Hutchinson's Teeth : A late clinical manifestation of congenital syphilis

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Background

Syphilis the infection caused by *Treponema pallidum* is increasing globally; leading in turn to an increase in the incidence of congenital syphilis (CS). CS is classified as (i) early or (ii) late depending on whether symptoms appear before or after 2 years of age. Early CS is a multisystem illness with snuffles, rash, skin desquamation, hepatomegaly and long bone abnormalities. Signs of late CS include Hutchinson's teeth, hearing loss and neurodevelopmental delay. Syphilis serology is routinely performed at the first antenatal appointment and includes Rapid Plasma Reagin (RPR), Treponema pallidum particle haemagglutination assay (TPHA), and *Treponema pallidum* IgM. We present two cases of CS treated at Rotunda Hospital Dublin, with characteristics of Hutchinson's teeth, a rare late manifestation of CS.

Case 1

A male infant born at 37+4 gestation delivered to a woman with no antenatal care. Maternal serology at delivery revealed positive TPHA and RPR (titre 1:16). Infant clinical evaluation was significant for growth restriction, excess nasal secretions, respiratory distress and skin desquamation. Infant serology showed RPR positive (titre 1:8). CSF RPR was also positive. Radiology demonstrated long bone lucencies and classic findings of CS (Image 1A&B).
The infant was treated with 10 days IV benzylpenicillin. Follow up RPR was negative at 6 months, but TPHA remains persistently positive after 18 months, confirming congenital syphilis.

At age 4 years dental abnormalities were noted including; peg shaped incisors and mulberry molars, features of Hutchinson's teeth (Image 2).

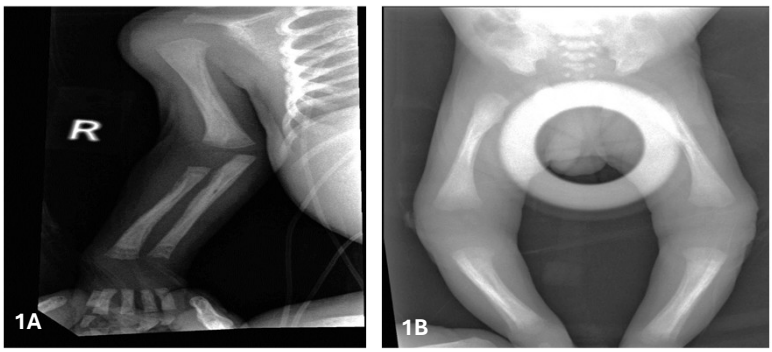

Image 1A: Metaphyseal lucencies in the distal humerus and 'sawtooth pattern' in right distal radius. 1B: metaphyseal lucencies in the distal femora and thick periosteal reaction in the diaphyses of the femora and tibia, pathognomonic for congenital syphilis.

Image 2: Abnormal dentition at 4 years old including peg shaped incisors and mulberry molars (noted as molars with extra cusps).

Case 2

A male infant born at 33 weeks gestation delivered to a woman who presented for antenatal care at 31 weeks gestation. Maternal serology showed a positive TPHA, RPR (titre 1:32), and positive *T. pallidum* IgM . She was treated with one dose of IM benzathine penicillin.
Infant clinical evaluation showed growth restriction, desquamating skin, petechiae, hepatosplenomegaly and respiratory distress. Infant serology at birth had a positive TPHA and RPR (titre 1:32), negative RPR on CSF. Radiology demonstrated long bone lucencies and significant pneumonitis (Image 3A&B) consistent with CS.
The Infant was treated with 10 days IV benzylpenicillin. Follow up RPR was negative at 6 months; TPHA persistently positive at 18 months.
At age 3 years he presented with small, widely spaced teeth and notched incisors, features of Hutchinson's teeth (Image 4).

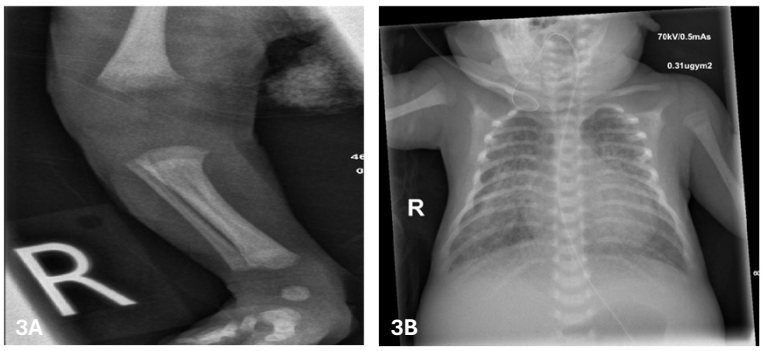

Image 3A: Long bone lucencies along metaphysis of distal femur, proximal tibia and fibula consistent with congenital syphilis. 3B: Chest Xray showing bilateral nodular density, confluent in right middle lung, consistent with pneumonitis of congenital syphilis.

Image 4: Signs of Hutchinsons teeth age 3: small, widely spaced teeth with notched upper incisors and abnormal pre molars.

Discussion

Hutchinson's teeth occur due to the invasion of *T. pallidum* proximate to the dental germ layers during fetal development. This results in 3 main dental abnormalities with peg shaped incisors (Hutchinson's incisors), Moon's molars (bud molars) and mulberry molars, seen in the patients described here (Images 2 and 4).

Hutchinson's teeth represent a rare manifestation of late CS. While most reported cases involve older children with undiagnosed and untreated CS, both patients described here had appropriate neonatal therapy and achieved negative RPR from 6 months of age. This suggests that CS can cause late sequelae despite adequate diagnosis and treatment in the neonatal period. Alternatively, these dental anomalies may point toward a different diagnosis which should not be evident in secondary teeth.