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RESEARCH ARTICLE

PINDBORG TUMOR PRESENTING IN EARLY ADOLESCENT- A RARE CASE REPORT

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Abstract

As infrequent as it may be, a calcifying odontogenic tumor As infrequent as it may be, a calcifying odontogenic tumor is more commonly seen in the late decades of life. It is a locally aggressive lesion that might interfere function by virtue of its expansile growth through the cortices of the involved jaw. This is a case report of Pindborg tumor in 11 year old male in the right mandibular body region and a rundown on the various investigations done and the final treatment plan adopted. A review of literature on the lesion and justification for the chosen treatment modality is included.

Introduction:-

Danish pathologist Jens J. Pindborg first described CEOT(calcifying epithelial odontogenic tumor) as a separate entity in 1958, as he reported a case series of three patients, two of whom presented with recurrent lesions. CEOT arises from the odontogenic epithelium and the remnants thereof, and though shows a wide range of occurrence, with reported cases as early as in 8th year of life to as late as 92 years, the peak incidence is noted around the 5th decade of life. The nomenclature is based on the histopathological (epithelial lined tumor) and radiographical (calcification) presentation of the lesion. Shafer et al coined the term Pindborgtumor.CEOT is a rare tumor accounting for 1% of all odontogenic tumors, usually seen in adults with only 7% of the reported cases occurring in children. Tumors reported in the pediatric group tend to have variable presentation either as an asymptomatic lesion found incidentally or appears as a painful growth. Radiologic features of CEOT also vary depending on the stage of the tumor. At an early stage, the tumor may present as a radiolucent unilocular or multilocular (soap-bubble) lesion, whereas as the lesion progresses, radiopacities increase. As the lesion has shown up to 14% local recurrence but a very benign indolent course of the primary lesion, the treatment modality ranges from simple enucleation to gross segmental resection and microvascularosteocutaneous flap reconstruction.[1]

Case Report

A11 years old male patient, presented to the department of Oral and maxillofacial surgery, with c/o painless swelling over right lower jaw since past one and a half months. Patient noticed a small pea sized swelling over the right lower front tooth region, in the lower vestibule, which was progressive in nature and had grown to larger dimension. The swelling on initial presentation was not associated with any symptom, however became painful and tender on palpation over time.

Pt was well nourished and moderately built. Had no other systemic conditions inhibiting treatment. Medical history was not contributory.

On extra oral examination, facial asymmetry was noted on the lower thirds of face, due to the localized swelling over the right mandibular body region. The swelling spanned approximately 3cms from midline of lower lip to mid

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body regionanteroposteriorly, and 2 cms in width Mediolaterally and 1.5cms in its height from the superior margin at the level of commissure of lip to inferior most margin at the lower border of mandible. The skin over the swelling was unremarkable.[Figure 1]

On palpation, the swelling was non-compressible, non-fluctuant and bony hard, associated with tenderness. The swelling was not fixed to the skin and had no local rise of temperature. No paresthesia noted over the distribution of inferior alveolar or mental nerve. There was no associated lymphadenopathy.

On intraoral examination, patient had sufficient mouth opening of about 44 mms. Pt was at his mixed dentition period and was noted to maintain a good oral hygiene. Swelling noted in lower right vestibule region, extending from central incisor to deciduous 2ndmolar region, obliterating the buccal vestibule. Mucosa was not breeched. No draining sinus was noted.[Figure2]

On palpation, it was hard in consistency, with no obvious areas of decortication. The swelling was tender on palpation, but not associated with paresthesia.No pus or signs of inflammation was noted. No tooth mobility, drifting, displacement or tenderness on percussion was elicited.

On clinical extra and intra oral examination, the lesion did not cross the midline.

On aspiration, scarce aspirate- which was mostly blood and mucous fluid, with no pus.

Investigation

An OPG was the initial radiographic investigation that was done. The radiograph revealed normal anatomy of all mandibular subunits, with a revelation of a mixed dentition status. The right lower body region showed a well-definedmixed radiolucent-radiopaque lesion, which was multilocular with coarse trabeculae and scattered foci of calcification. The lesion was enveloping an unerupted tooth and abutting other.[Figure 3]

Given the peculiarity of the lesion, we had decided to go ahead with further radio diagnosis with a CECT view. The well encapsulated nature of the lesion, with cortical expansion on the buccal side, with certain areas of erosion could be clearly demarcated. The lingual cortex was not expanded nor eroded by the disease process. The characteristic snow drivenappearance due to mixed radiopaque – radiolucent nature was evidentthroughout the lesion. An impacted tooth was noted in the substance of the lesion, clinically and radiographically corresponding to the permanent canine. The erupting first premolar lies in close approximation to the lesion. The inferior alveolar nerve canal was displaced inferiorly with no involvement of nerve in the lesion.

The CECT impression suggested a thin walled solitary expansileosteolytic bony lesion with tooth like structure in the floor along the inferior border of mandible, with no soft tissue and nerve involvement.[Figure 4]

A bone biopsy was done and the reports suggested a diagnosis favouring Calcifying epithelial Odontogenic Tumor based on the presence ofsheets of polyhedral epithelial cells in a fibrous stroma with prominent intercellular bridges with masses of calcification and hyalinised areas resembling amyloid like material seen amidst tumor islands.

Based on clinical, radiographical and histopathological correlation, a diagnosis of biopsy proven case of CEOT was established.

Treatment planning

Based on these finding a treatment plan was devised. All Routine investigations was done which includes chest X ray, 2D echo and complete blood picture. Paediatric and anaesthetic fitness for general anaesthesia was obtained. Keeping patients age under consideration, a minimalistic approach with thorough enucleation of the lesion under GA, with adjunctive chemical cauterization with a modified Carnoy's solution followed by PRP concentrate packed into the bony defect was done. [Figure5]

Surgical procedure

After the induction of GA, Nasal intubation was done and patient was painted and draped aseptically. LA infiltration using Lignocaine 2% with adrenaline (1:100000) was given at the lower vestibule region. Through an intraoral vestibular approach, with incision extending from 33 to 46, a mucoperiosteal flap was raised and the lesion exposed.

The buccal cortex was thin and had areas of fenestration, which was broadened with a knife. The well encapsulated tumor mass was exposed. It was slowly teased out, using a curette, being careful not to breach the lining of the mass. The permanent canine tooth was enucleated along with the tumor mass. The premolar teeth was exposed by the tumor process and hence was extracted as well. The inferior alveolar nerve was not exposed, meaning the canal was not deroofed by the disease process. Once the tumor was completely enucleated, the peripheral osteotomy procedure was undertaken using a mastoid bur. About 2mm bone was removed from all the bony walls. Next, a cotton pellet dipped in modified Carnoys solution was applied to all the bone wall, and left for 5 to 10 minutes. It was later thoroughly washed away with normal saline. After achieving haemostasis, Genta- beta wash was given. The patient's blood was withdrawn from antecubital fossa and PRF concentrate was packed into the defect. Water-tight closure was done using Vicryl 4-0. The patient was extubated and the post op period was uneventful.

Follow up

The patient is maintained on close follow up.[Figure6]The latest being 5 months post-surgery. Parameters including bone remodelling and paraesthesia over the operated site is assessed using clinical photographs and orthopantamographs. The patient had satisfactory healing over the intraoral incision site. The bony overgrowth underwent remodelling with improvement in facial symmetry over time. The patient had paraesthesia of mental nerve on the operated side, post-surgery. The areas of absolute anaesthesia was marked in each visit. The improvement in nerve disturbances was noted over time, with declining paraesthesia.

Discussion:-

Odontogenic tumours and tumour-like lesions constitute a group of heterogeneous diseases that range from hamartomatous or non-neoplastic tissue proliferations to benign neoplasms and finally malignant tumours with metastatic potential. CEOT is an epithelial odontogenic neoplasm, which is locally invasive and is characterized by the presence of amyloid material that may become calcified[2]. The first cases of CEOT was reported byThoma and Goldman in 1946 but it was then called as "adenoid adamantoblastoma". The calcifying epithelial odontogenic tumor (CEOT) was introduced to literature in 1992 by Dr. J JPindborg. In 1971 the term "calcifying epithelial odontogenic tumor" was generally accepted and adopted by the WHO.According to contemporary views, this tumour is considered to be a developmental disorder, arising from the remnants of the dental lamina epithelium.[3] Various histological variants including CEOT with cementum-like components, clear-cell CEOT (15 cases reported so far), CEOT-containing Langerhans' cells, combined epithelial odontogenic tumour (CEOT/AOT) and CEOT with myoepithelial cells are observed.What makes this tumor unique is its rarity in its occurrence, hardly comprising less than 1% of all benign odontogenic tumors of jaw, with a much less incidence in children.[4]The extra-osseous or peripheral variant is less common with an incidence of about 6% of all diagnosed cases, occurring more in the anterior gingival region, and diagnosed a little earlier than the intraosseous forms. CEOT has no gender predilection, and occurs equally in both the sexes. Premolar and molar regions are the commonest site of occurrence, although can occur at any site. Mandible is affected by intra-osseous lesions more frequently than maxilla, with a ratio of 2:1.The lesion can be a purely incidental finding owing to its asymptomatic presentation. Patients get alerted only when the slow growth produces large enough swelling for the patient to notice its presence intraorally, or when it causes facial asymmetry, paresthesia or occlusal disturbances due to drifting off of tooth.[5]

The larger or the mature tumour will be mixed radiolucent— radiopaque, although the early tumour may be completely radiolucent. CEOT is often associated with unerupted teeth. It may be unilocular and cystic in appearance. It can demonstrate a mixture of large and small multilocular spaces described as 'soap bubble' and 'honey comb' in appearance. The radiographic borders in almost all cases between surrounding tissues and tumour appear to be circumscribed and well defined. CEOT on CT examination demonstrating thinning and expansion of lingual and buccal cortical plates with well-defined mass containing scattered radiopaque areas of different size and signal intensity in mandible. Pindborgtumour on MRI reveals predominantly a hypointense lesion on T1-weighted images and mixed hyperintense lesion on T2-weighted images.[6]

The histologic feature pathognomic for the lesion is presence of sheets, nests and masses of polyhedral, eosinophilic epithelial cells which may show cellular abnormalities including giant cell formation and nuclear pleomorphism. Some cells increase in size and produce a homogeneous, eosinophilic, `amyloid-like' substance which may become calcified and which may be liberated as the cells break down. The true nature of the amyloid-like material is still unresolved.

Numerous surgical modality has been suggested, main determinants being size, location and general condition of patient. A small well circumscribed lesion can be treated with enucleation or curettage with adjunctive procedures of removing a thin layer of bone all around. Large tumors may require varying degrees of resection and various reconstructive modality based on the type of residual defect (from marginal resection to resection with continuity defect and disarticulation).

In one of the earliest review article, by C D Franklin and J JPindborg, they observed the treatment done for over 113 cases of CEOT, ranging from radiotherapy to seeding radium into the curetted area, both of which yielded no significant results. The initial treatment for most of the lesions which subsequently recurred has been conservative and has involved the use of curettage, enucleation, or simple excision. In a consideration of the recurrence rate for various treatments, it would seem that a marginal resection with a rim of apparently normal tissue (as determined radiographically or clinically at operation) would be most likely to meet with long-term success and should, therefore, be the treatment of choice.[7]

In the review article by H.P. Philipsen and P.A. Reichart, wherein 181 cases of CEOT cases were discussed, the authors noted that the clinically innocent behavior of the lesion can be used to the patients and surgeons benefit, and a mutilating procedures, such as wide resection or hemisection of the mandible, seem unwarranted. Enucleation with a margin of macroscopically normal tissue is, therefore, the recommended treatment for CEOT involving the mandible. Maxillary CEOT should, however, be treated more aggressively, as they tend to grow more rapidly and do not usually remain well confined. Generally, recurrences are rare. Five years is considered the absolute minimum follow-up period.[8]

The Carnoy solution is a substance used in the auxiliary treatment of maxillofacial tumors and cysts. It is applied to diminishing the recurrence rate of unicysticameloblastoma and KOT. The use of Carnoy solution is designed to eliminate remaining tumor tissue and promote a superficial chemically induced necrosis.

From various study, its noted that the recurrence of CEOT can happen as early as 5 to 6 months to as late as 18 years after primary treatment.

As per the collaborative study of 32 cases and review of literature by Dr José Alcides Almeida De Arruda et al, the overall prognosis of CEOT appears to be favorable, although extremely rare examples of aggressive or malignant CEOT have been described. The majority of cases of malignant CEOT have been reported in women with a mean age of 53.7 years. Occurrence in the mandible appears in almost 90% of reports. Metastasis to regional lymph nodes, lungs, and the vertebrae located close to the hip has been documented.

Conclusion:-

The treatment plan in our study is in line with most literature on the lesion, where in the primary occurrence is treated conservatively, with simple enucleation and adjuctives of peripheral osteotomy and carnoys cauterization. Reserving more aggressive treatment modality for only if the lesion recurs.

The main factors for this decision included the patients age and utilization of the excellentpeadiatric growth potential therefore and the fact that though locally aggressive, CEOT, is a rather well encapsulated indolent lesion, with a good prognosis. Given the lesion shows potential for recurrence, a close follow up, up to a minimum of 5 years is inevitable.

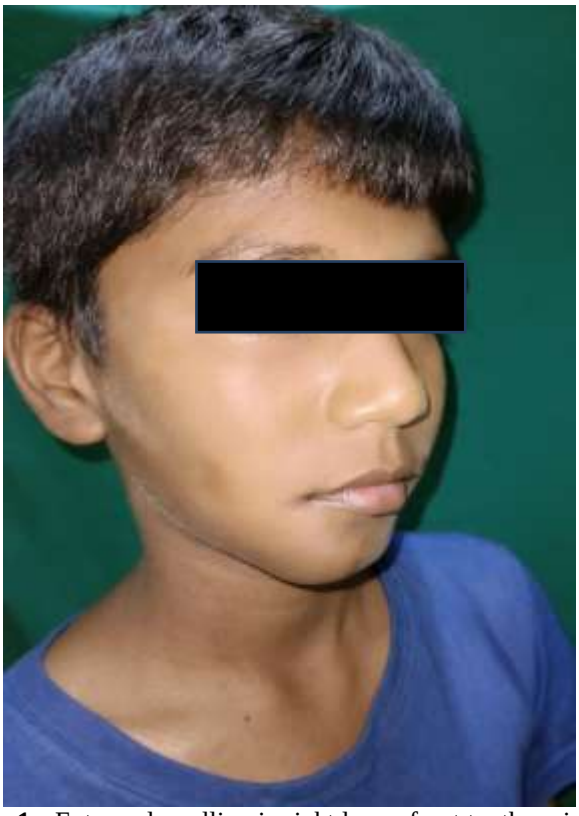

Fig 1:- Extaroral swelling in right lower front tooth region.

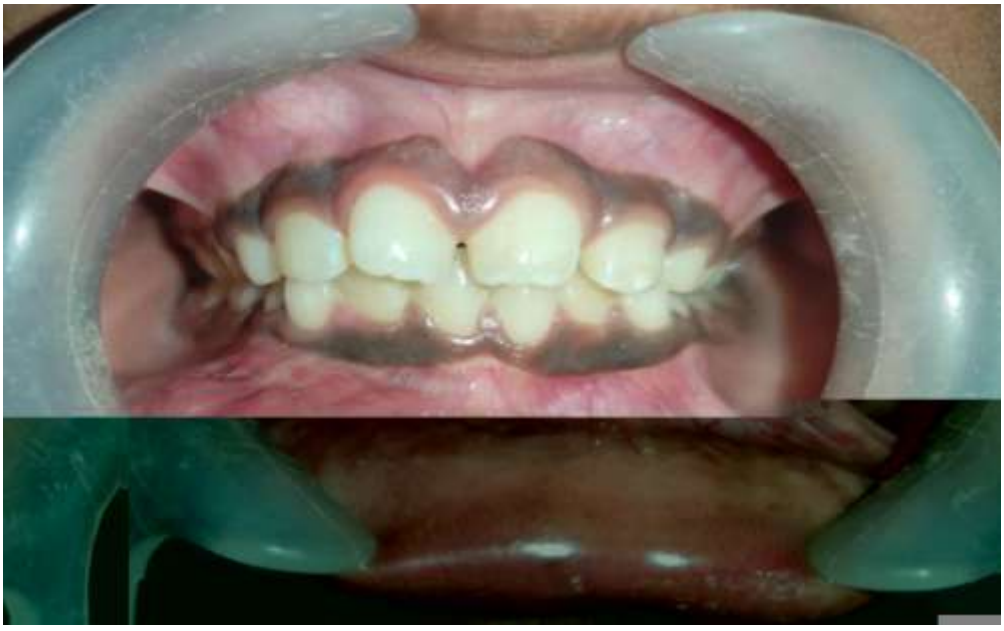

Fig 2:- Obliteration of right vestibule noted.

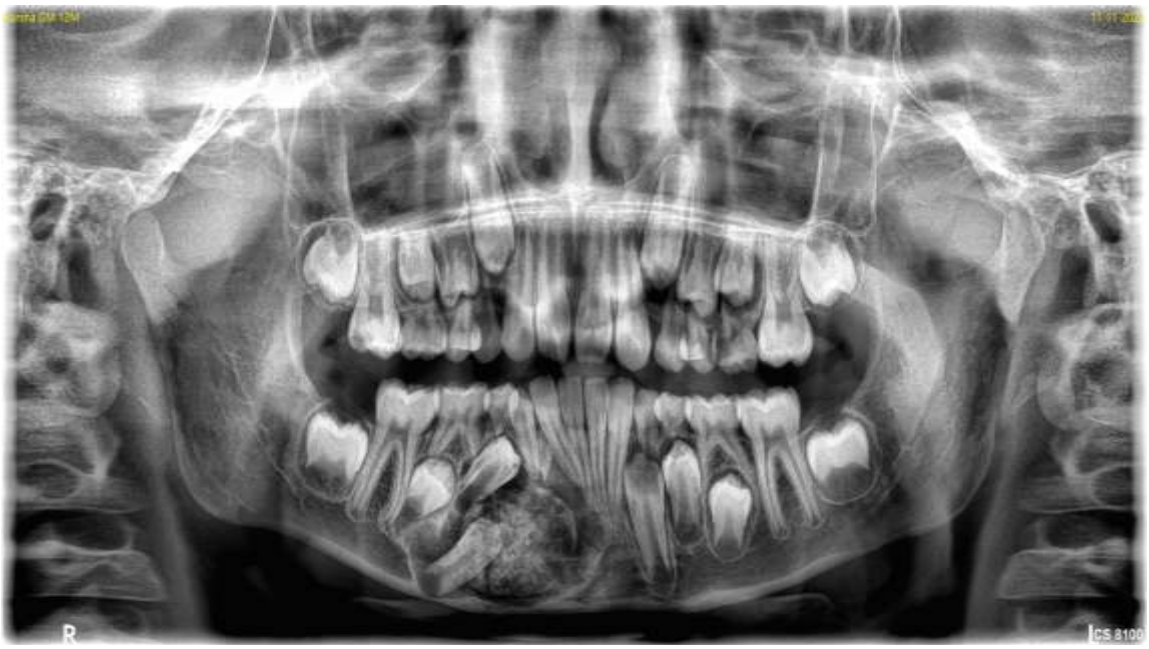

Fig 3:- OPG reveals mixed radiolucent radiooapque lesion right mandible.

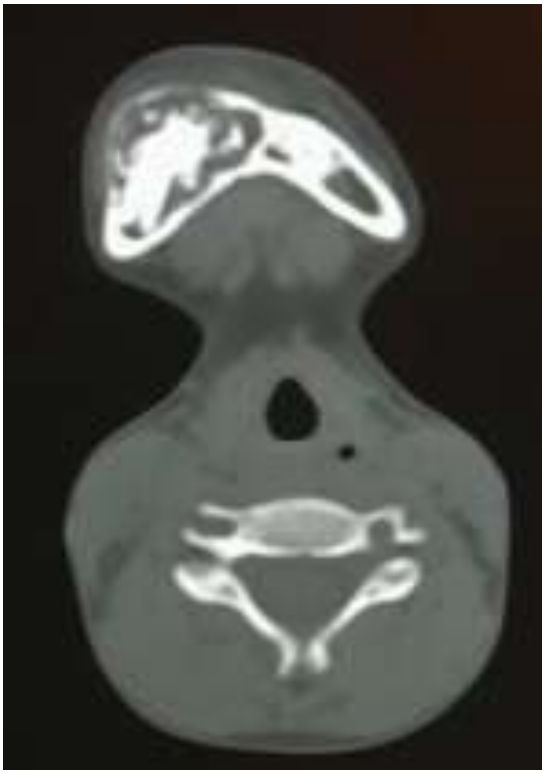

Fig 4:- CT reveals destruction of buccal cortical plate.

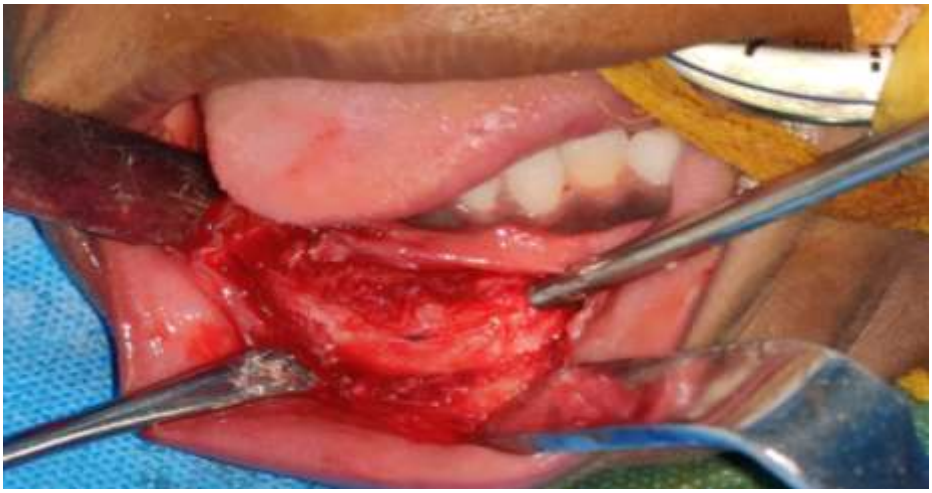

Fig 5:- Enucleation of the cyst.

Fig 6 a,b:- First follow up.

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