Crouzon Syndrome : A Case Report and Review of Literature-



Fig 1: Photograph of the patient showing repaired bilateral cleft lip,malocclusion of teeth,wide parrot beaked nose,bilateral proptosis and esotropia.

1) First is the release of prematurely fused sutures based on evidence of raised intracranial pressure. Surgery is mainly carried out early after 3-6 months [5]. The principle is the release of bony ankylosis by exposure of fused sutures via a coronal flap.

2) Craniofacial reconstructive surgery including advancement of the maxilla and frontonasal complex; and other surgeries depending upon the deformities in the patient like rhinoplasty, oculoplasty and cleft lip and cleft palate repair can be done.

Early and accurate diagnosis of a patient of Crouzon syndrome is essential. Genetic counselling plays an important role. The need, extent and timing of treatment depend upon the severity of the disease and age of the patient. For complete evaluation, optimum treatment planning and comprehensive services, a multidisciplinary approach to the management of a patient of Crouzon syndrome is needed.

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Fig 2 : Photograph of the patient showing repaired cleft palate, dental malposition and maxillary hypplasia.

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Clinical Report

DENTIGEROUS CYST: A RARE PRESENTATION

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Abstract: Though Dentigerous cysts are encountered not so frequently by the otolaryngologist, a giant dentigerous cyst causing facial deformity and requiring a major reconstructive surgery is very rare. One such case is described.

Keywords: Dentigerous cyst, Facial asymmetry, Reconstructive surgery.

INTRODUCTION

The dentigerous cyst, also known as the follicular cyst is defined as a cyst which envelops the whole or part of the crown of an unerupted tooth and is attached to its neck. Majority of these cysts are discovered accidentally on routine radiographs,

otherwise they escape detection and enlarge to produce expansion of the jaw.⁽¹⁾ The present case is being reported because of its rare mode of presentation.

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Indian Journal of Otolaryngology and Head and Neck Surgery Vol. 58, No. 4, October - December 2006-

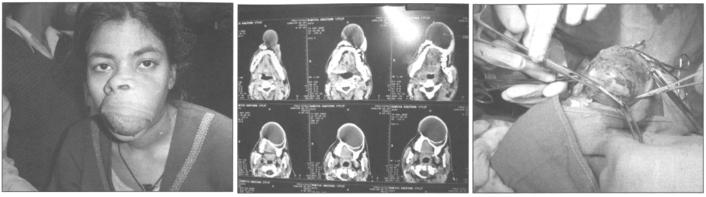


Fig-1: Clinical picture

Fig-II: CT scan picture

Fig-III: Intra operative picture

CASE REPORT

An 18 year old girl presented to the ENT OPD of Patna Medical College and Hospital with a large swelling in the left maxillary region with mild pain off and on for the last two years. About two years back she fell down from a tree and had trauma to teeth in upper jaw. A few days later she noticed a swelling in the upper jaw on the left side. The swelling progressed gradually, so she consulted a homeopathic doctor and had homeopathic medicines for some time. The swelling did not respond to the treatment and kept enlarging to such an extent that her face became distorted, so she came to ENT OPD, PMCH. There was no history of dysphagia.

On physical examination a large swelling was seen in the left maxillary region about 8x7cm in size grossly distorting the face, extending from the alveolar region below up to the lower eye lid above. Anteriorly it was protruding out of the upper lip for about 2cm (Fig-1). The nose was deviated to right and expanded by the mass. On anterior rhynoscopy the lateral wall and floor of the left nasal cavity was pushed by the swelling so that very narrow space was left in the nasal cavity. In the oral cavity the fleshy looking mass was protruding from the alveolar margin, there was no bulge of hard or soft palate. On palpation the mass was soft to firm in consistency in its anterior part while bony hard posteriorly. Temperature of overlying skin was not raised. The gingivolabial and gingivobuccal sulcus was obliterated by the swelling. There was no proptosis or facial nerve involvement. No significant cervical lynmphadenopathy was found.

CT scan of the skull showed a large expansile, non-enhancing, unilocular cystic lesion of 7.4x5.5 cm size, well marginated by cortical bone involving the alveolar ridge of left maxilla and distorting the adjacent anatomy (Fig-2). On the basis of clinical findings and CT scan the provisional diagnosis of odentogenic cyst was made and the patient was prepared for excision of the cyst under general anesthesia.

On the operation table incision was given on the mucosa overlying the swelling. Mucosa was separated from the cyst by blunt dissection (Fig-3). Now the cyst was aspirated. The aspirate was watery and straw colored. The cyst was dissected out completely. One tooth was found within the cyst (Fig-4). The antral wall was reshaped and intra nasal antrostomy was done. Excess tissue was cut and the wound sutured, Ryle's tube was inserted for feeding. On fourth post operative day two-three sutures slipped and a large oroantral fistula developed (Fig-5). The patient was discharged after one week and was called after one month for correction of the antral defect.

In the second stage operation, repair of the defect in anterior antral wall was done under general anesthesia. The incision was given on the projecting alveolar margin and mucosal flap elevated on the superior aspect. A curvilinear groove was made in the palatal bone and the bone was fractured and elevated superiorly, the elevated part now formed the anterolateral wall of the maxilla. The mucosal flap stitched with the superior part of mucosa. Ryle's tube feeding was given for one week. A small fistula developed in the post operative period that healed spontaneously otherwise the postoperative period was uneventful (Fig-6).

DISCUSSION

The dentigerous cyst is a common odontogenic cyst. It can occur at any age but has highest incidence in the third and fourth decades. It is more common in men than women. It originates after the crown of the tooth has been completely formed by accumulation of fluid between the reduced enamel epithelium and tooth crown.⁽²⁾ The cysts are twice as common in the mandible as in maxilla. The majority are associated with mandibular third molar then in decreasing frequency the maxillary canine, mandibular premolar and maxillary third molar, rarely they can be associated with supernumerary teeth. Radiographs show a radiolucent area associated with the crown of an unerupted tooth.

The cyst may be related to the crown of the tooth in three waysin central type, the cyst completely surrounds the crown of the tooth, in lateral type, the cyst projects laterally from the side of the tooth and dose not completely envelop crown, and lastly in circumferential type, the entire tooth appears enveloped by the cyst.⁽¹⁾

Histologically the cysts are lined by a layer of non keratinized stratified squamous epithelium. The lining is supported by fibrous connective tissue free from inflammatory infiltrate. The content is a protein rich yellow fluid with cholesterol crystals.

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Dentigerous Cyst : A Rare Presentation



Fig-IV- Excised cyst with a tooth

Fig.V- Residual defect after excision of cyst

Fig.VI- Post-op picture after 2nd operation

In children, best results are obtained by marsupializtion as enucleation may lead to damage to involved tooth. In adults the tooth is unlikely to erupt therefore enucleation together with removal of associated tooth is preferred⁽¹⁾. In the present case the cyst was very large and caused expansion of surrounding bones therefore enucleation was done in first stage and reconstruction in the second stage

CONCLUSION

It can be concluded that a simple dentigerous cyst can acquire a huge size and cause distortion of the face, so that it requires major reconstructive surgery for restoration of a normal face.

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Clinical Report

DISSEMINATED INTRAVASCULAR COAGULATION SYNDROME -ENT SURGEON'S PERSPECTIVE

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Abstract: ENT surgeon is seldom confronted with a systemic disease severe enough to progress to Disseminated Intravascular Coagulation syndrome (DIC), where he has to undertake a definitive surgical procedure for malignancy or perform an emergency tracheostomy. This calls for a judicious calculation of the pros and cons with an element of risk that needs to be accepted. We report two such patients who have been treated in such a scenario where the outcome varied depending on the severity and time of onset of the underlying coagulation disorder.

Keywords: Bleeding Disorders, Disseminated Intravascular Coagulation, Hepatitis B infection

Key for Abbreviations:

- DIC- Disseminated Intravascular Coagulation
- INR- International Normalized Ratio
- FFP- Fresh Frozen Plasma
- PT- Prothrombin Time
- PTT- Partial Thromboplastin Time
- APTT- Activated Partial Thromboplastin Time
- USG- Ultrasonography
- CT- Computed Tomography
- TFA- Tissue Factor Activity

INTRODUCTION

Disseminated intravascular coagulation is a condition characterized by widespread activation of coagulation process, which results in intravascular formation of thrombin and ultimately thrombotic occlusion of small and midsize vessels^[1,2].

CASE 1:

38 yr old male patient presented with complaints of difficulty in breathing for 4 months. He was asymptomatic till 6 months ago when he developed Hepatic Encephalopathy following Hepatitis

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Indian Journal of Otolaryngology and Head and Neck Surgery Vol. 58, No. 4, October - December 2006-