True Giant Cell Tumour of Maxilla

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INTRODUCTION

The giant cell tumors of jaw in general, include giant cell granuloma of peripheral and central type, cherubism, and brown tumor of hyperparathyroidism. The true giant cell tumor at an early age, is extremely rare. The relative rarity of incidence of this lesions is obvious from the Mayo Clinic Study by Austin et al (1959), wherein only two cases of true central giant cell tumor were seen as compared to 64 cases of giant cell granuloma. Cytogenesis of the multinucleated giant cells, so characteristic of these tumors, remains controversial. They are suggested to be derived either from endothelial cells or from the osteoclasts. Such tumors from bone, seldom if ever, involve any of the paranasal sinuses. At times the true giant cell tumor morphologically appears similar to, yet is clinically quite distinct from the granuloma. In view of its rarity such tumor in the maxilla of a child observed and successfully treated, is reported herewith.

Cash Report

This 6 years old male presented with progressive swelling of right upper alveolus and cheek for 14.50 months duration. He suffered with difficulty in mastication, when the mass suddenly increased within a weeks time to a sizable extent. There was no history of either trauma or bleeding. The personal and family history was also of no significance.

General examination revealed pallor and bad oral hygiene. There was bilateral submandibular and jugulo-digastric lymphadenopathy. These nodes ranged from 2 to 3 cms. in their maximum dimensions and were firm, mobile, tender and had raised local temperature. Otolaryngological examination revealed a dif-

fused non-tender swelling over the right cheek with the corresponding bulge over the alveolar region extending over the hard palate upto the midline. This bulge was tender above and covered with thin slough. There was no posterior extension beyond the maxilla and no mass was seen in the nasal cavity.

The routine blood counts and urinalysis were normal. The serum sodium, calcium and potassium were within the normal limits, but he had blood hemoglobin level of 7.0 mg%. The radiological examination of paranasal sinuses revealed only hazy right maxillary sinus with a soft tissue shadow, in relation to the upper alveolus.

The histopathological study of its biopsy revealed it to be composed of proliferated spindle shaped or elongated cells which were frequently intermingled with multi-nucleated giant cells. There was absence of collagen and osteoid material, suggesting it to be the true giant cell tumor. (Fig. No.1) There were no cells impinging closely to the vascular channels. Occasional haemosiderin pigment was also seen in stroma (Fig. No. 2)

The patient was put on hematinics and was also transfused blood to bring up the hemoglobin level up to 10 gm%. The lymphadenopathy responded well to a two week course of preoperative broad spectrum antibiotics. The tenderness and temperature disappeared and size was reduced to approximately 1 cm. Subsequently excision was done via a Danker's approach under general anaesthetic and endotracheal intubation. The growth was removed with the help of diathermy and sharp dissection including the alveolus, inferior maxilla, and homolateral palate almost up to the midline. The growth was found to be completely filling the maxillary antrum. After achiev-

Fig 1: *Histopathological examination showing abundant proliferating fibrous tissue amidst vascular spaces*

Fig 2: *High-power examination showing giant cells amongst proliferating fibrous tissue.*

ing complete haemostasis medicated packing of the cavity was done and the obturator was applied. The pack was removed after three days and the obturator was readjusted in the position. The patient was discharged with healthy healed cavity.

In follow up he revealed to have a recurrence within ten months at the same site. He had a facial swelling present over the right cheek extending up to the lateral border of nasal pyramid. Intraorally the mass was fleshy, non tender, bleeding on touch, covering the roof of antrum and extending to the ethmoidal region. The tumor was again excised from the ethmoids along with remnants of medial wall of maxilla. No dermal or muscle lining of the cavity was carried out at any time. The post operative phase was uneventful and the cavity healed very satisfactorily. The histopathological examination of this second specimen was essentially the same as the first one except for some more haemosiderin pigment signifying more bleeding. Several months follow up revealed the no recurrence though the cavity had somewhat contracted inspite the dental and hollow antral maxillary prosthesis. His health and general condition was very satisfactory till last follow up after about 3 years.

Discussion

The true giant cell tumor in a 6 years old child is

most unusual. These tumors are more commonly seen in patients over 20 years of the age. Only about 15% of the true giant cell tumors are observed in the first decade of life, particularly amongst the females. The mandible is the commonest site for giant cell granulomas, which though histologically similar, are not true neoplasms. The true giant cell tumors on the other hand involve both the upper (Hoffman et al, 1994) and the lower jaw. They have also been reported in skull base and temporal bone presenting with vertigo (McCluggage et al 1995).

The trauma is said to be as an aetiological factor for some of these tumefactions, but neither history of the injury nor any bleeding episode preceding its onset was reported in this case. The absence of the pain inspite of the massive bony expansion seen herewith is in favor of it to be the true giant cell tumor. Radiologically these giant cell tumors are said to be having a classical osteolytic and multiloculated appearance. Such picture was not present here instead with minimal bony destruction, a soft tissue mass was present, filling and expanding the maxillary antrum without any cystic area. This reflects to rather a rapidly growing potential of the tumor, which resulted in it attaining such a large size in shorter span of 14.05 months. Had it been slow growing lesion, the pseudo-ascinar appearance might have been expected.

The success of the first surgical procedure is the most important factor in determining the recurrence, in the larger series the recurrence rates range up to 34% after curettage and 7% following wide resection (McDonald et al, 1986).

The early and rapid tumor recurrence inspite adequate surgical excision, observed in this case, is also against it being a giant cell granulomas. Actually these latter tumefactions are satisfactorily treated only with simple local curettage. Its recurrence can not be assigned due to the surgical trauma, again reactivating it, as may sometimes be the case in the reparative granuloma. The true giant cell tumors of the extremities are known to metastasize in lungs (Von Hoeven et al, 1994). There was however neither lymphatic or pulmonary dissimulation in this case. It is perhaps the inadvertently left residue, might have got reactivated within 2 months which required more thorough excision. In fact the rapidity of recurrence or the reactivation of residue, stresses its aggressive yet basically benign nature.

The brownish discoloration of the tumor on gross appearance observed herein is suggestive of haemosiderin deposition subsequent to the bleeding, it is again a behaviour of rapidly growing tumor. The absence of any osteoid, collagen and immature bone are unusual features of the true giant cell tumor but may be due the rapid pace of growth in this case. There was absence of giant cells intimately adjacent to the capillaries, but old hemorrhagic foci were seen, as are well documented in true giant cell tumors (Gusek et al, 1958 and David and Korth, 1959). The lack of contact of the giant cells to the vascular endothelium, as seen in some true giant cell tumours, insured freedom from vascular dissimulation in this case. The closely matched (histologically) entity is the giant cell reparative granuloma which according to Waldon & Shafer (1966) and Auclair et al (1988) together is the variant of the same disease process modified by age of the patient and the site of occurrence Leban et al (1971) also considers that there is a considerable overlap

between the two lesions. The giant cell tumour is composed of larger giant cells with more number of nuclei. The type of giant cells observed comparatively amongst both the specimen of this particular case did not reveal any difference thus suggesting the recurrence of the previous tumefaction rather than occurrence of giant cell reparative granuloma. The more frequent production of osteoid or new bone and the more frequent occurrence of inflammatory components are seen in giant cell reparative granuloma. The present case neither revealed any significant osteoid formation nor any abundant occurrence of mononuclear cells, in both the tissue specimen. However neither of the specimen revealed any foci of necrosis which is more often seen in giant cell tumour. The more abundant occurrence of haemosiderin pigmentation in the second specimen as compared to the first one may signify a more rapid recurrence (resulting in more bleeding) or a coexistent reparative granulomatous process. This further emphasises the statement of Auclair et al (1988) who regarded these two as a variant of same disease process. The most constant feature of a giant cell tumour is a relatively even distribution of giant cells throughout the entire lesion, in contrast to the reparative granuloma wherein the distribution is uneven and aggregated around the areas of haemorrhage. Both the specimen revealed relatively similar even distribution within any predominant concentration around the haemosiderin pigment. The tumour was of grade 2 as per the grading proposed by Jaffe et al (1940) based on the stromal component. As the clinical behaviour of these variants differ, there is a need to differentiate the two in order to plan out the management for achieving best prognosis.

The clinical behavior of this rapidly reactivating residue, inspite of a thorough and adequate local clearance of this lesion is unlike that of giant cell reparative granuloma. The absence of pain and systemic metastasis and lack of cachexia observed herein, is also against it being a malignant disease. The anemia resulted from nutritional deficiency, subsequent to his inability for proper mas-

tication and intake in present of intra-oral tumefaction with super added infection. It appears to be an unusual presentation of the true giant cell tumor.

This is therefore observed to be an unconventional and rare true giant cell tumor occurring

in a male child which is aggressive, locally destructive and non-metastasising lesion, without classical radiological picture. Its rapidly recurrent nature requiring radical surgery for cure may be related to enhanced growth potential of children at such age.

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