

THIRD GENERATION INFANTILE SYPHILIS— AN UNUSUAL PRESENTATION

Ratan Singh, R. C. Sharma and M. C. Baruah

*Department of Dermatology and Venereology, Maulana Azad Medical College
and Associated Irwin and G. B. Pant Hospitals, New Delhi, India*

Abstract. A case of third generation early congenital syphilis with unusual features is reported and discussed. The infant presented with Livedo reticularis and an ulcer on the right forearm since birth; the underlying radius and ulna showed osteomyelitic changes with sequestrum formation and a pathological fracture. The rest of the skeletal system was normal. The blood STS (VDRL) was reactive at 1:64. The child showed no other evidence of congenital syphilis. Livedo reticularis and bony lesions resolved promptly with penicillin therapy, but abnormal movements at the wrist joint persisted. The mother had pathognomic Hutchinsonian incisors and a reactive VDRL test.

Key words: Syphilis congenital; Osteomyelitis; Livedo reticularis

Before the controversy of transmission of syphilis to the third generation could be finally settled, the introduction of penicillin in the therapy and control of syphilis recorded such a precipitous fall in the incidence and prevalence of syphilis all over the world, that even second generation syphilis almost disappeared from the clinical scene and third generation syphilis did disappear entirely. The last report on third generation syphilis in the medical literature appeared in 1965 (3, 8). With the resurgence of syphilis throughout the world, however, both second and third generation syphilis are again being seen: the latter very infrequently, of course.

CASE REPORT

An 18-day-old male (Fig. 1—III, 12); the first child of the family, was admitted to the Dermatology and Venereology Department on 5th April, 1972, with an ulcer over the right forearm, present since birth. A whitish membranous patch was noticed at the site, removal of which revealed an ulcer underneath.

The propositus' mother (Fig. 1—II, 5), aged 18 years

and third child of her parents, had classical Hutchinson's teeth and open-bite deformity (Fig. 2). She had no history of abortion or miscarriage. There was no other evidence of congenital syphilis. Her blood STS (VDRL) was reactive at 1:64. She had never received any formal or "happen chance" treatment.

The father (Fig. 1—II, 11) aged 22 years had no history of genital sore, urethral discharge, inguinal bubo or extra-marital exposure. He was in perfect health and his blood STS (VDRL) was persistently non-reactive.

The grandmother (Fig. 1—I, 2), aged 50 years, had no complaints and her serum VDRL and FTA-ABS were non-reactive. She had had eight pregnancies, no abortion and no miscarriage. Four children (Fig. 1—II, 4, 8, 9) outcome of second (twin), sixth and seventh pregnancies, died of marasmus during infancy; only 2 children (Fig. 1—II, 6, 10), aged 17 and 10 years, born of fourth and eighth pregnancies could be examined; both had blood STS (VDRL) non-reactive and no evidence of congenital lues.

The grandfather (Fig. 1—I, 1) never attended for medical checkup.

Examination (5.4.1972)

The infant appeared healthy and fairly well developed (weight 3 kg). A granulomatous ulcer 3 cm x 2 cm was seen

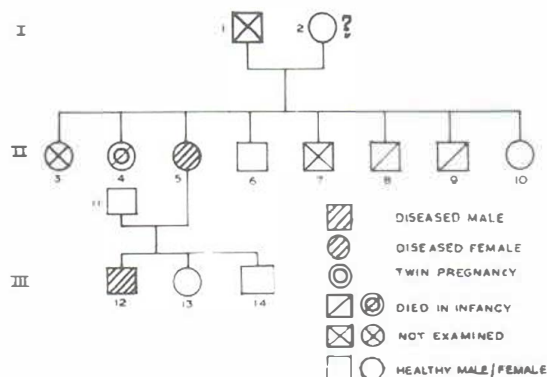


Fig. 1. Pedigree. Third generation syphilis.

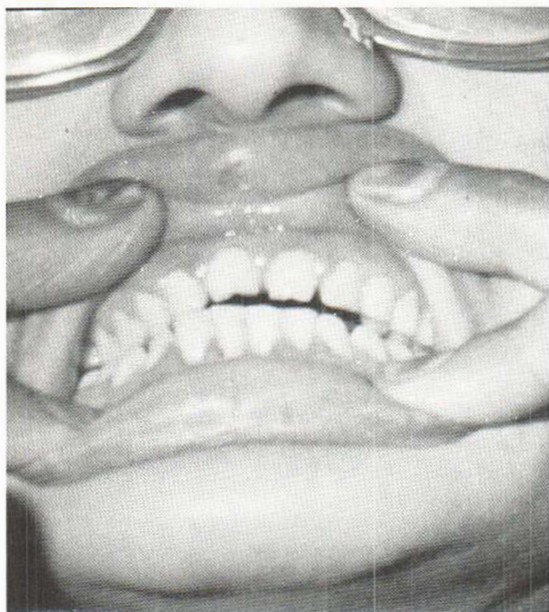


Fig. 2. Pathognomic Hutchinson's teeth and open-bite deformity of the mother of the infant.

on the flexor aspect of lower third of the right fore-arm. The floor of the ulcer was partly covered with slough and at one point a probe could be easily passed down to the underlying bone. Abnormal movements were present above the wrist-joint and bones appeared fractured. There was no tenderness or swelling at the ends of long bones. Livedo reticularis was present on the skin, all over the trunk and extremities. The liver was palpable 1 cm below the costal margin. No other clinical lesion was detected. The serum VDRL was reactive at 1:64. Darkfield examination of the ulcer did not show any *Treponema pallidum*. Pus culture from the ulcer was sterile.

Radiological examination

Dislocation of right infra-radio-ulnar joint was seen, with fractures in the shafts of radius and ulna. There was marked periosteal reaction and sequestrum formation in both the bones (Fig. 3). A radiograph of the entire skeletal system of the infant did not reveal any other bony lesion.

Treatment

The child was given procaine penicillin, 200 000 units intramuscularly, daily for 20 days and a plaster of paris cast applied to the right fore-arm. The mother was given PAM (procaine penicillin G in oil with 2% aluminium monostearate) 600 000 units intramuscularly, daily for 15 days.

PROGRESS AND FOLLOW-UP

At the time of discharge on 25.4.72, the ulcer on the right fore-arm had completely healed, but abnormal

movements above the wrist persisted; livedo reticularis had also completely cleared.

The family was seen again in February, 1974. Except for the persistence of abnormal movements above the right wrist, the child was in perfect health. His VDRL was non-reactive. Radiography of the right fore-arm showed absorption of the lower half of radius with cortical thickening of ulna and deformity at the wrist. Unruptured upper incisors appeared radiologically normal.

During this period, the mother had had a second pregnancy and delivered at full term a healthy female child in November, 1973. This child had no evidence of congenital syphilis and her serum VDRL was non-reactive. Mother's VDRL was reactive at 1:32. Father's STS was again non-reactive.

The mother was seen subsequently in May, 1976 with her third child, then 2 months old, the outcome of her third pregnancy. The mother's STS (VDRL) was reactive again at 1:32. The infant appeared in perfect health and its VDRL was non-reactive.



Fig. 3. Radiograph (taken on 5.4.72) of the right fore-arm of the infant showing dislocation of right infra-radio-ulnar joint with fractures in shafts of radius and ulna. Marked periosteal reaction and sequestrum formation also seen in both bones.

DISCUSSION

Fournier (2) and Finger laid down stringent criteria for the diagnosis of third generation syphilis. According to them, acquired syphilis must be demonstrated in the grandmother and preferably also in the grandfather. But we agree with Ogilvie that the evidence of syphilis in the grandparents should be considered of corroborative rather than of absolute value and it is not essential to demonstrate syphilis in one or other grandparent, as it is undoubtedly possible to diagnose congenital syphilis in a parent by its pathognomic signs and symptoms. The presence of classical Hutchinsonian teeth and open-bite deformity along with reactive serum VDRL at 1:64 constitutes undisputable evidence of congenital syphilis in the mother. Nabarro (5) also observed typical Hutchinsonian teeth in mothers of 15 out of 42 cases of third generation syphilis and considered them one of the most important single diagnostic signs of congenital syphilis. Moreover, in the present case the mother's VDRL has persistently been reactive in dilutions ranging from 1:64 to 1:32, even 4 years after adequate therapy. This serum-fastness is further evidence of long-standing infection in the form of congenital syphilis in the mother. It is well known that positive STS in recently acquired infection or super-infection promptly reverts to negative after adequate therapy. This, consequently also excludes the remote possibility of super-infection of the mother during her adolescence. It is true that the serum VDRL and FTA-ABS tests on the grandmother and VDRL of her 2 children are non-reactive, but is a moot point whether her 2 other children, who could not be examined, are also free from syphilis. It is also a well recognized fact that a syphilitic mother need not necessarily transmit the infection to each of her offspring. Further, several cases are on record, where, in cases of both uniovular and binovular twins, one of the twins has escaped infection (6, 7, 12). How far syphilis was responsible for causing marasmus and death in the four infants of the grandmother is again speculative, but cannot be excluded. Bruusgaard's study revealed that 27.9% of untreated syphilitics have spontaneous cure and even the blood STS becomes non-reactive; it is possible that the grandmother with non-reactive STS belongs to this group: of the 14 cases of third generation syphilis reported by Dennie & Pakula (1), the STS of the grandmother was non-reactive in

one. Seronegative maternal (11) and Ocular syphilis (10), using modern treponemal tests are also well documented and the grandmother with non-reactive FTA-ABS tests is another instance of this anomaly. According to McLean (4) bony lesions in syphilitic fetuses are always widely distributed and in the first month of life, the lesion of a single bone is never due to syphilis. McLean also held the view that osteomyelitis and periostitis never presented alone during the neonatal period and were always associated with osteochondritis. The infant reported here presented with severe syphilitic osteomyelitis leading to pathological fractures of radius and ulna, completely sparing the rest of skeletal system. The radiograph also showed the presence of sequestrum in both radius and ulna, which again is a very unusual feature. The negative darkfield examination of the osteomyelitic lesion does not exclude syphilis. Sing et al. (9) reported a case of infantile congenital syphilis with orchitis and hyperostotic lesions confined to the upper ends of both humeri only. Both livedo reticularis and the ulcer on the right forearm promptly resolved with anti-syphilitic treatment. Livedo reticularis as a manifestation of congenital syphilis is very infrequently seen.

ACKNOWLEDGEMENTS

Thanks are due to the Institute of Venereology, Government General Hospital, Madras, for performing the FTA-ABS test on blood of the grandmother.

REFERENCES

1. Dennie, C. C. & Pakula, S. F.: Congenital Syphilis, p. 437, Lea & Febiger, Philadelphia, 1940.
2. Fournier, A.: L'hérédité syphilitique (ed. Portalier), Paris, 1891.
3. Lampert, F. & Mayer, H.: The transmission of congenital syphilis to the third generation. *Med Klin* 60: 1526, 1965.
4. McLean, S.: The roentgenographic and pathologic aspects of congenital osseous syphilis. *Amer J Dis Child* 41: 130, 363, 1931.
5. Nabarro, D.: Congenital Syphilis, p. 392, 393. Arnold Publ. London, 1954.
6. Penrose, L. S.: Congenital syphilis in a monovular twin. *Lancet* i: 322, 1937.
7. Rajam, R. V. & Rangiah, P. N.: Prenatal syphilis in one of twins. *J Ind Med Ass* 27: 186, 1952.
8. Rutherford, H. W.: Two cases of third generation syphilis. *Br J Vener Dis* 41: 142, 1965.
9. Singh, R., Kaur, D. & Parmeswaran, M.: Infantile congenital syphilis presenting with bilateral orchitis. *Br J Vener Dis* 47: 206, 1971.

10. Smith, J. L.: The current status of ocular syphilis. *Surv Ophthalmol* 14: 176, 1969.
11. Tinkler, A. E.: Sero-negative maternal syphilis. *Br J Vener Dis* 44: 136, 1968.
12. Wolk, E.: Congenital syphilis in identical twins with dissimilar serologic reactions. *Arch Dermat Syph* 43: 491, 1941.

Received February 1, 1977

R. Singh, M.D.
Department of Dermatology & Venereology
Maulana Azad Medical College & Irwin Hospital
New Delhi-11 0002
India