary arteries, pulmonary trunk, and renal, mesenteric, and basilar arteries; arterial and arteriolar nephrosclerosis; old pyelonephritis; adenomatous polyp of ileum; and diplomyelia beginning at level of tenth thoracic segment and extending distally. No abnormality was noted in the vertebrae, medually canal, or dura mater.

Complete doubling was noted grossly in the lower third of the spinal cord (Figures 1, 2), and both of the separated cords were enclosed within a single dura and pia-arachnoid. No abnormality was noted in the medulla or the upper thoracic segments of the cord (Figure 3). The anterior median fissure in the lower thoracic portion of the cord was expanded to the shape of a deep U, and the transverse diameter was increased to more than twice the anteroposterior diameter (Figure 4). The peripheral nerves all emerged medially from the anterior nerve roots. Distal to the above level, the spinal cord was completely doubled (Figure 5). Each cord presented a pair of anterior horns and a pair of posterior horns, although the medial anterior horn of the left cord and the medial anterior and lateral posterior horns in the right cord were slightly incomplete; and each had a Clarke's column and a central canal. The pia-arachnoid was not separated, and there was a bundle of nerve roots between the two cords. At the level of the lower end of the thoracic segment, each cord appeared to be normally formed and displayed a central canal, paired anterior horns and roots, posterior horns and roots, and Clarke's columns. In the right cord, however, the central canal was enlarged, the medial Clarke's column was defective, and the cells of the medial anterior horn were ill-defined. The pattern was similar from the lumbar region to the sacral region.

臨床経過 ジギトキシンとテオコリンを使用した．1962年11月心筋梗塞の症状があった．また1963年11月には呼吸困難を訴えた．検査結果は，左右心臓の拡大と右の胸水，著明な腹水，下肢の浮腫，尿量の減少を認めた．ジゴシンの投与やイグロシンの注射，ダイアモックスの経口投与で胸水と腹水は著明に減少した．8か月後1964年7月2日急死した．臨床診断は糖尿病，冠状動脈硬化症，心筋梗塞，慢性腎盂腎炎およびキンメルスチール・ウイルソン症候群であった．

解剖([redacted])に際しての所見は心筋梗塞(左心室前後壁，心室中隔，右心室の乳頭筋の陳旧性瘢痕)，両側性心室拡大(心重量 450 g)，大動脈，冠状動脈，肺動脈幹，腎，腸間膜および脳底動脈の中等度ないし高度の動脈硬化症，動脈および細動脈性腎硬化症，陳旧性腎盂腎炎，回腸の腺腫様ポリープおよび第10胸髄の高さから脊髄下端までの diplomyelia であった．脊柱，脊柱管および硬膜には奇形は認められなかった．

脊髄の下部⅓には肉眼的に完全な重複があり(図1，2)，おのおのは単一の硬膜とクモ膜に包まれている．延髄や上部胸髄には全く奇形はなかった(図3)．胸髄下部の前正中裂は深いU字型に拡大し，その横径は前後径の2倍以上となる(図4)．末梢神経は前神経根から内方へ出ている．この高さから下部は脊髄は完全に二分されている(図5)．双方の脊髄は一対の前角と一対の後角をもっており，左の脊髄の内側の前角と右の脊髄の内側の前角および外側の後角はやや不完全である．また双方の脊髄は一個のクラーク氏柱と一個の中心管をもっている．クモ膜は分かれておらず，2つの脊髄の間に神経根束を認める．胸髄の下端の高さでは各脊髄は正常のようにみえ，中心管，一対の前角および前根，後角および後根，およびクラーク氏柱をもっている．しかし，右側の脊髄では中心管は拡大し内側のクラーク氏柱は欠けている．また内側の前角の神経細胞は不明瞭となっている．この様子は腰髄より仙髄にかけて同様である．

Histologic findings included severe aortic and coronary atherosclerosis with areas of ulceration or calcification of intima; atherosclerosis of the pulmonary trunk, with areas of thrombosis and recanalization; severe renal atherosclerosis, hyalinization of arterioles, and globular areas of hyaline glomerulosclerosis characteristic of diabetes mellitus; focal pelvic inflammation and irregular cortical inflammation and deep scarring consistent with old pyelonephritis; and cerebral atherosclerosis, with areas of hyalinization of the arterial walls.

組織学的所見は大動脈および冠状動脈の動脈硬化症と内膜の潰瘍化および石灰化，肺動脈幹の硬化症と血栓および再疎通，腎動脈の強い硬化症と小動脈の硝子化，糖尿病に特徴的な糸球体の硝子化，腎盂炎，および腎盂腎炎に相当する不整形の皮質の瘢痕形成と炎症，および動脈壁の硝子化を伴う脳動脈硬化症などである．

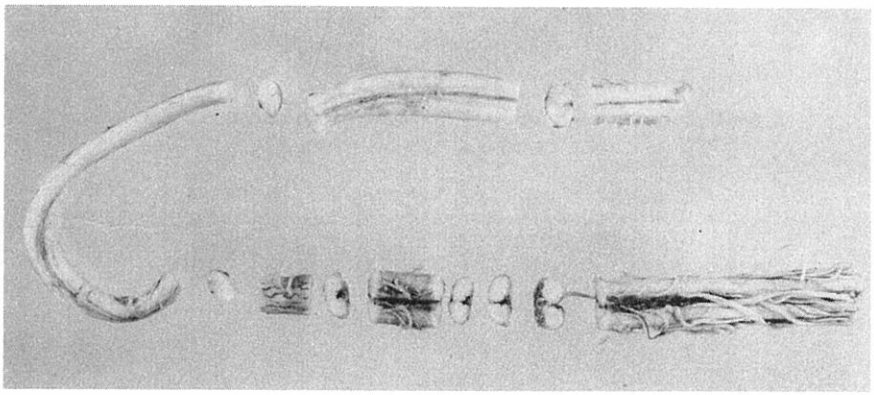

Figure 1 Spinal cord partially sectioned
図1　脊髄，一部横断面を示す

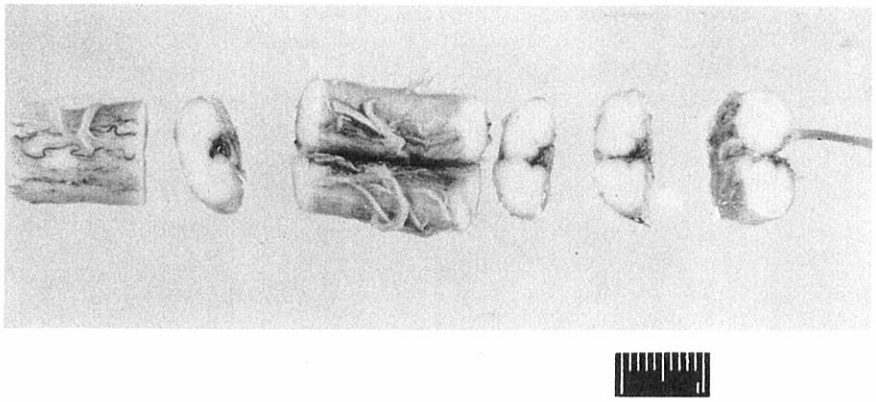

Figure 2 Anterior of spinal cord, from region of tenth lower thoracic to lumbar region
図2　脊髄前面，第10胸髄より腰髄部までを示す

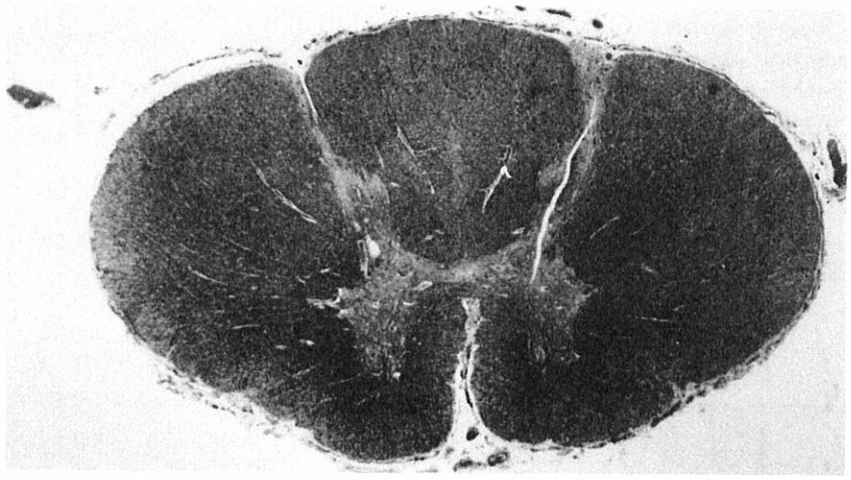

Figure 3 Cervical portion of spinal cord. No remarkable change is noted (myelin stain)
図3　頚髄横断面，著変なし（髄鞘染色）

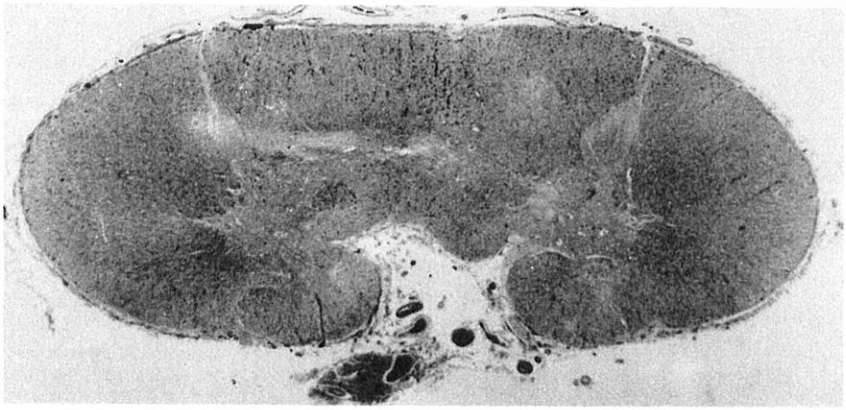

Figure 4 Lower thoracic portion of cord shows incomplete doubling (myelin stain)
図4　胸髄下部，不完全 doubling （髄鞘染色）

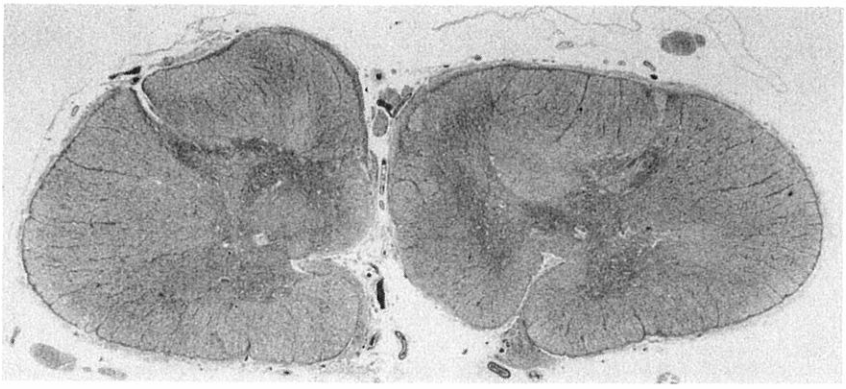

Figure 5 Lower end of thoracic region of cord shows complete doubling and complete separation. The separate portions are directed toward each other (myelin stain)
図5　胸髄下端，完全 doubling と分離．左右の脊髄は互いに内方に回転して向き合っている（髄鞘染色）

COMMENT

Diplomyelia is a congenital malformation characterized by duplication of the spinal cord to form two perfect segments. It has also been called doubling, reduplication, bifurcation, or diastematomyelia. The last term refers to fissuring of the cord by a variety of mechanisms and does not imply that the two segments of cord, if completely separated, are perfect. The proper designation for a case such as the one presented here, according to Herren and Edwards,[1] would be congenital diastematomyelia with diplomyelia. We shall, however, use the simpler term *diplomyelia.*

The table presented in the article by Herren and Edwards indicates that the ages at autopsy of persons with this anomaly are widely distributed, ranging from 5 months of fetal life to 76 years. Approximately 50% of reported cases, however, were either fetuses or infants less than 1 year old, and in most of the cases reported the sex or race is not mentioned. No previous report of diplomyelia in a Japanese has been located.

The majority of cases of diplomyelia are associated with spina bifida, and a smaller number are complicated by meningocele or meningomyelocele. Syringomyelia, hydromyelia, anencephaly, acrania, hydrocephaly, and cyclopia have been reported in a few instances; and kyphosis, scoliosis, and occasionally club foot, appear to be secondary complications associated with spina bifida and peculiarities of innervation to the extremities.

The extent of doubling ranges from only a small part of the cord to involvement of its entire length. Whereas 36 of 50 reported cases (72%) of diplomyelia exhibited doubling of only a portion of the spinal cord, in the remaining 14 (28%) the entire cord was doubled distal to the point of first division. The sites of doubling occurred predominantly in the lumber segment, approximately as often in the thoracic as in the sacral region, and rarely in the cervical portion. A few patients had symptoms of diminished muscular power and disturbances of sensation, but most patients had no complaints referable to the cord.

In the development of the fetus, the formation of the spinal cord precedes that of the spinal column, and normally in the evolution of the spinal cord from the neural plate, only a single primordium is seen. In this condition, however, Holmdahl and Ikeda[2] postulate that the lower portion of the cord develops from a group of primitive nerve cells separate from the primordium of the upper portion of the cord. These form two or more neural canals which later join to form the primordium of the single central canal from the upper portion of the cord. They explain that

考　察

Diplomyelia は脊髄が重複するという先天的な奇形である．これは doubling，reduplication，bifurcation または diastematomyelia ともいわれる．最後の名称は奇形発生機序が異なって脊髄の重複化が区々の場合につけられるものであって，そこの分化が完全である場合にはこの語は使用しない．ここに述べる症例は Herren と Edwards [1] によれば diplomyelia を伴った先天的 diastematomyelia と呼ばれるものである．われわれはここでは単に diplomyelia と呼ぶ．

Herren と Edwards の表によるとこの奇形の年齢分布は広く，胎生5か月から76歳にまで及ぶ．しかし，報告例の約50%は胎児か1歳以下の乳児である．性や人種についてはこれに触れた報告はほとんどない．日本ではこれまでに diplomyelia の報告例はない．

Diplomyelia の大部分は脊椎分離症を合併し，また少数のものは脳膜脱出や脳膜脳髄脱出を合併している．脊髄空洞症，脊髄水腫，無脳症，無頭症，脳水腫，単眼症などの合併も報告されている．亀背，脊椎側弯症および足弯曲症も脊椎分離症の二次的合併症としてみられ，四肢の神経支配に異常を示す症例も報告されている．

Doubling の範囲は脊髄の小部分から全体に至るものまでさまざまである．50例の diplomyelia の報告例中36例（72%）は脊髄の一部にのみ doubling を示し，残りの14例（28%）は最上端から最下端まで doubling を示している．Doubling を示すところは腰髄に多く，胸髄と仙髄とはほぼ同様の割合である．そして頚髄ではまれである．一部の症例では筋力の低下や知覚障害があるが多くの場合は脊髄に関連した訴えはない．

胎児の成長において脊髄の形成は脊柱管のそれに先行する．そして脊髄は原始神経板より分化し，そこでは単一の原基しかみられない．しかし，Holmdahl と池田 [2] によると，脊髄の下部は上部の原基より分かれた原始神経細胞の集団から発生するという．これらが2つまたはそれ以上の神経管を形成し，その後これらが単一の神経管と

doubling occurs when fusion of the cavities does not proceed satisfactorily.

SUMMARY

A 65-year-old woman who had suffered from diabetes mellitus and hypertension for 10 years was hospitalized at the Hiroshima Railway Hospital. In July 1964, she had a heart attack and suddenly died. The principal clinical diagnoses were diabetes mellitus, coronary atherosclerosis, myocardial infarcts and Kimmelstiel-Wilson syndrome.

The principal autopsy findings were myocardial infarcts, bilateral ventricular hypertrophy and dilatation, atherosclerosis of various arteries all over the body and diplomyelia, beginning at the level of the lower thoracic segment and extending distally. Both of the separated cords appeared to be normally formed and both showed inward twisting which was more marked distalwards. Each was covered by an independent arachnoid but enclosed within a single dura mater. No malformation or deformity was noted in the vertebrae, etc. This rare malformation of the spinal cord is probably the first case to be reported in Japan. A review of the literature was made on the frequency, site, and nature of this disease.

なる．そしてかれらは doubling は空洞（原始神経管）の融合が不完全のときに起こると説明している．

要　約

65歳の女性で10年来糖尿病および高血圧症のあることを指摘され，広島鉄道病院内科に入院加療中であったが1964年７月２日心臓発作をおこし急死した．おもな臨床診断は糖尿病，冠状動脈硬化症，心筋梗塞およびキンメルスチール・ウイルソン症候群であった．

おもな病理解剖所見は心筋梗塞，両側性心室肥大および拡張，全身諸動脈の動脈硬化症などのほかに胸髄下部以下の脊髄の完全二分症（diplomyelia）があり，二分した左右の脊髄はほぼ正常の形態を示し，双方とも内方へ回転していて，この度合は下方にいくにしたがって強い．各々はそれぞれ独立したクモ膜に覆われているが，硬膜は一枚で双方を包んでいる．脊柱その他には奇形や変形は認められなかった．本症は，脊髄奇形中希有なものであり，本邦第１の報告例と思われる．本症の発生頻度，部位および形態などにつき文献的考察を加えた．

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（脊椎分離症と脊髄空洞症を伴った脊髄の重複）