CLINICO - PATHOLOGIC - CONFERENCE

(A weekly clinical and pathologic case conference part:cipated jointly by members of the Departments of Internal Medicine and Pathology of the Chulalongkorn Hospital, Faculty of Medicine, Chulalongkorn University, and by the third and fourth year medical students).

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(Headache – Vomiting – Papillaedema – Neurosensory deanfess – Facial Paresis and Sudden death).

This unfortunate 40 year old woman who was previously well had 3 year history of slowly progressive intermittant headache, 6 months history of dimming of vision, poor appetite and weight loss; 3 months double vision and 2 days continuous headache with vomiting, numbness of the left side of the face and arm. During this period she frequently visited Chulalongkorn Hospital where the diagnosis of neurasthenia or psychoneurosis were repeatedly entertained. Not until her last visit on April 12, 1971 while seen by the enthusiastic intern, that bilateral full blown papilledema was discovered. She was admitted.

Physical examination revealed a pleasant, cooperative and alert middle age woman in no acute distress.

BT 36.2°C. PR 84/min. BT 130/80 RR 24/Min.

General physical finding was unremarkable. Neurological examination disclosed normal mental status. There was no neck stiffness and the Kernig's sign was negative. Gait and speech were normal Cranial Nerve (C) examination revealed :–

C_2 – Visual acuity deficit, bilalerally right 6/18 O.U. left 6/24 O.U. The visual field showed concentric narrowing. Fundoscopic examination disclosed full blown papilledema with exudatation.

$C_{3,4,6}$ – The right pupil was 3 mm. and sluggishly reactive. The left pupil 2 mm. promptly reactive. Extraocular movement was normal.

C_5 – Pain sensation and corneal reflex was diminished on the left side.

C_7 – There was mild right facial weakness of lower motor neurone type.

C_8 – There was neurosensory deafness on the right sibe.

The caloric test using cold water showed no response in the right ear, the left ear was normal.

The rest of the cranial nerves were normal. The motor, sensation and cerebellar functions were within normal limits. The deep tendon reflexes were slightly and equally hyperactive. The plantar responses were down. Laboratory findings showed normal blood and urine examination.

Chest X – ray revealed normal heart and lung shadow. Plain skull film disclosed demineralization of sella turcica which was suggestve of increased intracranial pressure. The electroencephalogram showed minimal diffuse dysrhythmia.

The brain scan showed slight increase scattering uptake in both cerebral hemisphers.

Bilateral carotid angiogram displayed bilateral hydrocephalus, elevation of carotid siphons bilaterally which was consistent with intrasellar lesion. Ventriculogram also showed evidence of communicating hydrocephalus. No definite evidence of a space occupying mass was seen. Ventricular pressure was 270 mm. water The cerebrospinal fluid was clear, colorless containing 25 mg% of protein, 45 mg % of sugar, 28 lymphocytes and 24 red blood cells/cu. mm.

RISA scanning done two weeks later revealed enlarged left lateral ventricle. No entry of RISA in the right lateral ventricle. Marked delayed absorption of CSF. The spinal fluid presure was 180 mm water The clear and colorless CSF contained 8 neutrophils and 12 lymphocytes/cu. mm, 12–5 mg % protein, 65 mg % of sugar.

Having found no demonstrable cause of hydrocephalus. Prednisolone was started initially 40 mg/day on 24.5.71 Throughout her hospitalization, she always appeared fully alert, pleasant and seldomly complained of vaque headache which was easily relieved by analgesic. No change in neurological finding was noted except for the improvement in vison, which was 6/12 O.U. No apparent change in eye ground, She was discharged from the hospital on September 28, 1971.

Apparently the patient had done well at home until Oct. 5, 1971, while having supper she suddenly experienced severe generalized headache and became unconscious later. She was rushed to the hospital where she was found to have respiratory arrest. Spontaneous respiration cameback after a short resuscitation. Neurologically, the patient was deeply comatose. There was decortication in response to deep pain stimulus. There were bilateral full blown papilledema. The left pupil was 4 mm. and the right was 3 mm. indiameter, nonreactive. Doll's eye was negative. Two hours later decerebrate rigidity was noted. Ventricular tap was done and revealed a pressure of 370 mm. water. Bloody fluid was thoughttobe probably the effect of trauma (evidenced by decreasing rbc 8,000 to 4,000 /cu. mm. inbottle 1, day 3). The patient had respiratory arrest again while being retapped and finally expired on October 7, 1971.

Dr. C. Souwanwela : From the subjective symptoms alone, there is no doubt at all that this 40 year old man was having a generalized increase intracranial pressure, at least during the last several months pior to admission. The finding of full blown papilledema bilaterally make this diagnosis unmistakebly. I am not particularly think that double vision or diplopia is helping much. Monocular diplopia is rare and commonly followed primary diseases of the eye proper. Binocular diplopia, on the other hand, is caused by failure or weakness of the occular musculature. This is quite unlikely in our patient since the neurological examination disclosed intact function of all three nerves namely C3, C4 and C6. I have experienced that failure of focusing or sometimes, simply a blurring of vision may be misinterpreted by the patient of diplopia. I would consider this as the only explanation for double vision stated by our patient here.

It is not clear, from the information provided whether the increase of the intracranial pressure has been there since the beginning of the patients siokness 3 years ago, or the patient really had psychoneurotic headache first and the increase pressure is of something else occuring separately only during the past few months before admission. I will come back to his later.

The neurological examination showed a normal mental status. This is important to me which will be eloborated later on. The findings of reduction in visual acuity as well as of the visual fields do not add much to the diagnosis–except to confirm that there is an increase in intracranial pressure. I can not explain why the right pupil is larger that the left one especially when there is no positive finding pertaining to C3, anywhere from the periphery to the Ettinger–Westhpal nucleus in the brain. I would therefore try not to think about it.

There is a definite impairment of left C5 as evident by the findings of diminished pain sensation and corneal reflex on the left side. Right facial weakness of lower moter neurone type was detected. There is an impairment of both the vestibular and auditory functions of the right C8 as elicited by the tests and mentioned in the protocol. I discard lesion of the inner ear as the cause of C8 deficit because of the presence of increase intracranial pressure.

Considering that the patient is having impairment of C5, C7 and C8 at the same time, a lesion located within the brain–stem is quite logical. There are several points against this posibility, however. A lesion in the brain stem capable of producing C7 deficit and impairment of both the vestibular and anditory nuclei must be quite large and in such a case, long tract signs must be more or less positive. Also, in such case, the facial weakness must be of upper moter neurone type. If all of the information given in the protocal is correct, the lesion tn our patient could not be located within the brain–stem. It might be around there somewhere in the vicinity of brain–stem especially in cerebropontine angle or at the basal cistern.

A lesion located at cerebropontine angle would fit well if there is no C5 impairment which there is in our case, and it is on the opposite side too. I would like therefore to scratch this out unless there are two lesions in both angles. The mechanism of the cause producing lesion in this patient seem to be moving; at first, while located in the lower part of the base of brain, it produces impairment of right C7 and C8. Then, moving a little upward, it crosses the mid–line from right to left causing a deficit in the left C5.

What, then could the cause which behaving in such a mechanism be. This is not an easy question considering that there in also an increase in intracranial pressure for a long period of time. A space occupying lesion in the brain–stem is out because of the absence of long tract signs. The only thing that can produce both the increase pressure and cranial nerve deficincy as such, is cerbrospinal fluid. An obstruction of the flow at the base of brain causing accumulation of fluid in the ventricles can answer most of the questions in our case.

A congenital malformation occuring at the age of 40 is not heard of except for one condition–an Arnold–Chiari malformation which can be found at this age. Stretching the brain stem and the cranial nerves at the base of the brain while at the same time blocking the CSF flow may be responsible for all of the findings here. The condition is so rare, however, that a diagnosis should never be attempted unless there really is no other way out.

Infective condition is also possible. Tuberculous areachoiditis with basal cranial nerve involvement is quite common in Thailand. However, the history of sickness is too much too long. I have never known of tuberculous meningitis producing increase intracranial pressure yet the patients mental condition is still undisturbed as it is noted in this patient.

Purulent meningitis is out. No acute symptoms were present neither was there any sign of meningeal irritation. Slow progressive fungal infection causing adhesive arachnoiditis and healed process of any infectious conditions of the meninges however may produce this type of lesion as well.

The most likely possibility in my differential diagnosis is a parasitic disease. This is not only that the patient is living in this part of the world where parasitic infestations are ubiquitous, but also the peculiar mechanism of the disease. Cysticercosis, being the most common organism, is strongly possible in this case. Three types of cerebral cysticercosis have been classified by a report from South America. They are:–

1. Parenchymatous type of which the cysts are located in the brain tissue. The main symptoms are dementia and mental disorders.

2. Ventricular type inwhich the cysticercus is located within the ventricle. Signs of increase intracranial pressure, on and off, are chief findings.

3. Subarachnoid type which in known as cysticercus latimosum. Usually, the cyst is localized in the subarachnoid space, at the base of the brain, producing adhesive arachnoiditis and CSF pathway blockade.

The last type is what I have in mind as the most possible diagnosis in this patient.

Other unusual possibilities such as vascular lesions and neoplasms should be mentioned for the sake of completeness. The only blood vessel out–side the brain–stem is the basillar artery and branches. Aneurysms and ectasia of basillar artery could produce similar clinical manifastations as found in our patients. Ectasia and torthousity of basillar artery has been recently reported from Sweden. The pulsating artery in the fourth ventricle produces partial obstruction of the CSF flow and generalized increase intracranial pressure.

Brain tumor capable of producing hydrocephalus must be quite large and if so, localizing brain stem signs such as nystagmus etc. should present. However, if the tumor is small and multiple, features similar to what manifested by our patient may be found.

Before giving final clinical diagnosis, I would like to know the result of various investigations done in this patient.

Dr. N. Suwanwela : Plain skull X-ray revealed separation of bony sutures compatible with increase intracranial pressure. Carotid angiograms showed lateral ventricles emlargement, more or the right side.

Ventriculographic studies revealed dilatation of frontal horn of right lateral, third and fourth ventricles. The left lateral ventricle appeared normal. Cerebral aqueduct was not visualized but contrast is passable. There is in the right lateral film, a shadow in the area of massa intermedia which could be a soft tissue mass or tumor that may be responsible for dilatation of the right foramen of Monroe.

Dr. M. Posayachinda : Brain-scannings revealed slight increase up-take in both cerebral hemisphere and up-take in the left lateral ventricle at the cranial floor. Isotope study showed absence of radio-activity in subarachnoid space even 48 hours after injection. There is marked delayed of absorption of CSF.

Dr. C. Suwanwela : Spinal fluid examination was not remarkable much. There is some increase in lymphocytes in the CSF. Spinal fluid lymphocytosis may be encountered in variables of pathology from chronic infection to carcinomatosis.

The special investigations confirm my clinlical impression. The final episode of the patient is interesting a sudden onset of sovere headache, coma which followed soon by death. Two possibilities may be explained:

1. Massive spontaneous subarachnoid hemorrhage or rupture of an abscess.

2. A sudden blockade of CSF pathway with marked prompt increase in pressure. However, in my experience, the headache in such mechanism should not be abrupt and intense or followed by sudden death such as found in our case.

If the patient did have cysticercosis in the ventricle however, this final episode may be explained. Cysticercosis tends to be multiple. The floating cysts may suddenly block the CSF pathway while other cysts may be localized in the third ventricle, at base of brain with adhesion arachnoiditis and still others may localed somewhere else. This is my first choice of diagnosis. Other possible diagnoses are ectasia of basal artery with massive subarachnoid hemorrhage, basal arachnoiditis and multiple small tumors of tumor-like conditions in the basal part of the brain are also possible.

Dr. S. Shuangshoti : The brain shows recent basal subarachnoid hematoma around the pons and cerebellum. At the upper pons there is a whitish area underneath a mass of clotted blood. This area is corresponded to a small elongated tissue projecting into both cerebropontine angles. Under microscopic examination, this whitish tissue represents a flat worm, folded upon itself and embedded within the meningeal tissue. Numerous calcified spots are observed within the worm parenchyma.

The external surface shows thick cuticular layer. The inner part is composed of loose areolar tissue and mesenchyme. We have consulted parasitologists for identification of the parasite and tentatively reported to be a sparganum of a tape worm. The worm is located at the base of the brain amidst area of massive subarachnoid hemorrhage of which, no difinite point of bleeding could be found despite careful examination. The major blood vessels are all intact. It is possible

that a small artery may be eroded by the inflammatory process and ruptured and the area is completely obscured by mass of clotted blood. Microscopically, however, we are not able to find arteritis.

This is our second case in the past 5 years. Two years previously a case of cerebral sparganosis was discussed in a CPC and the clinical diagnosis then was tuberculous meningitis.

Sparganosis is not rare in Thailand. Tansurat[1] has reviewed literatures in this country and cases were found to be reported from all parts of the country.

The sparganum is a solid larval tape – worm belongs to a genus Spirometra. Its first intermediate host is a cyclop and the second intermediate host is frog or snake. Man is accidental second intermediate host when inadvertently eating cyclop infested with procercoid larva. The larva then penetrates the intestinal wall and become excysted as sparganum in any organ. Man can also become infected by eating raw or partially cooked frog containing sparganun.

Final Anatomical Diagnosis

Primary

Encysted tape worm larva, probably sparganosis, subarachnoid space at base of brain stem and cerebro – pontine angles,

Hydrocephalus, moderate;

Recent subarachnoid hemorrhage, severe;

Ref. 1. Tansurat, P: Human sparganosis in Thailand. J. Med. Assoc. Thailand **49**: 391 – 1966.