Original Article - Prospective Study

Reconstruction in oral malignancy: Factors affecting morbidity of various procedures

Access this article online
Website: www.amsjournal.com
DOI: 10.4103/2231-0746.175748
Quick Response Code:

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ABSTRACT

Aims and Objective: (1) To study the age and sex distribution of patient with oral malignancies. (2) To analyze various types of surgery performed. (3) Evaluation of reconstruction and factors affecting complications and its relation to the type of reconstruction. **Materials and Methods:** Cases of oral malignancies, undergoing surgery for the same in Sri Aurobindo Medical College and PG Institute, Indore from the period from October 1, 2012, to March 31, 2015. **Results:** Out of analysis of 111 cases of oral malignancy, 31 (27.9%) cases were in the fifth decade of life with male to female ratio 1.9:1. The commonest site of cancer was buccal mucosa. Forty-seven cases (43.2%) were in stage IVa. Diabetes was the most common co-morbidity reported, accounting for 53.9% of cases with reported morbidity. Tobacco chewing was the common entity in personal habits. All the cases underwent neck dissection along with resection of the primary. Hemimandibulectomy was the most preferred form of primary resection accounting for 53.15% (59 cases), followed by wide resection of primary 27% (30 cases). Pectoralis major myocutaneous (PMMC) flap only was the most common reconstruction across the study population. PMMC alone accounted for 38.7% (43 cases). The infection rate was 16.21%. PMMC alone accounted for 5 out of 18 (27.8%) of total infection rate, and 4.5% of the total study population. PMMC + deltopectoral accounted for 5 out of 18 (27.8%) of total infection rate, and 4.5% of the total study population. **Conclusion:** PMMC is a major workhorse for reconstruction with better functional outcome and acceptance among operated patients.

Keywords: Deltopectoral flap, oral malignancy, pectoralis major myocutaneous flap

INTRODUCTION

Head and neck cancers account for 23–25% of all cancers occurring in different sites and oral cancers account for 50% of these or 12.5% of the whole body. The prevalence in subsites of the oral cavity as lower lip 0.32, upper lip 0.01, anterior tongue 2.81, lower alveolus 1.84, upper alveolus 0.25, floor of mouth 0.44, buccal mucosa 4.82, hard palate 0.32, retromolar space 0.51, and base tongue 1.36[1] (these figures are a percentage of all cancers in the body). Globally, over 300,000 people are diagnosed with oral cancer each year, and it is the eighth most common malignancy worldwide.[2] There is considerable geographic variation, oral cancer being the third most common cancer in South East Asia. The age/standardized incidence rate for oral cancer in SE Asia is as high as 25/100,000 per annum.

Oral cancer is more common in males, patients usually presenting in the sixth–seventh decade of life, although the incidence of oral cancer in young people seem to be increasing.[3,4] Smoking and alcohol consumption are the major etiological factors in the development of oral cancers.[5] Apart from squamous cell

Cite this article as: Chakrabarti S, Chakrabarti PR, Desai SM, Agrawal D, Mehta DY, Pancholi M. Reconstruction in oral malignancy: Factors affecting morbidity of various procedures. Ann Maxillofac Surg 2015;5:191-7.

carcinoma, the most common oral cancer is mucoepidermoid cancer arising out of the minor salivary glands.

Reconstruction after oral cancer resections should aim at maintaining the functional integrity of the different structures of the oral cavity which includes pliable buccal mucosa for adequate mouth opening, jaw for stable mastication, dental rehabilitation, tongue of adequate size, shape, and mobility for speech and swallowing and floor of mouth should be able to hold food or saliva without leak. The tissue requirement at the time of reconstruction can be broadly looked at in terms of the quantum of soft tissue required and the skeletal framework that needs to be reconstructed. The reconstructive ladder starting from skin grafts and ending with free flaps may not always be able to be followed due to anatomical and functional requirements of the defects. Skin grafts may be useful in only select cases and defects such as the small and shallow defects in the floor of the mouth or cheek. Local flaps such as nasolabial flaps provide thin, reliable skin tissue suitable for repairing, only again, in small defects. Most often tissue will need to be brought into the region to repair larger defects with the use of pedicle or free flaps. The pedicle flaps commonly used for oral reconstruction include a pectoralis major myocutaneous (PMMC) flap, forehand flap and platysma myocutaneous flaps[6] and skin flaps like a deltopectoral (DP) flap, latissimus dorsi. Micro vascular free flaps have allowed great flexibility to the reconstructive surgeon to import composite tissues matching the requirements at the site better than other methods and have become the method of choice in a great number of defects. This becomes more significant in reconstructing bony defects.

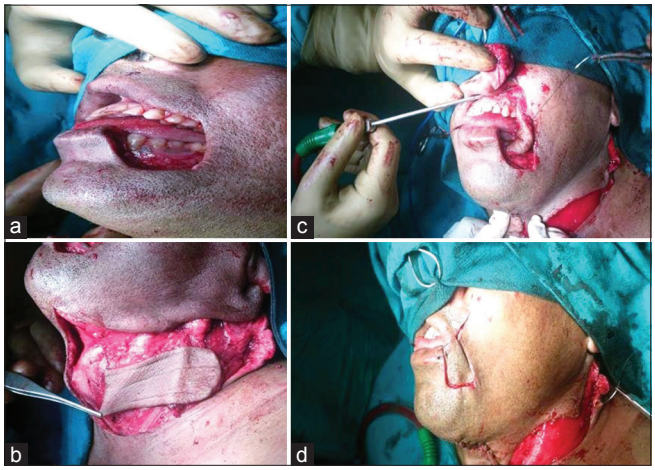

Figure 1: (a) Intraoperative photograph of postexcision surgical defect of carcinoma of left side of lower lip, (b) intraoperative harvesting of platysma flap, (c) intraoral placement of platysma flap, (d) intraoperative closure of surgical defect with nasolabial flap

Figure 3: (a) Bipedal pectoralis major myocutaneous flap donor site incision, (b) intraoperative photograph of postexcision surgical defect, (c) intraoperative photograph of placement of bipedal pectoralis major myocutaneous flap, (d) postoperative photograph

Aims and objectives

- To study age and sex distribution of patient with oral malignancies
- To evaluate the type of surgery performed
- Evaluation of reconstruction options of oral malignancy surgery
- To study factors affecting the complications and its relation to the type of reconstruction.

Figure 2: (a) Immediate postoperative photograph of reconstruction with pectoralis major myocutaneous and deltopectoral flap with split skin grafting of deltopectoral donor site, (b) postoperative day 14 photograph

Figure 4: (a) Postoperative photograph of forehead flap reconstruction, (b) follow-up visit of same patient at 4th month postsurgery

Figure 5: (a) Carcinoma of left buccal mucosa with orocutaneous fistula postchemoradiation, (b) intraoperative photograph of postexcision surgical defect (c) intraoperative skin marking of latismus dorsi flap donor site, (d) intraoperative scalp flap donor site marking, (e) postoperative photograph and (f) postoperative follow-up vist at 1 month

Figure 6: (a) Reconstruction with nasolabial flap, (b) reconsruction with Abbe estlander flap, (c) postexcision primary closure for verrucous carcinoma of lower lip, (d) intraoperative photograph of postmaxillectomy local recurrence excision and forehead flap

MATERIALS AND METHODS

The present study comprises cases of oral malignancies undergoing surgery in the Department of Surgical Oncology at Sri Aurobindo Medical College and PG Institute, Indore, Madhya Pradesh in a study period from October 1, 2012, to March 2015 [Figures 1-8].

A pretested proforma was used to collect the relevant information by interviewing, clinical examination of patients, and noting relevant investigations required for treatment. Postoperatively patients were evaluated clinically on a monthly basis in the 1st year, 3 monthly in the 2nd year and 6 monthly in the 3rd year. Additional investigations were ordered as deemed necessary only after clinical examination. Patients were enrolled as and when required when they presented with the following inclusion and exclusion criteria.

Inclusion criteria

In the study, all the cases diagnosed with oral malignancy and subjected to relevant investigations and underwent surgery were included.

Exclusion criteria

- Cases which had extensive nature of the disease
- Inoperable and managed with definitive or palliative radiotherapy (RT) or concurrent chemotherapy (CT) and RT.

RESULTS

Out of 111 cases, 31 cases (27.9%) were in the fifth decade of life with male predominance and male to female ratio 1.9:1. The most common site for oral malignancy in this study was buccal mucosa accounting for 28.2% of the cases [Tables 1-3].

Forty-seven cases were staged as stage IVa; overall stage IV accounted for 43.2% of the total cases [Table 4]. 87.4% cases were newly diagnosed carcinoma oral cavity.

Surgery was the most common upfront treatment modality with seven cases accounting for 6.3% cases. Out of the operated

Table 1: Age wise distribution of oral malignancy

Age (years)	Number of patients	Percentage
<20	0	0
20-29	7	6.3
30-39	18	16.2
40-49	31	27.9
50-59	24	21.6
60-69	22	19.8
70-79	9	8.1
>80	0	0

Table 2: Sex wise distribution of oral malignancy

Sex	Number of patients	Percentage
Male	73	65.7
Female	38	34.3

Table 3: Site wise distribution of oral malignancy

Site	Number of patients	Percentage
Upper lip	2	1.8
Lower lip	4	3.6
Anterior part of tongue	20	18
Lower alveolus	25	22.5
Upper alveolus	6	5.4
Floor of mouth	4	3.6
Buccal mucosa	32	28.2
Retromolar trigone	14	12.61
Base of tongue	4	3.6

Table 4: Stage wise distribution (AJCC 2010) of oral malignancy

Stage AJCC (2010)	Number of patients	Percentage
I	21	18.9
II	16	14.4
III	24	21.6
Iva	47	42.3
IVb	1	0.9
IVc	0	0
Incomplete staging	2	1.8

Figure 7: (a) Neck dissection skin flap dehiscence, (b) postprimary closure dehiscence, (c) pectoralis major myocutaneous flap and deltopectoral flap failure, (d) pectoralis major myocutaneous flap donor site abscess collection

cases, four cases were operated for the same with unknown margin and lymph node status. One case underwent total glossectomy followed by carcinoma of retromolar trigone after 6 months; one case had undergone wide excision of carcinoma buccal mucosa right side 18 years back followed by carcinoma of left lower alveolus of the left side. One case had undergone wide local excision with modified radical neck dissection (MRND) followed by concurrent chemoradiation for carcinoma buccal mucosa, now with a recurrence of disease and orocutaneous fistula. Out of the three cases receiving radiation, two were defaulters [Table 5]. Twenty-four cases (21.6%) had both hypertension and diabetes, whereas diabetes was the most common co-morbidity reported, accounting for 48% of the total cases and 53.9% of cases with reported morbidity [Table 6]. All 111 cases had some form of addiction. One hundred and ten cases (99.09%) gave a history of tobacco chewing in the form of gutkha, khaini, mawa.

All the cases underwent neck dissection, either in the form of MRND or supraomohyoid neck dissection. Hemimandibulectomy was the most preferred form of primary resection accounting for 53.15% (59 out of 111), followed by wide resection of primary 27% with 30 cases [Table 7]. PMMC flap only was the most common reconstruction across the study population. PMMC alone accounted for 38.7% (43 out of 111) followed by primary closure 20.7% (23 out of 111). PMMC + DP reconstruction was performed in 14 cases (12.6%), PMMC + forehead flap in 8 out 111 (7.2%) and in one case PMMC + split skin graft (SSG) (0.9%). Hence, overall PMMC was a used as single or in combination for reconstruction in 66 out of the total 111 cases, i.e., 60% of the total study population [Table 8].

Major complication rate was 8.1% (9 out of 111). The minor complication rate was 12.61% (14 out 111). Karnofsky performance scale score 60 and eastern cooperative oncology group Grade 2 was associated with highest complication rate (major + minor) 69.6% (16 out of 23) [Table 9].

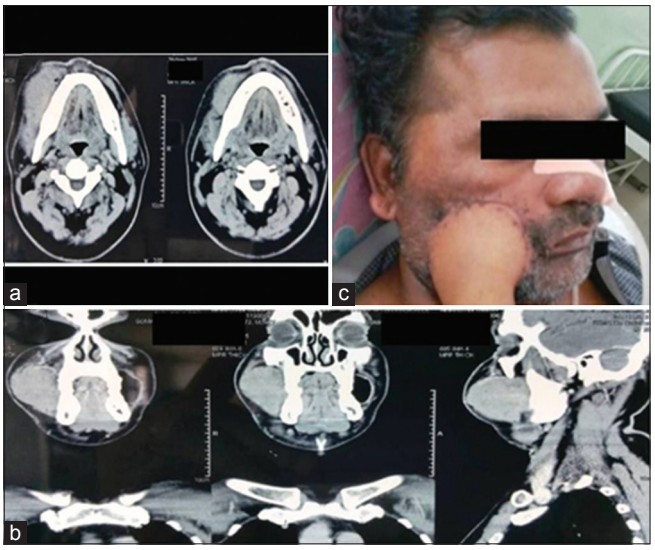

Figure 8: (a and b) Contrast-enhanced computed tomography neck showing large infiltrative hypodense enhancing mass lesion epicentered around the right gingivobucal sulcus with extension into the ipsilateral retromolar trigone without erosion of the alveolar process of mandible, (c) postoperative photograph of reconstruction with pectoralis major myocutaneous flap and deltopectoral flap on day 10

Table 5: Previous treatment history

Previous treatment	Number of cases	Percentage
CT	3	2.7
CT + RT	1	0.9
RT	3	2.7
Nil	97	87.4
Surgery	7	6.3

RT=Radiotherapy; CT=Chemotherapy

Table 6: Co-morbidity

Co-morbidity	Number of cases	Percentage
Diabetes mellitus	48	43.2
Hypertension	41	36.9
Diabetes mellitus + hypertension	24	21.6
None	22	19.8

Table 7: Type of primary surgery

Type of primary surgery	Cases	Percentage
Hemimandibulectomy	59	53.1
Marginal mandibulectomy	2	1.8
Segmental mandibulctomy	4	3.6
Bite-composite resection	1	0.9
Maxillectomy	6	5.4
Wide excision	30	27
Total glossectomy	9	8.1

Pectoralis major myocutaneous flap

PMMC was used as only or in combination with other flaps in 66 out of 111 of the study population (60%). Four cases of total flap necrosis were noted, accounting for 6.06% of the total reconstructive procedure involving PMMC and 3.6% of the total reconstructive procedures performed. Two cases of partial flap necrosis were noted, accounting for 3.03% of the total reconstructive procedure involving PMMC and 1.8% of the total reconstructive procedures performed. Three cases of

epidermolysis were noted, accounting for 4.54% of the total reconstructive procedure involving PMMC and 2.7% of the total reconstructive procedures performed. Dehiscence was noted in five cases, 7.5% of the total reconstructive procedure involving PMMC and 4.5% of the total reconstructive procedures performed. PMMC related suture dehiscence accounted for 35.7% (5 out of 14) of the overall dehiscence rate of 12.6% (14 out of 111) of the study group.

Deltopectoral flap

DP flap was a part of reconstruction in 15 out of the 111 cases (13.51%) of the study population. Two cases of flap necrosis were noted, 13.33% of the total DP flap reconstruction group and 1.8 of the total study population. Dehiscence was noted in two cases, 13.33% of the total DP flap reconstruction group and 1.8 of the total study population. DP flap related suture cut dehiscence accounted for 14.2% (2 out 14) and 1.8 of the total study population.

Radial free forearm flap

Radial, free forearm flap, was a part of reconstruction in 3 out of the 111 cases (2.7%) under study. One out of the three radial forearm free flap reconstruction underwent total flap necrosis. 33.3% of the total radial free flap group and 0.9% of the total study population.

Primary closure

Primary closure was performed in 20.7% (23 out of 111) of the study population. Dehiscence was observed in three cases, 13.04% of the primary closure group, 21.4% (3 out of 14) of the suture dehiscence group, and 2.7% (3 out of 111) of the study population.

Criteria for infection

- Fever over 38°C
- Leukocytosis
- Localized erythema, induration or purulent discharge.

The overall infection rate in this study was 16.21%. PMMC alone accounted for 5 out of 18 (27.8%) of total infection rate, and 4.5% of the total study population. PMMC + DP accounted for 5 out of 18 (27.8%) of total infection rate, and 4.5% of the total study population. PMMC + forehead flap accounted for 1 out of 18 (5.5%) of the total infection rate and 0.9% of the total study population. PMMC overall contributed to 55.55% (11 out of 18) of the total infection group [Table 10]. The most common organism was *Escherichia coli* 55.5% (10 out of 18), *Staphylococcus aureus* 22.2% (4 out of 18), diptheroids 22.2% (4 out of 18), and methicillin-resistant *S. aureus* 16.7% (3 out of 18).

DISCUSSION

Primary tumors of the oral cavity may be derived from the mucosa, salivary glands, neurovascular tissues, bone or dental tissues. Over 90% of the tumors of oral cavity are squamous cell carcinomas. The principles of reconstruction in the oral cavity following oral cancer excision remain to maximize the functional results. Functions of the oral cavity include the articulation component of the speech, tongue mobility to propel food and clear the oral cavity food debris, and mobility of tongue tip to prevent the pooling of saliva in the sump areas of the anterior and lateral floor of mouth.

In this study, the youngest patient was 22 years while the eldest was 78 years. The mean age of patients was 45 ± 4.3 years. Majority of patient belonged to the fifth decade in life. Franceschi *et al*.[3] and Shiboski *et al*.[4] reported the majority of oral cancers in the sixth decade of life. The male: female sex ratio being 1.9:1. The male to female ratio reported by large-scale epidemiological studies and national cancer registries varies from 2:1 to 15:1 depending on the site of disease.[7] The most common site of oral

Table 8: Reconstruction procedure

Type of reconstruction	Number of cases	Percentage
PMMC only	43	38.7
Primary closure	23	20.7
PMMC + DP	14	12.6
PMMC + forehead flap	8	7.2
Radial free forearm flap	3	2.7
SSG	5	4.5
DP only	1	0.9
Lateral tongue flap	5	4.5
Forehead flap	4	3.6
Nasolabial flap	2	1.8
PMMC + SSG	1	0.9
Total	111	

PMMC=Pectoralis major myocutaneous flap; DP=Deltopectoral flap; SSG=Split skin graft

Table 9: Complication following reconstruction

Type of reconstruction	Major complication	Minor complication	Infection
PMMC only	4	3	5
Primary closure	0	3	4
PMMC + DP	4	3	5
PMMC + forehead flap	0	1	1
Radial free forearm flap	1	0	0
SSG	0	1	1
DP only	0	0	1
Lateral tongue flap	0	1	1
Forehead flap	0	0	0
Nasolabial flap	0	1	0
PMMC+SSG	0	1	0

PMMC=Pectoralis major myocutaneous flap; DP=Deltopectoral flap; SSG=Split skin graft

Table 10: Infection related to reconstruction

Type of reconstruction	Infection
PMMC	5
Primary closure	4
PMMC + DP	5
PMMC + forehead flap	1
Radial free forearm flap	0
SSG	1
DP only	1
Lateral tongue flap	1
Forehead flap	0
Nasolabial flap	0
PMMC + SSG	0
Total	18

PMMC=Pectoralis major myocutaneous flap; DP=Deltopectoral flap; SSG=Split skin graft

malignancy in this study was buccal mucosa seen in 28.2% of the cases. This was followed by lower alveolus seen in 22.5% and anterior part of the tongue in 18% cases. Upper lip cases were 2, i.e., the lowest accounting for 1.8% of the total 111 cases studied. In western studies, the tongue and floor of mouth is the most common site for malignancy in the oral cavity. However, the retromolar trigone and buccal mucosa are the most common sites of primary in the world where chewing of tobacco and areca nuts is common.[8] This trend was observed in our study.

Stage IV accounted for 43.2% of the total cases and was the highest among oral malignancy. Sankaranarayanan *et al*.[9] reported the distribution of oral malignancy in the Indian population as stage I (25%), stage II (17%), stage III (18%), stage IV (33%), and unknown (8%). Diabetes was the most common co-morbidity reported, accounting for 48% of the total cases and 53.9% of cases with reported morbidity. Mangrulkar and Khair.[10] had noted an increase in postoperative morbidity in diabetic patients as compared to nondiabetic, but the increase was statistically insignificant.

All 111 cases had some form of addiction. Tobacco chewing in the form of guthka, khaini, mawa was the most common entity in personal history accounting for 99.09% which was analyzed in term of the period of exposure. Smoking alone accounted for 36% (40 cases) of the study population. Twenty-five out of 40 smokers were current smokers (one or more cigarettes per day or some day). Seventeen out of the 25 (68%) current smokers had major or minor surgery related complications. Smoking along with alcohol consumption and tobacco chewing was noticed in 16.21% (18 cases). Ten out of 18 were current smokers (55.5%) out of which 8 had minor/major surgery-related complication. Therefore, overall 35 out of the 111 (31.53%) were current smokers out of which 23 patients had major + minor complication (65.71%), which was quite high compared with the overall complication rate (major + minor) of 20.72%. Several large studies have demonstrated that current smoking increase mortality in head and neck cancers.[11-14] Forty Nine (44.1%) of the total study population had a history of alcohol consumption. Thirteen of the 49 (26.49%) patients had postoperative infection compared to 5 out of 62 (8.06%) patients without a history of alcohol consumption. Prospective and retrospective studies[15] demonstrate a 2-fold to 3-fold increase in postoperative morbidity in alcohol abusers, the most frequent complications being infections, bleeding, and cardiopulmonary insufficiency. Wound complications account for about half of the morbidity. As all the patients with a history of alcohol intake had history of tobacco consumption and/or smoking, it was difficult for us to comment on the role of alcohol on the complication rate.

87.4% cases were newly diagnosed carcinoma oral cavity. Surgery was the most common upfront treatment modality with seven cases accounting for 6.3% cases. Neoadjuvant CT accounted for 2.9%, neoadjuvant radiation 2.7%, neoadjuvant chemoradiation 0.9% case population under study. This was low when compared to data from Tata Memorial Centre, Mumbai for the year 2013–2014 where neoadjuvant CT followed by surgery accounted for 20% cases. Radiation followed by surgery was accounting for 20% of the case of Head and Neck Department. Number of cases who received upfront treatment was few in number so their relation vis-a-vis postoperative morbidity could not be commented upon.

PMMC only was the most common reconstruction across the study population accounting for 60% of the total study population. Four cases of total flap necrosis were noted, two cases of partial flap necrosis were noted, and three cases of epidermolysis were noted, of the total reconstructive procedure involving PMMC. PMMC overall contributed to 55.55% (11 out of 18) of the total infection group. Female sex accounted for 5 out of the 9 (55.5%) major complication cases, presumably because of the interposition of breast tissue between the muscle and skin pedicle. Kroll *et al*.[16] reported in his study of 168 cases, the overall complication rate of 63%, total flap loss of 2.4%, more in the subgroup of patients with the larger tumor. They observed that RT had no effect on PMMC flap vitality, thereby inferring PMMC to be a sturdy flap. They also noted a predilection of flap necrosis in the female sex. McLean *et al*.[17] in a 16-year study of 136 reconstruction using PMMC note a complication rate of 13%. Zou *et al*.[18] studied 24 cases of PMMC reconstruction with a complication rate of 62.5%, 2 cases (8.3%) of flap necrosis, 5 (20.8%) cases of partial flap necrosis and 3 cases of $<40\%$ flap necrosis. Baek *et al*.[19] in their study of 133 cases of PMMC reconstruction reported total necrosis in 1.5% and partial necrosis in 7%, dehiscence in 13% cases. Mehrhof *et al*.[20] in their study of 73 cases of PMMC reconstruction reported total necrosis in 12.3% and partial necrosis in 12.3%.

Functional outcome in PMMC reconstruction in relation to oral continence were normal 4 (6.1%) cases, occasional drooling in 46 cases (69.7%), continuous drooling 16 (24.2%). PMMC + DP accounted for 10 out 16 patients with continuous drooling, 4 out of the 46 patients with occasional drooling. All PMMC + forehead flap (8) had occasional drooling. Deglutition with full diet in 13 (19.6%) patients, soft diet 38 (57.8%) patients, tube feeding 15 (22.8%). All the patients on the postoperative full diet had PMMC only reconstruction. All the patients on tube feeding had either PMMC + DP or PMMC + forehead flap reconstruction had a flap related complication. Speech was normal in 4 (3.6%), easily intelligible 33 (50%), poor but intelligible 20 (30.3%), and unintelligible 9 cases (13.6%). Normal to easily intelligible were all PMMC only reconstruction.

Esthetic outcome of the various flaps are as follows:

- PMMC only (n = 43): Acceptable 31 (76.7%), good 7 (16.2%), poor 2 (4.6) and failure 3 (6.9%)
- Primary closure (n = 23): Acceptable 8 (34.8%), good 14 (60.9%) and failure 1 (4.3%)
- PMMC + DP (n = 14): Acceptable 2 (14.2%), poor 7 (50%) and failure 5 (37.8%)
- Radial free forearm (n = 3): Acceptable 1 (33.3%), poor 1 (33.3%) and failure 1 (33.3%),
- Nasolabial (n = 2): Acceptable 1 (50%) and poor 1 (50%) , Forehead flap (n = 2): Poor 4 (100%),
- Lateral tongue flap (n = 5): Acceptable 1 (20%) and poor 4 (80%),
- SSG (n = 5): Good 1 (20%), failure 1 (20%) acceptable (2%) and poor 1 (20%).

Overall acceptability was 62.12% in the PMMC reconstruction group, representing 36% (40 out of 111) of the study group.

Our study was comparable to a study performed on PMMC only reconstruction by Tahir *et al*.[21]

DP flap was a part of reconstruction in 15 out of the 111 cases (13.51%) of the study population. Two cases of flap necrosis were noted, 13.33% of the total DP flap reconstruction group. Dehiscence was noted in two cases, 13.33% of the total DP flap reconstruction group. DP alone accounted for 1 out of 18 (5.5%) of the total infection rate and 0.9% of the total study population. PMMC + DP accounted for 5 out of 18 (27.8%) of total infection rate. Therefore, 6 out of 15 reconstructions with DP were infected. The infection rate of DP reconstruction was 40%.

Our findings were similar to a study on DP flap by Gilas[22] who in his study of 678 DP flaps in 604 patients over 20 years reports a flap necrosis rate of 16.6% and overall complication rate of 51.4%.

Radial, free forearm flap, was a part of reconstruction in 3 out of the 111 cases (2.7%) under study. One out of the three radial forearm free flap reconstruction underwent total flap necrosis, 33.3% of the total radial free flap group. Flap survival was 66.7%. Chen *et al*.[23] in a study of 38 radial free forearm reconstruction in 37 patients reported a flap survival of 92% with donor site complication such as poor appearance (8%), reduction of grip strength (11%), and partial skin loss in donor site (11%). The small size of our group prevented us from coming to any conclusion of statistical significance.

CONCLUSION

PMMC which is a major workhorse for head and neck reconstruction formed the majority of reconstruction in this study. Functional outcome after PMMC alone or in combination was acceptable. PMMC reconstruction has a high rate of esthetic acceptance among operated patients. All of the patients with forehead flap reconstruction were not satisfied with the final outcome due to the unsightly donor site. Smaller defects closed by DP flap are relatively accepted better by the patients than with larger flaps. Complications of reconstruction increase with smoking and tumor stage at initial presentation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Acknowledgment

Staff and Management of SAMC and PGI, Indore.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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