RESEARCH PAPER | Medical Science | Volume : 2 | Issue : 3 | December 2012 | ISSN - 2249-555X

Total Maxillectomy with Orbital Clearance for Squamous Cell Carcinoma of Maxilla - Our xperience with two Cases and Literature Review

KEYWORDS: Total maxillectomy, Orbital exenteration, Orbital clearance, Reconstruction.

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ABSTRACT *Malignant tumours of ethmoid and maxillary sinus frequently involve the orbit. Orbital involvement is an important prognostic predictor of recurrence-free, disease-specific, and overall survival. We report two cases of maxillary sinus squamous cell carcinoma infiltrating the orbital floor who underwent total maxillectomy with orbital clearance operated at our institute. Various studies showed that orbital preservation as opposed to orbital exenteration or clearance does not result in significant differences in local recurrence or actuarial survival. Small defects following orbital exenteration may be reconstructed with a temporalis muscle flap. Microvascular free-tissue transfer is the best option for repair of defects following orbital exenteration and total maxillectomy, although an obturator still has a role in selected patients in low resource setups.*

Background and aims:
Malignant tumours of sinonasal tract usually present as advanced disease because early diagnosis is difficult. Radical surgical excision remains the mainstay of the treatment. The classic surgical treatment for sinonasal tumours in close proximity to the orbit involved radical excision with orbital clearance or exenteration. After the introduction of orbital preservation surgery, the indications and need for removal of orbital structures have diminished.

Nonetheless, despite several reports claiming the effectiveness of various treatment strategies, the issue remains controversial. The two main points to be considered in planning the surgical excision are the oncological safety of orbital preservation and the functional outcome in preserved eyes. Management of such case is challenging as it is a multimodality treatment that involves oncosurgeons, prosthodontist, radiation oncologist and speech therapist.

We report two patients with advanced squamous cell carcinoma of maxilla with orbital involvement managed at our institute. Clinical features, management and outcomes of treatment are presented here with a review of literature.

Materials and methods:
Patients with biopsy proven SCC of maxilla who presented to the surgery outpatient department of Shrimati Kashibai Navale Medical College and Hospital, Narhe, Pune were included in our study. Patient records were reviewed with respect to the demographic data, clinical presentation, pathological features and treatment outcomes.

A Medline literature search was performed using the MeSH terms "sinonasal tumours" and "maxillectomy" and "orbital clearance" or "orbital exenteration."

Case presentations
Case 1
40 year lady presented with progressive swelling of the left malar area since 8 month. There were no symptoms related to vision, nasal bleeding or difficulty in mouth opening besides pain. On clinical examination, the growth was arising from the left maxilla with involvement of orbital floor. On anterior rhinoscopy the growth was abutting the nasal septum, no diplopia and trismus. CT scan showed 6cm x 5 cm x 4cm lesion arising from left maxilla with erosion of all the walls of maxilla and infiltration of orbital floor and abutting the extraocular muscles and extending into the soft tissues of infratemporal fossa. Incision biopsy was done that reported as squamous cell carcinoma. She underwent total maxillectomy with orbital clearance. Intraoperative frozen section confirmation of periorbita was done to detect infiltration. Optic nerve margins were also confirmed with the same. The entire defect was covered with split skin graft and the cavity was packed with roller guaze pack. Post operative period was uneventful. Oral feeding was started on 5th post operative day and the graft take was 80%. Histopathology showed squamous cell carcinoma, all the bony, soft tissue and mucosal margins were negative. She completed adjuvant radiotherapy after which she was referred to prosthodontist for reconstruction of the maxilla and eyeball with a prosthesis. Following completion of the treatment she is on regular follow up since 6 month and is recurrence free.

Figure 1: CT scan showing erosion of orbital floor (Case 1)

Figure 2. Surgical defect after total maxillectomy and orbital clearance (Case 1)

Volume : 2 | Issue : 3 | December 2012 | ISSN - 2249-555X

Case 2
54 year gentleman presented with progressive swelling of the left malar area since 4 month. There were no other symptoms besides the swelling. On clinical examination, the growth was arising from the left maxilla with involvement of orbital floor. Following thorough investigation and work up he underwent total maxillectomy with orbital clearance. Intraoperative frozen section confirmation of periorbita was done to detect infiltration. Optic nerve margins were also confirmed with the same. Post operative period was uneventfull. Oral feeding was started on 5th post operative day and the graft take was 70%. Histopathology showed squamous cell carcinoma, all the bony, soft tissue and mucosal margins were negative. He completed adjuvant radiotherapy and presently being referred to prosthodontist for the reconstruction of the maxillary and orbital defect with a prosthesis.

Figure 3. Appearence of patient following closure of the defect (Case 2)

Discussion and review of literature:
All types of malignant tumours are encountered in the nose and paranasal sinuses. Of these tumours malignant melanoma has the worst outcome, while esthesioneuroblastoma and chondrosarcoma has the best prognosis [1-7].

The two most common histologic types of sinonasal tumours are adenocarcinoma and squamous cell carcinoma. Adenocarcinoma has a better disease-specific survival and recurrence-free survival than squamous cell carcinoma [2].

The incidence of orbital invasion by malignancies of the sinonasal tract varies with the site of origin, histology, and the aggressiveness of the tumor.

Tumors invade the orbit via preformed pathways, via neurovascular structures, or by direct extension through the bone. Tumor extension into the orbit occurs very commonly in ethmoid tumors, because of the thin lamina papyracea separating the two structures.

Invasion of the orbital wall is present in 66% to 82% of the patients with ethmoidal malignancy [1-2], with involvement of the orbital periosteum in 30% to 50% of patients [2, 3,9]. Orbital invasion (bone erosion/invasion) occurs in 60% to 80% of maxillary sinus malignancies [10].

The periorbitum is a barrier against invasion, but once the tumor has infiltrated this robust periosteum it gains access to a space that lacks barriers to check local tumor spread. An important distinction should be made between erosion of the bony orbital wall, infiltration of the perisoteum, and deeper invasion of the orbital soft tissues. Intraoperative frozen section confirmation is required to determine tumor infiltration of the orbital periosteum and periorbital tissue.

The term ''orbital exenteration'' is normally applied to complete removal of the contents of the orbit, including the eyelids whereas ''Orbital clearance'' is a procedure in which the globe, muscles, fat, and periorbita are removed, while the lids, and the palpebral conjunctiva are preserved.

Iannetti et al [1] has described 3 stages of orbital invasion: grade I, erosion or destruction of the medial orbital wall ; grade II, extraconal invasion of the periorbital fat ; grade III, invasion of the medial rectus muscle, optic nerve, ocular bulb, or the skin overlying the eyelid. Grade III orbital invasion warrants orbital clearance or exenteration.

Effect of orbital involvement on patient survival
Orbital involvement is associated with a significant reduction in survival both in ethmoid and maxillary sinus tumors. Most authors also found orbital invasion to have a deleterious impact on the outcome of maxillary sinus tumors [12, 13, 14]. In a review of 57 patients who underwent maxillectomy, multivariate analysis confirmed that skull base and orbital involvement were the only factors significantly associated with disease-specific survival.

Involvement of the orbit was associated with a 5-year survival of only 17%, as opposed to 49% when there was no invasion [14]. On the other hand, no survival benefit was achieved by orbital clearance, only 11% of patients with orbital involvement remained alive after 5 years despite complete extirpation of orbital contents [14]. Orbital invasion also was an independent prognostic factor in a series of 95 tumors of the maxillary sinus, and T4 tumors with orbital invasion had a worse prognosis than other T4 tumors [13].

No definite consensus has been reached on the degree of orbital invasion that is oncologically safe when sparing orbital contents. Different indications for orbital clearance have been proposed based on involvement of periorbita, orbital fat, extraocular muscles, or orbital apex. Thus, a selection bias exists in all of these studies because the tumors in more advanced stages (ie, orbital apex invasion) with expected worse outcomes were treated with orbital clearance, whereas those with more favorable orbital extension were treated with more conservative approaches.

Effect of orbital clearance on local recurrence
In a retrospective review of 111 patients with maxillary sinus tumors who had invasion of the orbital floor, it was reported that when clearance was performed, the 5-year survival rate was 27.3% and the rate of recurrence within the orbit was 12.5%. In patients with preservation of the orbital contents the corresponding rates were 34.8% and 8.6%, respectively, with no significant differences between these groups [16].

Several authors have advocated an eye- sparing approach in the treatment of sinonasal tumors with periorbital involvement based on similar oncologic results with the less radical procedures [17-20].

In a review of 53 patients with squamous cell carcinoma of the maxillary antrum, clearance was performed in 25 patients for disease invading the orbit with gross involvement of periorbita, whereas the other 28 patients with disease that either did not fully invade through bone into the orbit or invaded the orbit without gross periorbital involvement were treated by orbital preservation. Again, the results demonstrate no significant difference in local recurrence or actuarial survival between the orbital preservation and orbital clearance groups [20].

A meta-analysis of disease-free survival and local recurrence in 170 patients with orbital invasion by squamous cell carcinoma revealed that patients in whom the orbital contents were preserved had 5-year survival and local recurrence rates of 41%and 20%, versus 37% and 36%when orbital clearance was undertaken [10]. These results do not demonstrate any significant difference in local control or actuarial survival. However, it should be noted that these studies were retrospective and that patients who underwent orbital clearance generally had more advanced orbital involvement.

Eye function following conservative procedure
Imola and Schramm [15] reported functional results in pa-

Volume : 2 | Issue : 3 | December 2012 | ISSN - 2249-555X

tients in whom the orbit had been preserved. Overall eye function was reported as functional without impairment in 54%, functional with impairment in 37%, and nonfunctional in 9%.

The most common abnormality was globe malposition in 63% of patients and was associated with lack of adequate rigid reconstruction of the complete orbital floor or multisegmental orbital defects, with persistent diplopia in 9% of patients.

Radiation therapy increased the risk of ocular complications, in particular optic atrophy, cataracts, excessive dryness, and ectropion. Stern et al [20] reported that only 17% of patients, who had the orbital floor resected without an attempt to reconstruct the orbital floor, retained significant function in that eye. On the other hand, patients who had an intact bony orbital floor and when radiation fields did not include the eye had minimal problems.

Eye function is thus strongly influenced by the position of the resected orbital segment, with abnormal eye function in patients undergoing total maxillectomy without reconstruction of the orbital periosteum and bony floor. However, eye function in patients subjected to total ethmoidectomy or resection of the lateral wall is almost always normal [21]. It follows that large defects resulting from complete orbital floor resection or resections involving two or more orbital walls and large portions of orbital periosteum should undergo reconstruction.

Reconstruction of orbital floor defects

Reconstruction of orbital floor after total maxillectomy should provide support to the orbital contents, reconstruct the palatal surface, achieve facial symmetry and a good aesthetic result. Repairing lost orbital support decreases the risk of globe malposition, diplopia, and disturbance of extraocular muscle function, as well as lid malposition and ectropion resulting in exposure keratitis [15].

Minimal bony resection such as isolated orbital wall (lateral or medial) or small orbital floor defects do not require any kind of reconstruction. Resection of medial orbital periosteum can be repaired with split skin or fascia lata with minimal morbidity.

Larger defects in the orbital floor can be repaired using a thick fascial sling tightly secured to the margins of the bony defect. With subtotal or total floor defects (>75% surface area) and multisegmental defects, orbital floor and one or more walls, some form of rigid reconstruction is advisable.

Primary reconstruction of total maxillectomy defects with preservation of orbital contents remains a complex problem without a perfect solution. These methods include skin graft or a temporalis muscle sling, the sheath of the upper portion of the rectus abdominis muscle or other related procedures, but they have also resulted in complications, such as enophthalmos, diplopia, and facial deformity.

Nonvascularized bone grafts or a titanium or synthetic polyethylene mesh, in conjunction with a soft tissue free or pedicled muscle flap, can be used to reconstruct the orbital floor. Split ribs, iliac crest grafts, or even vascularized calvarial bone flaps, [22] radial forearm osteocutaneous flaps [23, 24] or coronoid-temporalis pedicled rotation flaps [25] have been used for reconstruction of the orbital floor.

Reconstruction of orbital exenteration or clearance defects

Maxillectomy defects become more complex when critical structures such as the orbit, globe, and cranial base are resected. Rectus abdominis microvascular free-tissue transfer for repair of orbital exenteration or clearance defects with or without total maxillectomy is a safe and reliable alternative to the use of the temporalis muscle pedicled flap.

It provides a larger volume of well-vascularized tissue and greater placement flexibility, and the long vascular pedicle facilitates the use of multiple donor vessels within the head and neck, which is an advantage in previously irradiated patients. Microvascular free flap reconstruction provides an expeditious and immediate means of reconstruction of surgical defects, where soft tissue replacement and lining in multiple areas are required.

Orbital prostheses can be applied to the socket once healing has occurred. Ideally, these can be secured with osteointegration, which can be done primarily at the time of the resection or as a secondary procedure after completion of adjuvant radiotherapy.

Conclusion

Orbital preservation can be done when the periorbita is not infiltrated by the tumor without affecting the cure or local control. Large multi-institutional studies are needed to address the issue of orbital preservation in malignant sinonasal tumors because the existing data is based on retrospective analysis and non randomized studies.

Orbital reconstruction is essential for large defects resulting from total orbital floor resection or resections involving two or more orbital walls, to prevent displacement and dysfunction of the eye.

Volume : 2 | Issue : 3 | December 2012 | ISSN - 2249-555X

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