Three Cases of Pure Neuritic (PN) Leprosy at Detection in Which Skin Lesions Became Visible During Their Course

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Pure neuritic leprosy (PN leprosy) is a type of leprosy with nerve involvement, but without obvious skin lesions. It is not uncommon in the south of Bangladesh,¹⁾ but its nature is less recognized than that in other types of leprosy. Male is dominant on its occurrence with higher disability grading. The histopathological study shows that the entire spectrum can be observed in nerves of PN leprosy.^{2) 3)} There was no relation among clinical parameters, such as the number and distribution of affected nerves, the immune response and its histopathology.³⁾ Therefore the treatment of PN leprosy is not well-established at field level. Out of 1,741 newly detected cases by Dhanjuri Leprosy Project-Khulna Branch (PIME Sisters) in Khulna, the south of Bangladesh from 1994 to 1998, 141, or 8.10 % were diagnosed as PN leprosy. 6 cases out of 1,741, or 0.34% were canceled afterwards because of wrong diagnosis, of which one misdiagnosed as PN leprosy. Three cases out of 140 of primary neuritic leprosy proved to have obvious skin lesions some time after their treatment started. The details of the three cases are described in this paper.

Case 1.

Birth: 1971, Age: 25 years old at detection, Sex: Male, Race: Bengali, Birth Place: Pathikhalghata, Kathalia, the south of Bangladesh. Registered on 3/Sep./1996, Family history: His wife was detected as BT/PB leprosy on 28/Nov./1996. His son is described in Case 2. below.

History of present illness:

The disease began one year before detection. At first he got pain on the left hand and the ulnar nerve trunk. It was followed by the pain in bilateral radial, ulnar, median, radial cutaneous and common peroneal nerves. He first consulted a doctor at the Barishal Medical College Hospital near by and was told to take some medicine for one month. Afterwards he came along from far to Khulna for treatment voluntarily.

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Clinical findings and its course:

Peripheral nerve involvement, manual muscle testing(MMT) and disability grading are shown in Table 1, 2, and 3, separately. Bilateral hands and foot-soles had sensory deficit. The muscle wasting of bilateral interosseous, thenar muscles and hypothenar was already shown. There was no skin lesion seen.

He was diagnosed as PN leprosy and WHO/MB regimen was started under steroid cover owing to ongoing neuritis. The routine slit skin smear was 4+ positive(Average of 3 sites) at detection. After 4 weeks' treatment, he developed ENL reaction and also the skin infiltration of extremities became visible. And then the classification was changed from PN to BL leprosy. He was released from treatment (RFT)

on 2/Feb./1998 after taking 16 doses of WHO/MB regimen, because the shortened period(12 doses) of WHO/MB regimen started to be applied in Bangladesh in January 1998. Disability grading at RFT was the same as it was at detection. The very pale skin infiltrations of BL leprosy were not recognized by any staff including a medical officer at detection.

Case 2.

A son of Case 1., Birth year: 1989, Age: 7 years old at detection, Sex: Male, Race: Bengali, Address: Pathikhalghata, Kathalia, the south of Bangladesh. Registered on 4/Oct./1996, Family history: His father got leprosy described above in Case 1. His

Table 1. Peripheral nerve involvement of case 1.

Peripheral Nerve	Enlarged		Tender	
	R	L	R	L
Ulnar nerve	(+)	(++)	(+)	(++)
Median nerve	(+)	(++)	(+)	(++)
Radial nerve	(+)	(+)		
Radial cutaneous branch	(+)	(+)		(+)
Common peroneal nerve	(+)	(+)		
Posterior tibialis nerve	(+)	(+)		

Table 2. Manual Muscle Testing of case 1.

	R	L
ADM	2	0
APB	2+	0
TA	5	4
Peroneal Group	5	4

ADM: Abductor Digiti Minimi

APB: Abductor Polisis Brebis

TA: Tibialis Posterior

Table 3. Disability Grading of case 1.

	R	L
Hand	2	2
Foot	1	1
Eye	0	0

mother was detected as BT/PB leprosy on 28/Nov./1996.

History of present illness:

The disease began 6 months before detection. At first he got the right ulnar, median and common peroneal nerves enlarged with pain. Though he did not have any visible skin lesions, his father suspected him to have caught leprosy and brought him to a leprosy clinic.

Clinical findings and its course:

Peripheral nerve involvement is shown in Table 4. Sensory deficit was unclear because he was 7-year old child at detection. MMT seemed to be normal. The routine slit skin smear was not taken because of his young age. There was no visible skin lesion at detection. He was diagnosed as PN leprosy and WHO/MB regimen was started under steroid cover owing to ongoing right ulnar nerve neuritis. On 16/July/1997 small hypesthetic hypo-pigmented maculae were discovered in the right cheek, chin, shoulder and heel at a clinic and the routine slit skin smear was negative. Skin lesions showed any signs of inflammation. They did not look like ENL reaction which patient's father had. The classification was

changed from PN to BT leprosy. He was released from treatment (RFT) on 3/Feb./1998 because of the shortening of the period of WHO/MB regimen. Disability grading was 0 both at detection and at RFT.

Case 3.

Birth year: 1962, Age: 35 years old at detection, Sex: Male, Race: Bengali, Address: Khalishpur, Khulna, the south of Bangladesh. Registration on 20/Feb./1997.

History of present illness:

The disease began with hypesthesia of the left second toe 6 months before detection. He developed multiple nerve involvement gradually afterwards. He came to a leprosy clinic voluntarily.

Clinical findings and its course:

The right ulnar, common peroneal and posterior tibial nerves were enlarged with tenderness at registration. A big toe of the right foot, the second toe and a sole had sensory deficit. The routine slit skin smear was negative at detection. Disability grading was 1 because of the anesthesia of a foot sole. He was diagnosed as PN leprosy and WHO/PB regimen was

Table 4. Peripheral nerve involvement of case 2.

	Enlarged		Tender	
	R	L	R	L
Ulnar nerve	(++)	(+)	(+)	(+)
Median nerve	(+)		(+)	
Radial nerve	(++)		(+)	
Common peroneal nerve	(+)			

Table 5. Summary of 3 cases.

CASE	AGE, SEX	No. of nerves involved	Treatment	BI	Skin lesion	Class	Remark
1	25yo M	12	MB→MB	4+	infiltration	PN→BL	ENL, Neuritis
2	7yo M	4	MB→MB	(-)	hypopigmented macules	PN→BT	Neuritis
3	35yo M	3	PB→MB	(-)	ill defined hypopigmented macules	PN→BT	

BI: Bacterial Index (Average of 3 sites)

started. On 12/June/1997 when he completed 5 months' PB regimen, several skin lesions were discovered on bilateral forearms at a clinic, which were hypesthetic hypo-pigmented maculae with their margins ill-defined. There was no sign of inflammation around skin lesions and no further development of nerve damage on limbs. The classification changed from PN to BT and WHO/MB regimen was started. A slit smear test was negative again on 17/July/1997. He was released from treatment (RFT) on 2/July/1998.

Discussion

Pure neuritic leprosy(PN leprosy) has been paid less attention than other types of leprosy, mainly because it lacks eruptions and is thought to be less important. Therefore, the less interest of field workers has been given on it. PN leprosy sometimes escapes for detection because the criteria of nerve involvement of it is not well defined. Also the clinical course of PN leprosy after its treatment being started has not been well described. These cases described above were detected as PN leprosy and treatments were started but skin lesions were recognized afterwards during their treatment.

Case 1 was a rare case with skin smear positive. Because the skin smear of PN cases is usually negative, the skin smear positivity was not expected.^{2) 4)} Skin lesions were not localized and the changes were minimal, which were hardly recognized on patient's

dark skin. It could have been recognized together with the positive result of skin smear. Because the number of nerves involved was as many as 12 with symmetrical distribution and the slit skin smear was 4+(average of 3 sites) at detection, this case was thought to be quite advanced one. He could have come to a clinic when he got only a few nerves involved, if he had had proper knowledge of PN leprosy(Table 5).

Case 2 developed BT skin lesions 9 months after detection. It was interesting that he had neuritis in the right ulnar nerve at detection but that skin lesions did not show any signs of inflammation during its course after they were recognized. Case 1 and 2 were father and son, and they might have common genetic effects that skin lesions were difficult to recognize.

Case 3 also developed BT skin lesions 5 months after detection. This was the case which wrong treatment was given to due to wrong diagnosis given at detection.

The incidence of PN leprosy is high in the south of Bangladesh. The proportion of PN cases to newly detected cases was once 6.16% (46 out of 747 newly detected cases from June 1992 to Nov. 1993, M:F=33:13 in Khulna.1). It was 7.87% (137 out of 1,741 cases from 1994 to 1998, M:F=79:58) for the last 5 years (Table 6).

At field level the diagnosis of PN leprosy is only made on such clinical findings as thickened nerves, tenderness or sensory/motor paralysis. Over-diag-

Table 6. PN leprosy among new cases detected (1994-1998)

	Total	PN leprosy	Percentage
1994	392	34	8.67%
1995	494	38	7.69%
1996	346	37	10.69%
1997	282	15	5.32%
1998	227	13	5.73%
Total	1,741	137	7.87%

nosis can happen, for example when a great auricular nerve could be misdiagnosed a leprosy lesion in case it is visible or palpable. The self-healed cases to leave affected peripheral nerves thickened can be taken as PN leprosy. The detection of PN leprosy seems to be later than that of other types in classification of leprosy. The average time lag between recognition of symptoms by patients and detection was 28 months in our series and 29 months in India.^{1),2)} The mean age at detection was older as well. The main reason of delayed detection can be due to poor awareness of PN leprosy both in patients and in medical staff. The information on the symptom of PN leprosy is usually neglected in the health education on leprosy. The WHO/PB regimen tend to be considered for PN leprosy in Bangladesh,⁵⁾ which is sometimes inadequate such cases presented in this paper.

From our experiences among newly detected pure neural leprosy patients, there are a few of cases which skin lesions are missed at detection or skin lesions become visible afterwards, those cases may not be given proper treatment as well as case management.

More attention should be paid to PN leprosy for maximum detection and prevention of disability.

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経過中に皮疹の存在を認めた純神経型ハンセン病 (Pure Neuritic Leprosy) の3症例

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皮疹を認めず、末梢神経症状のみを呈する純神経型ハンセン病(pure neuritic leprosy or PN)はバングラデシュ南部では稀ではない。1994年から1998年の当施設の新患登録統計1,741名中では141例(8.10%)がPNと診断された。この内1例は診断の誤りとして登録を抹消された。また、3例は、経過中に明らかな皮疹を認めたため他の病型に変更された。このためPNの占める割合は137例(7.87%)であった。これらの経過中に皮疹を認めた3例について、詳細を報告した。ハンセン病コントロールにおける住民に対する啓発活動の中で、PNは無視されがちであり、それゆえ発見も遅れ勝ちである。このため末梢神経麻痺による障害度も高い。ことにPNの中で、経過中に皮疹の存在を認める症例や、初診時に皮疹の存在を見落としている症例が少数存在することがあり、治療も変更せざるを得ないことがあるので、注意を要する。

症例1は、25才のベンガル人男性。広範囲なBLの皮膚浸潤を初診時に診断できなかった例であり、多発性神経症状よりWHO/MBを開始した。初診時の菌検査は4+(3箇所)の平均値であった。治療開始4ヶ月後、ENLを併発し、同時に皮膚の浸潤も確認された。

症例2は、症例1の子、7才男児。初診時、皮疹を認めず、右側の尺骨神経炎を起こしていた。WHO/MBとステロイド剤による治療開始の9ヶ月後にBTと思われる皮疹を初めて確認した。皮疹確認時の菌検査は陰性であった。

症例3は、35才のベンガル人男性。初診時、右側の尺骨神経、総腓骨神経、後脛骨神経の腫脹と圧痛を認めた。菌検査は陰性であった。PN/PBとしてWHO/PBの治療が開始された。5ヶ月後、両側の前腕に数個の知覚麻痺を伴う境界不明瞭な脱色素斑を認めたため、診断をBT/MBに変更し、WHO/MBの治療をやり直した。

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