DISEASES OF BONE

Dept.of Oral Pathology
& Microbiology

Learning Objectives

At the end of the lecture student should be able to describe

- Clinical features, oral manifestations,

  radiographic features, histopathological features,&

  treatment of Cherubism & Cleidocranial Dysplasia

INTRODUCTION

- A fibro-osseous lesion of the jaws involving more than one quadrant that stabilizes after the growth period, usually leaving some facial deformity & malocclusion
- It is a rare developmental disease involving jaws

- Cherub meaning plump-cheeked little angels (Renaissance paintings)
- Autosomal dominant pattern of inheritance
- The clinical alterations typically progress until puberty , then stabilize & slowly regress.

CLINICAL FEATURES:

- Occur in children (mean age – 7yrs) but can be noticed as early as 1st yr of age. Milder cases may not be detected until the patients reaches 10-12 yrs of age.
- A painless bilateral expansion of the posterior mandible is the most common early manifestation.

- The bilateral bony expansion imparts a ***"chubby"*** facial appearance
- Extensive maxillary involvement causes stretching of the skin of the upper face to expose the sclerae. This results in an ***"eye upturned to heaven"*** appearance

- Lesion tend to involve ascending ramus & body of the mandible.
- Extensive bone involvement causes a marked widening & distortion of alveoli.

CHERUBISM

Fig. 8.23 Cherubism: (left) this child has the characteristic chubby appearance due to expansion of the mandible in the region of the angle; (right) ten years later, the appearance is entirely normal. Nevertheless, giant cell areas were still active and proliferated through an extraction socket.

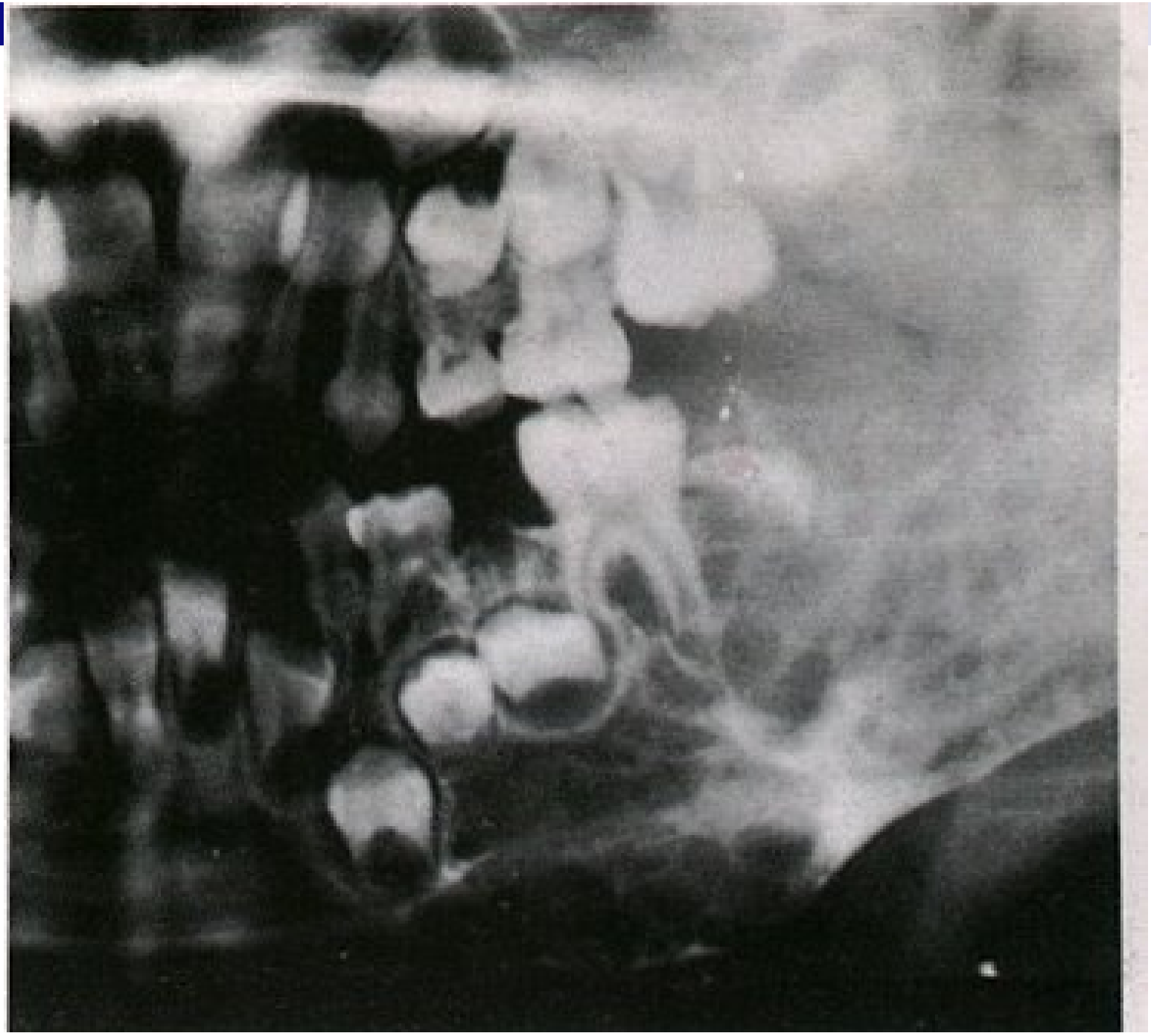
- All 4 quadrants may be simultaneously involved.
- Palatal vault may be obliterated.
- Premature exfoliation of deciduous teeth may occur as early as 3 yrs of age.
- Developing teeth are often **displaced** & **fail to erupt & may be malformed**

LAB FINDINGS:

- Serum Alkaline phosphatase levels may be elevated
- Serum Calcium & Phosphate levels are within normal limits

RADIOGRAPHIC FINDINGS:

- Multilocular expansile bilateral radiolucencies (rarely Unilocular)
- The borders are distinct & divided by bony trabeculae giving a **"Soap Bubble"** appearance
- Unerupted teeth are often displaced & appear to be floating in the cyst-like spaces- **'floating tooth syndrome'**



HISTOPATHOLOGY :

- Consists of highly cellular & vascular fibrous tissue containing variable numbers of ***multinucleated Giant cells.***
- The Giant cells tend to be small & are usually focally aggregated
- Blood vessels are numerous & typically surrounded by a cuff of ***eosinophilic fibrin-like*** material which appears to be perivascular collagen

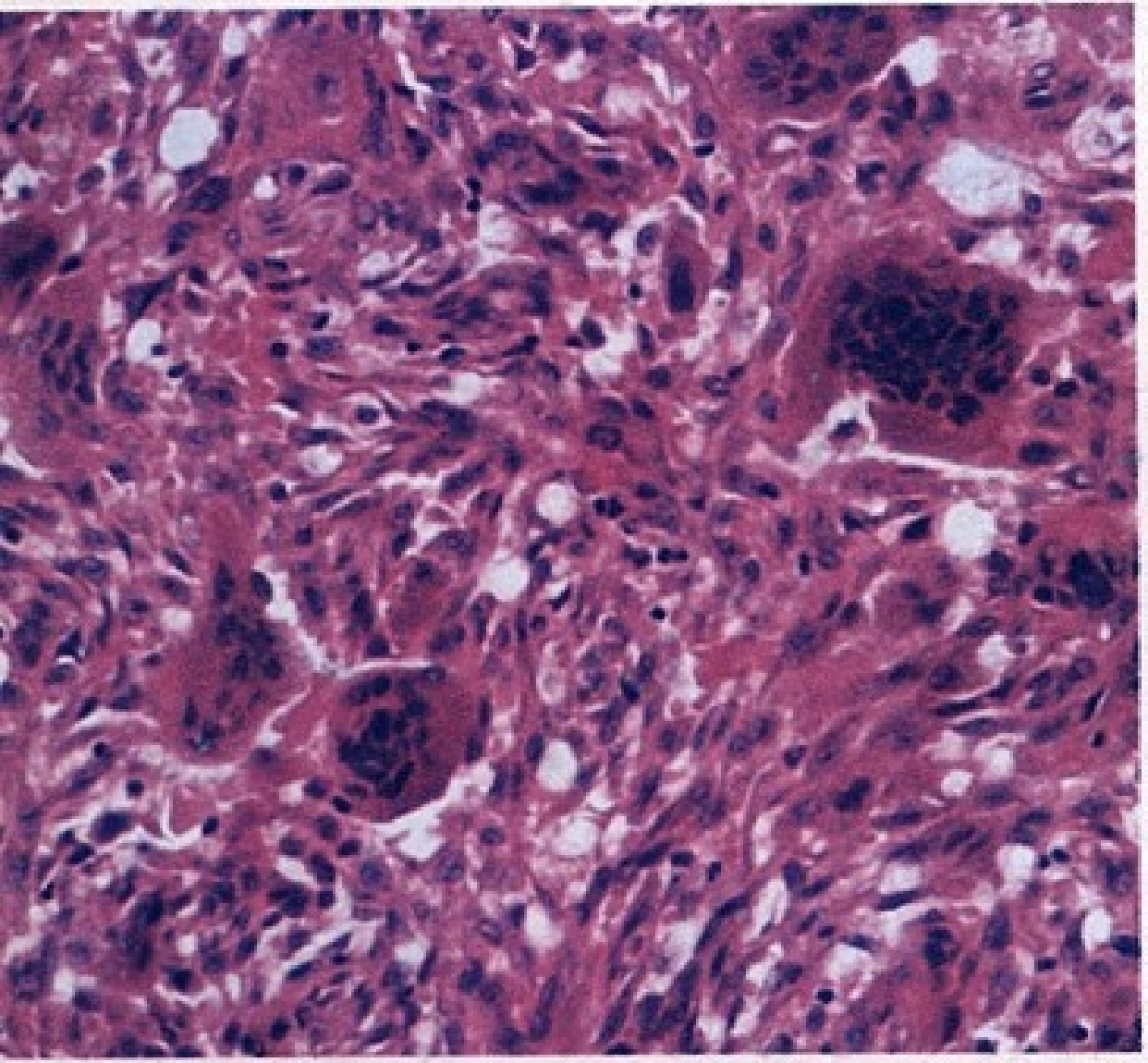


Fig. 8.25 Cherubism: characteristic giant cells in a spindle cell vascular matrix. These appearances are indistinguishable from hyperparathyroidism or central giant cell granuloma.

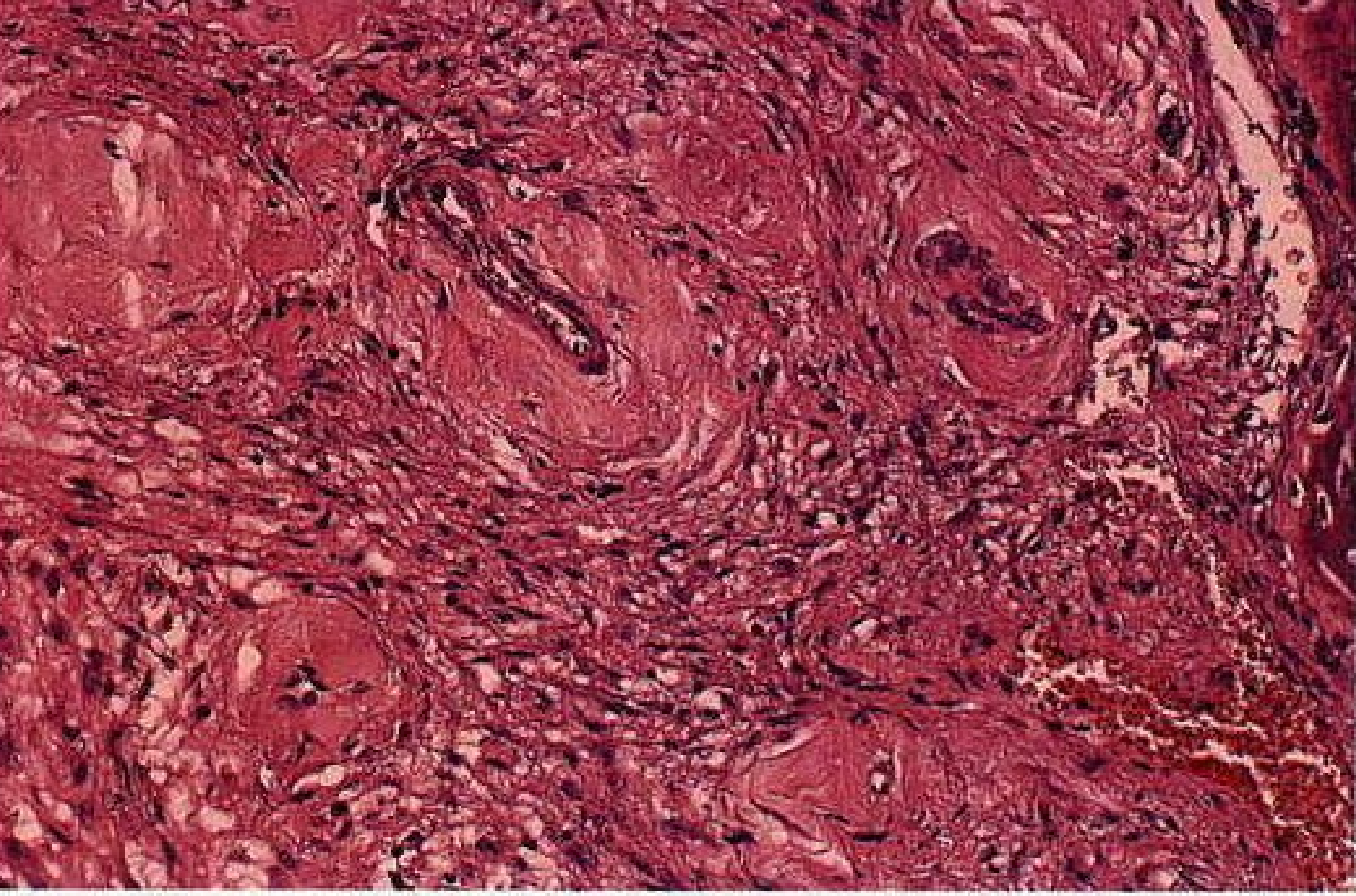


Fig. 17.3 Cherubism. Typical vascular cuffing in a fibrous area.

- Foci of extravasated blood are commonly present
- In long standing cases (resolving lesions) the tissue becomes more fibrous, the number of Giant cells decreases & new bone formation is seen in the form of small irregular bony trabeculae.

TREATMENT:

- Usually self limiting & regressive
- Cosmetic surgery for improving esthetics & function

*** The association between Cherubism & ***Noonan syndrome*** have been reported; but they are two separate diseases

CLEIDOCRANIAL DYSPLASIA
(Marie & Sainton's syndrome)

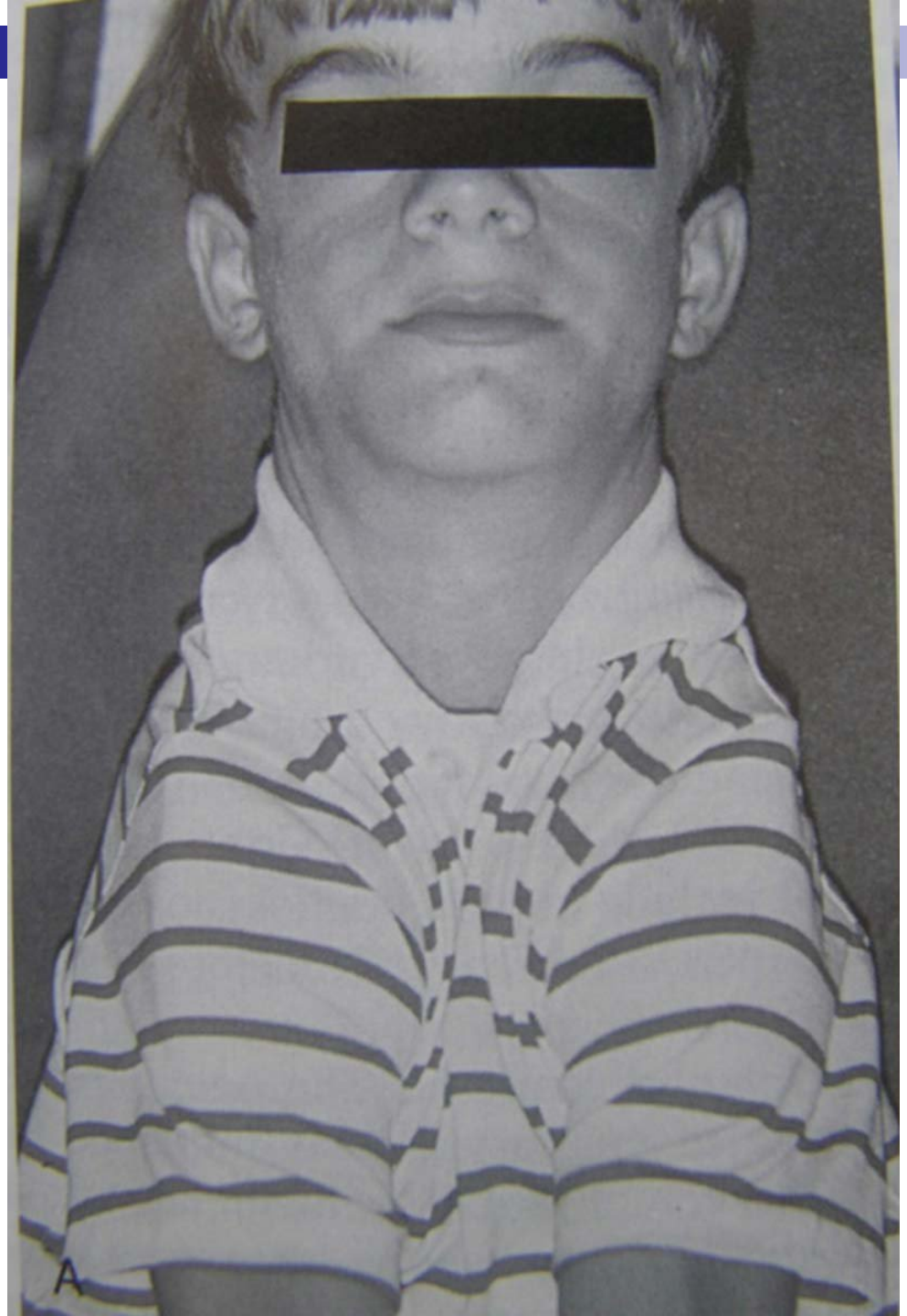
- It is a disease of unknown etiology which is often but not always hereditary.
- The disease shows an autosomal dominant inheritance pattern.
- Chiefly involved bones are ***skull*** & ***clavicles***, although a wide variety of anomalies may be found in other bones.

CLINICAL FEATURES:

- Short stature & have large heads
- Pronounced frontal & parietal bossing with underdeveloped or narrow paranasal sinuses.
- Ocular hypertelorism is frequently present.
- In the skull the fontanels often remain open or at least exhibit delayed closing.

- Sutures also may remain open & wormian bones are common.
- Mid-facial skeleton tends to be hypoplastic, and this combined with normal mandibular growth results in a relative prognathism.

- Patients neck is long, the shoulders are narrow & show marked drooping.
- Patients show an unusual mobility of their shoulders because of the **absence or hypoplasia of the clavicles**. (in some instances patient approximates the shoulders in front of chest)


A

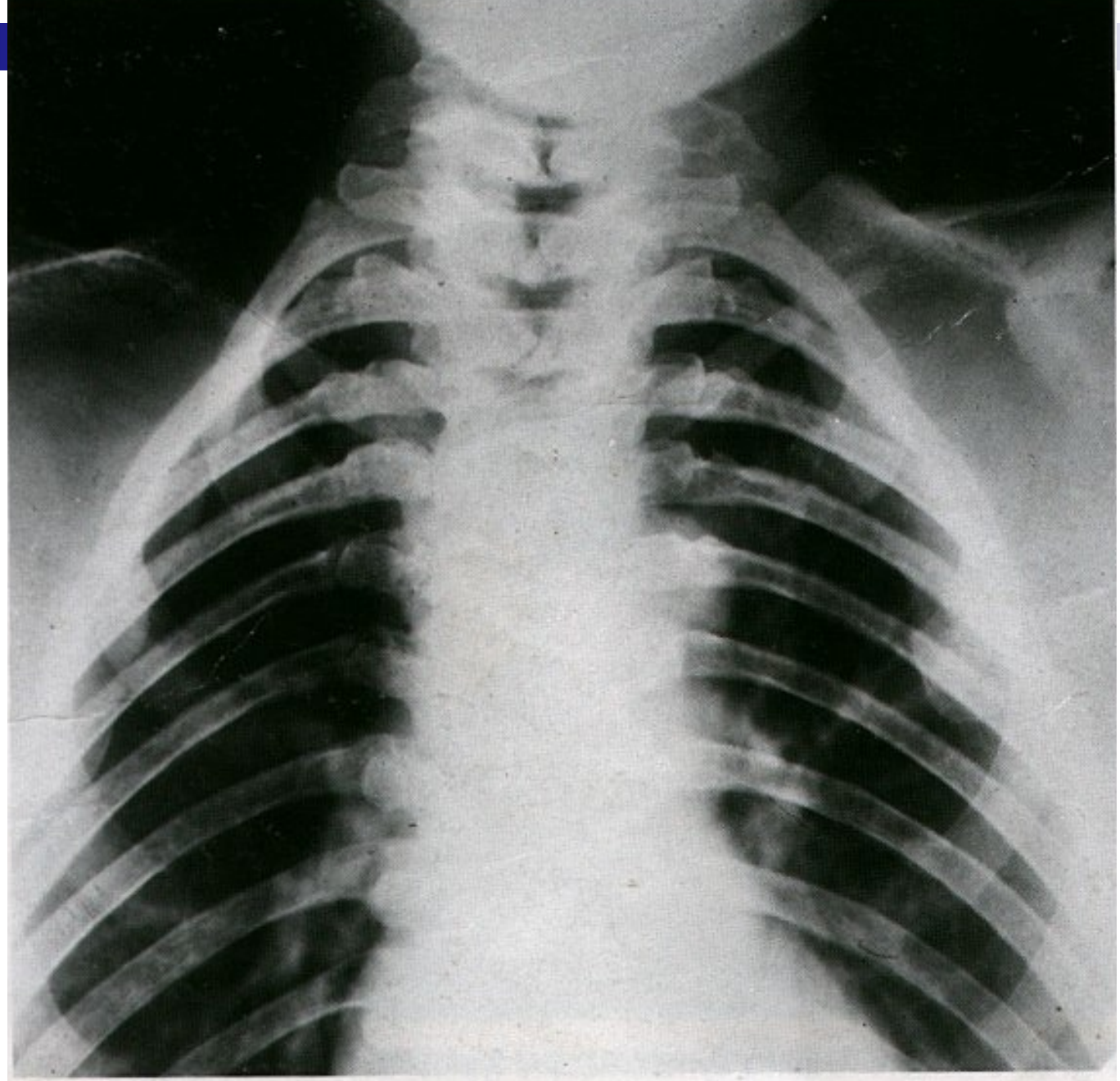


Fig. 2.24 Cleidocranial dysplasia: the radiograph shows absence of clavicles in this disorder.

ORAL MANIFESTATIONS:

- High, narrow, arched palate.
- Sometimes cleft palate is present
- Maxilla is underdeveloped
- Mandibular prognathism is seen
- Prolonged retention of deciduous teeth & subsequent delayed eruption of permanent teeth

- Numerous unerupted supernumerary teeth are present
- The roots of the teeth are often short & thinner than usual & may be deformed
- Absence or decreased amount of cementum is seen.

Fig. 2.25 Cleidocranial dysplasia: many buried teeth can be seen in his radiograph. Only a few had erupted

Summary

- Introduction,Clinical features, oral manifestations, radiographic features, histopathological features,& treatment of Cherubism & Cleidocranial Dysplasia

BIBLIOGRAPHY

- Text book of oral pathology Shafer's, 5 & 6th edition
- Oral & Maxillofacial Pathology A Rationale for Diagnosis & Treatment. R E Marx 1st edition
- Color Atlas of Oral Diseases Cawson, R. 2nd & 5th edition
- Oral and Maxillofacial Pathology Neville, Brad W. 2nd
- Lucas's Pathology Of Tumor's of the Oral Tissues

Thank You