Peripheral Giant Cell Reparative Granuloma of the Sino-orbital Region—A Giant Lesion: A Rare Case Report

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ABSTRACT
Peripheral giant cell reparative granulomas (PGCRGs) are benign tumors which spread locally and progress aggressively. The origin of the multinucleated giant cells is not established till date, and hence various theories suggesting the origin from osteoclasts based on the immunohistochemical features, or from mononuclear phagocyte system. The treatment of choice is surgical excision with adequate margins and the recurrence rates are usually below 10% as per the reported literature. We present a 6-year-old child who presented with a rapidly progressive sino-orbital mass leading to proptosis and lateral displacement of the right globe. He underwent combined approach (open and endoscopic assisted) for complete excision of the lesion. The histopathological features were consistent with PGCRG.

This case is being highlighted with the aim of developing insights about the clinical presentation and management of PGCRG.

Keywords: Embolization, Endoscope, Giant cell granuloma, Sino-orbital.


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INTRODUCTION
Giant cell reparative granulomas are rare lesions of head and neck areas. These are benign fibro osseous lesions which present typically as rapidly increasing tumors with associated cortical bone defects. Peripheral giant cell reparative granulomas (PGCRGs) are extremely rare in the paranasal sinuses and are usually seen involving the mandible and maxilla in the head and neck region.

Initially, PGCRG were thought of giant cell tumor variants and in 1953, Gaffe introduced the term PGCRG to the scientific community. It is usually thought to arise as a result of reparative hyperplastic response to an insult to the area concerned. Although the lesion is extremely rare, it has been reported to be involving the orbit, paranasal sinuses, temporal bone, calvaria, mandible and maxilla in the head and neck region.

The clinical presentation varies as per the site involved and the anatomical structures present in the vicinity. The treatment of choice is surgical resection of the lesion but a limited role of radiotherapy has also been suggested for tumors nonapproachable or resectable surgically. However, sarcomatous changes have been shown to occur in those irradiated.

This case is being reported in view of its extent and unusual presentation. This case also suggests that the treating clinician need to consider PGCRG as a probable differential diagnosis for any rapidly growing sinonasal lesions in children.

CASE REPORT
A 6-year-old boy presented to our outpatient department (OPD) with complaints of right-sided watery discharge from the eye followed by swelling in the sino-orbital region for 3 months. He also complained of occasional few spontaneous right nasal bleed with associated pain but no associated visual disturbances.

Clinical examination revealed proptosis of the right eye, pushing the eyeball laterally and outwards, with widening of the nasal bridge and bulge in the medial canthus (Fig. 1). A slough covered polypoidal mass was noted filling the entire right nasal cavity on anterior rhinoscopy. Ophthalmologist examined reported nonaxial proptosis with 6/18 vision in right eye.

Magnetic resonance imaging (MRI) with contrast and computed tomography (CT) scan was done which suggested a relatively well-defined expansile multiloculated lesion in the right sinonaso-orbital region (Figs 2 and 3). On T2-weighted images, it was heterogeneous and had fluid-fluid levels, hypo- and hyperintense areas; post Gadolinium scans showed enhancement of the
septations. Medially, the lesion was causing breach of the right medial orbital wall with the mass filling and expanding the right nasal cavity causing obliteration of the right nostril. Laterally, the lesion was causing mass effect on the right orbit displacing the rectus muscles and optic nerve laterally with resultant proptosis. Posteriorly, the lesion invaded part of sphenoid sinus on the right side. Superiorly, the lesion caused thinning of the right superior orbital wall in the medial aspect. However, there was no intracranial extension. Inferiorly, the lesion was eroding the medial aspect of inferior orbital wall and was filling the right maxillary sinus. The lesion also displaced the right optic nerve laterally.

As the lesion was suspected to have a high vascular supply, it was decided to embolize the feeding vessels preoperatively. Right external carotid angiogram revealed tumor blush arising from the internal maxillary artery. Progreat micro catheter and gelfoam was used to selectively embolize this vessel under fluoroscopic guidance and a single 3 mm coil was deployed in this artery. Another meningeal branch of the right external carotid artery was seen to supply the lesion and this was embolized selectively using gelfoam slurry. Post-procedural angiogram showed 90% reduction in the tumor blush.

Embolization was immediately followed by lateral rhinotomy incision with superior extension beneath the eyebrow (Lynch-Howarth approach) with endoscopic assistance for complete excision of the mass was made. The tumor was fleshy, lobulated vascular mass with blood filled cystic areas involving right nasal cavity, right maxillary sinus pushing the middle turbinate medially, breaching lamina papyracea and involving superior, medial and inferior areas of orbit causing displacement of orbital contents laterally. Tumor excised in toto (Fig. 4). Bony remodelling of frontal sinus was noted. Laterally, displaced orbit was secured back into the normal place by help of hitch stitches placed between the medial canthal ligament and periosteum and the wound was closed in layers (Fig. 5).

Histopathological examination revealed a submucosal lesion covered by respiratory epithelium, composed of loose spindle cell proliferation, with numerous osteoclast giant cells, in a hemorrhagic stroma, containing scattered mast cells and foci of reactive new bone (Figs 6 to 9).

**DISCUSSION**

Peripheral giant cell reparative granuloma is a nonneoplastic reactive tumor that is less common in head and neck region. It occurs mostly in the mandible and maxilla in the head and neck regions. The next most common site described is the hand and feet. Peripheral giant cell
reparative granuloma can be encountered in all age groups, however, usually more likely to be seen in young adults and children.\(^8\)

Despite being a benign lesion, the aggressive growth rate makes these behave as clinically malignant. A very few of these have been found to involve the paranasal
sinuses and the orbit. These lesions have varied clinical presentation depending on the location afflicted and the surrounding anatomical structures.29

Lesions located in the sinonaso-orbital region can present as pain, diplopia, proptosis, hypertelorism, facial deformity, nasal obstruction, epistaxis and peri-orbital swelling.6

Basically, these lesions belong to the parent group of giant cell reparative granulomas (GCRGs). Giant cell reparative granulomas are of two types namely: central-endosteal and peripheral-soft-tissue type.10 The central type is usually seen in the age group of 10 to 20 years but can occur in all age groups. This subtype is generally found to involve the mandible. The peripheral type is seen usually involving gingival and alveolar mucosa. This subtype is encountered more commonly in females below 30 years of age. Histologically, there exists no difference between central and peripheral giant cell reparative granulomas.

As per the clinical behavior, these are also categorized into two groups as aggressive and nonaggressive types. The nonaggressive type has as low course and is usually painless, whereas the aggressive type is painful and has an rapid growth pattern with an increase recurrence chances.11

The differentials of PGCGRG of sino-orbital regions are aneurysmal bone cyst (ABC), giant cell tumor (GCT), lymphangioma and brown tumor. The expanded lytic, cystic appearance with fluid-fluid levels can be seen with ABC, GCT and lymphangioma with vascular components could have hemorrhagic areas within. Usually, ABC arises from vertebrae and long bones, involves the host bone and is also uncommon in facial bones. Histologically, it can be differentiated from giant cell tumor by the absence of sheets of plump mononuclear spindle to ovoid stromal cell proliferation.12 To differentiate it from brown tumor, it is important to note that the serum levels of Ca, P and PTH, are within normal limits in cases of GCRG.12

Clinical entities like squamous cell carcinomas, sinonasal sarcomas, such as lymphomas, sinus nerve sheath tumors, inverted papilloma, extramedullary plasmacytomas, minor salivary gland tumors, esthesioneuroblastomas, fibrous dysplasias, melanomas, and hemangiopericytoma are a few other differentials that are to be considered when evaluating a rapidly progressive sino-orbital mass.

The treatment of choice for well-defined paranasal sinus lesions is en bloc resection and excision. When treating such extensive lesions in pediatric age group, embolisation prior to the procedure allows the blood loss to be minimized as even a small amount of blood loss can be relatively significant.

Additionally, the use of endoscope gives an added advantage to resect these tumors from difficult areas, such as anterior skull base as was evident in our case. As mentioned before the unresectable or surgically unapproachable areas can be irradiated, but sarcomatous changes have been well documented in these cases. However, no malignant transformation has been reported for those undergoing irradiation. The reported recurrence rate has been usually between 11 and 35%. Various studies have also suggested use of intralesional steroids, oral steroids, alpha interferon, phenol and ethanol treatment with bone grafting and several others, however, none of the mentioned above have turned to be conclusive.13,14

CONCLUSION

Peripheral giant cell reparative granulomas can rarely present as rapidly growing sinonasal tumors; its diagnosis is mainly based on radiological and histopathological features. The treatment of choice is mainly surgical excision with adequate margin clearance. In case of extensive lesion combined endoscopy with external approach by otorhinolaryngologist with the assistance from ophthalmologist is to be considered for a successful outcome.

REFERENCES


