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INFANTILE CONGENITAL SYPHILIS – A RARE CASE REPORT

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ABSTRACT

The syphilis, sexually transmitted disease very rarely observed in infantile stage. A twenty five days old preterm male child with history of vaginal delivery presented with complaints of abdominal distension, low-grade fever and excessive crying. VDRL was reactive for both mother and infant. Radiograph supported osteochondritis.

KEYWORDS: Congenital syphilis, infant, VDRL, radiography.

INTRODUCTION

The sexually transmitted disease - Syphilis is a systemic human disease due to *Treponema pallidum* and it is classified as acquired or congenital. Acquired syphilis (usually by sexual contact) is divided into early and late syphilis. Early syphilis includes primary, secondary and early latent syphilis.[1] Congenital syphilis is a syphilitic infection and rare disease that occurring during intra-uterine life through transplacental transmission from infected mother where the primary skeleton presentation is also unusual. It is divided into early (first 2 years) and late including stigmata of congenital syphilis. The recognition and prompt treatment are necessary to avoid devastating consequences. Differential diagnosis includes trauma, Caffey's disease, scurvy and hypervitaminosis D.[2]

Although the rate of congenital syphilis is declining in developing countries, a significant increase has been observed in the underdeveloped countries,[3] inspite of the widespread use of penicillin to treat syphilis since the early 1950s. Late congenital syphilis (recognized 2 or more years after birth) is a very rare clinical entity.[4,5] Estimated cases of congenital syphilis is about 25-75% of exposed infants and about 10-12% of infants born to infected mother will die if not treated.[6] Incidence of congenital syphilis is on increase at Eastern Europe and

central Asia.[7] In Africa, the reported sero-positivity is 3-18%. From Delhi 82 (1965-1978) and 7 (1980-1990) were reported.[6] We reported here a case of early congenital syphilis who presented at twenty five days preterm child with abdominal distension and fever.

Case presentation

A twenty five days old preterm male child with history of vaginal delivery presented with complaints of abdominal distension, low-grade fever and excessive crying. On general examination there was no pallor, jaundice, lymphadenopathy, maculopapular rash or hepatospleenomegaly.

Radiograph of both lower limbs (Fig-A) was done which shows bilaterally symmetrical multiple areas of erosion and destruction of the metaphyses of distal femur suggestive of Osteochondritis. Laboratory investigations showed normal hemoglobin, WBC, platelet counts and fundus examination revealed no abnormality but VDRL test for the patient and the mother were reactive.

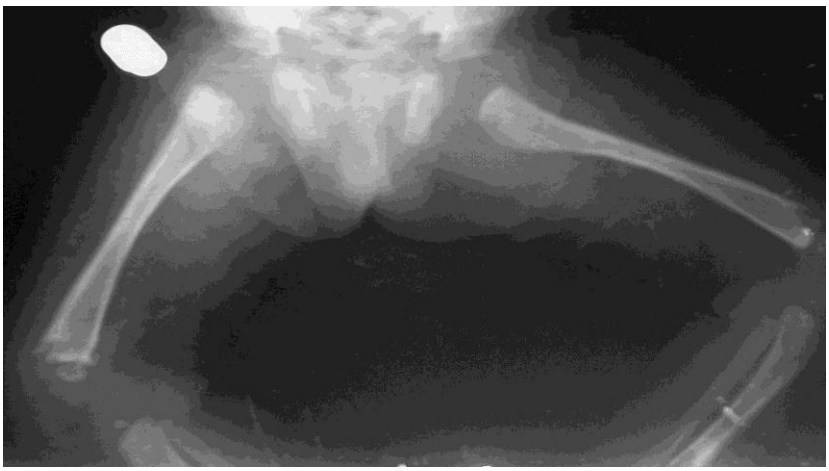

Figure 1: Plain radiograph of both knee joints

[The radiograph of both knee joints showing irregularity and sharp projection from the distal metaphysis of bilateral femur (osteochondritis). Solid periosteal reaction (periostitis) seen surrounding the distal metaphysis of right femur].

The radiographic features of the disease may mimic many other conditions such as multifocal osteitis, rickets, scurvy, neuroblastoma and battered baby syndrome. The treatment suggested are long acting benzyl penicillin G of 2.4 million units as first choice of treatment followed by doxycycline, is the preferred tetracycline with good penetration into the CSF), and erythromycin, both taken orally. Penicillin allergy or parenteral treatment also refused where

doxycycline 200mg daily (either 100 mg twice daily or as a single 200 mg dose) orally for 14 days and azithromycin 2g orally single dose are given.

Because of improved ante natal care, incidence has been brought down. Even then congenital syphilis infection occurs, necessitating that the antenatal screening should be strengthened. Good antenatal screening can prevent this preventable disease. Antenatal screening is easily done with blood VDRL test, a cheap, easy to perform, do not need sophisticated equipments and can be done in small labs. Antenatal screening for syphilis is mandatory because adverse pregnancy outcomes are 12 times higher. Further to prevent scars and deformities of congenital syphilis timely treatment right at the time of pregnancy is important.

DISCUSSION

The one way of transmission of infection is through transplacental circulation where the risk depends on the stage of the disease and the rate of transmission is 30%.[8,9] If the primary infection in the mother occurs longer time before pregnancy, the outcome is benign with respective rate and severity of infection. The risk of transmission is highest with primary and secondary syphilis during pregnancy and diminishes as the duration of latent syphilis increases.[9] The outcome of the pregnancy in untreated primary and secondary syphilis are 25% risk of still birth 14% risk of neonatal death, 41% risk of alive baby with infection and 20% baby without infection.[10] In untreated late syphilis, the risk is reduced with 12% of still birth, 9% of neonatal death, 2% of infected child and 77% chance to give birth of normal child. Women who manifest the disease after delivery may also transmit the infection.[9,10]

In case of congenital syphilis the disorder originates from transplacental migration of the Treponemas and invasion of the perichondrium, periosteum, cartilage, bone marrow, and sites of active endochondral ossification.[11] The spirochetes inhibit osteogenesis. In general the clinical presentation of the congenital syphilis showed signs of congenital syphilis in a neonate including a bullous rash, anemia, jaundice and hepatospleenomegaly.[12] The infant is often small for dates and may have feeding difficulties. More commonly the syphilitic infant appears normal at birth, and presents in the first three months of life with: failure to thrive, a rash resembling that of secondary syphilis, with desquamation involving palms and soles, persistent nasal discharge; and anemia or hepatosplenomegaly.[13,14]

Late congenital syphilis in a child or an adolescent corresponds to tertiary syphilis in an adult. Manifestations include hutchinsonian triad, consisting of Hutchinson's teeth, interstitial

keratitis, and nerve deafness may appear. Additional manifestations include fissuring about mouth and anus, anterior bowing of the lower leg, saddle nose, and perforation of palate.[9] Congenital syphilis has been divided on the basis of the presentation of the child clinically as well as radiological manifestation as early and late congenital syphilis. The late congenital syphilis in as child or adolescent corresponds to tertiary syphilis in an adult and radiological changes resemble the changes observed in acquired syphilis. Bone disease, although primary skeletal involvement is rare[2] and usually asymptomatic, it is the most common early manifestation. It is a bilaterally symmetrical polyostotic condition which affects mainly tubular bones, but any bone may be affected. The bones of the lower limbs are most commonly affected.[11]

The outstanding feature of syphilitic osseous disease is the presence of multiple bone involvement, and the predominant skeletal lesions are metaphysitis and periostitis. Metaphysitis is seen in 76.3%. There are two forms of metaphyseal involvement; altered mineralisation at the zone of provisional calcification and frank bone destruction. Areas of calcification may be increased at the growing zones and present as single or double transverse densities. Periostitis of the long bones, with or without metaphyseal abnormalities, is radiologically evident in more than 90% of cases, and may present clinically as pseudoparalysis of one or more limbs. The prognosis is very much better in those presenting in the postneonatal period. Periostitis is seen in 69.1% of cases. Osteitis is noted only in 6.0% cases.[9,11]

Some 30% of the cases had combined lesions of metaphysitis, osteitis and periostitis. The periosteal reaction was mild (a single layer) or severe in which a lamellar form also known as cloaking ‘coffin formation’ or the ‘periostitis of Pehu’. Periostitis can relate to infiltration by syphilitic granulation tissue. The long bones and, less commonly, the flat bones are affected.[14] Reparative periostitis is a second variety of periosteal response that is noted about healing foci of osteochondritis. Metaphyseal lesions were either productive or destructive and appeared as lucent or dense bands. Alternating bands produced a sandwich appearance on radiographs.[6,9] The epiphysealmetaphyseal junction of tubular bones, the costochondral regions, and in severe cases, the flat bones is affected. In the growing metaphysis of the long bones, widening of the provisional calcification zone, serrations, and adjacent osseous irregularity are seen. Erosive lesions develop along the contour of the bone at the metaphyseal-growth plate junction.[3] Erosions of the metaphysis were common and

symmetrical erosions in the upper medial tibia (Wimberger's sign). Metaphyseal serrations, the sawtooth appearance of the metaphysis is called as Wegner's sign.[3,14]

Diaphyseal osteomyelitis (osteitis) relates to the extension of granulation tissue. Osteolytic lesions with surrounding bony eburnation and overlying periostitis are seen. Osteitis commonly presented as a diffuse moth-eaten rarefaction of the shaft.[15] The other radiographic osseous features are joint changes, pathological fractures, dactylitis and skull lesions. Osteitis commonly presented as a diffuse moth-eaten rarefaction of the shaft. The other radiographic osseous features are joint changes, pathological fractures, dactylitis and skull lesions. Fractures through a defect in the shaft and joint involvement are also common. Joint changes include effusion and subluxation of shoulder, knee and hip. Erosion of the articular surface of the sigmoid notch of the ulna; this was the only articular surface involved.[16] Syphilitic dactylitis are rare. Skull lesions may be purely sclerotic or may present as a combination of sclerosis and osteolysis. In purely sclerotic lesions, new bone may be laid down in the frontal and parietal regions, so producing the 'hot cross bun' skull.[15,16] As per the case mentioned in this study, there are varied radiological presentations of congenital syphilis. The above-mentioned patient presented with radiological features of Osteochondritis - bilaterally symmetrical with periostitis.

REFERENCES

1. Walker GJ, Walker DG. Congenital syphilis: a continuing but neglected problem. Semin Fetal Neonat Med, 2007; 12: 198-206.
2. Toohey JS. Skeletal presentation of congenital syphilis: case report and review of literature. J Pediatr Orthop, 1985; 5: 104-6.
3. Gurlek A, Alaybeyoglu NY, Demir CY. The continuing scourge of congenital syphilis in 21st century: a case report. Int J Paediatr Otorhinolaryngol, 2005; 69: 1117–21.
4. Larsen SA, Steiner BM, Rudolph AH. Laboratory diagnosis and interpretation of tests for syphilis. Clin Microbiol Rev, 1995; 16: 1-21.
5. Monica C, Bineeta K, Preena B. Congenital syphilis, still a reality in 21st century: a case report. J Med Case Rep, 2007; 1: 90-4.
6. Phiske MM. Current trends in congenital syphilis. Ind J Sex Transm Dis, 2014; 35: 12-20.
7. Herbert LJ, Middleton SI. An estimate of syphilis incidence in Eastern Europe. J Global Hlth, 2012; 2: 402-7.

8. Vasquez MO, Dickson GSM, Salas JG, Rodriguez MAJ, Arria M. Congenital syphilis in Valera, Venezuela. J Trop Pediatr, 2007; 53: 274-7.
9. Sena AC, Muth SQ, Heffelfinger JD. Factors and the sociosexual network associated with a syphilis outbreak in rural North Carolina. Sex Transm Dis, 2007; 34: 280-7.
10. Genc M, Ledger WJ. Syphilis in pregnancy. Sex Transm Infect, 2000; 76: 73-9.
11. Sharma M, Solanki RN, Gupta A, Shah AK. Different radiological presentations of congenital syphilis: four cases. Ind J Radiol Imag, 2005; 15: 53-7.
12. Kim JK, Choi SR, Lee HJ, Kim DH, Yoon MS, Jo HS. Congenital syphilis presenting with a generalized bullous and pustular eruption in a premature newborn. Ann Dermatol, 2011; 23: 127-30.
13. Singh AE, Romanowski B. Syphilis: review with emphasis on clinical, epidemiologic and some biologic features. Clin Microbiol Rev, 1999; 12: 187-209.
14. Karthikeyan K, Thappa DM. Early congenital syphilis in the new millennium. Pediatr Dermatol, 2002; 162: 197-9.
15. Rasool MN, Govender S. The skeletal manifestations of congenital syphilis. J Bone Joint Surg, 1989; 71: 752-5.
16. Wantanabe A, Felipe S, Peter SV, Blazar P, Hiroshi Y. Ulnar sided wrist pain. II. Clinical imaging and treatment. Skeletel Radiol, 2010; 39: 837-57.