11 Open Surgery in Rhino-Orbito-Cerebral Mucormycosis

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The corona virus disease 2019 (COVID-19) infection is caused by the unique severe acute respiratory syndrome coronavirus 2 [1]. COVID-19 infection is associated with a wide spectrum of bacterial and fungal infections. Currently, Indian subcontinent has noticed a surge in mucormycosis. COVID-19-associated mucormycosis (CAM) may be induced by its mutant strain [2], its impact on innate immunity, generated cytokines, and increased risk of diabetes by selective damage of insulin-producing cells in the pancreas [2, 3]. Irrational use of steroids and other COVID-associated treatment has come up as a major risk for CAM.

The progression of CAM is variable amongst the patients. The progression pattern of disease depends on the patient's immune status and the status of comorbidities. Rapidly progressive course mandates aggressive medical and surgical treatment mandatory. Control of underlying immune-compromising condition is initiated as emergency. This chapter will focus on rhino-orbito-cerebral mucormycosis (ROCM). Endoscopic approach has emerged as standard of care for excision of fungal debris, necrotic soft tissues and bone, and also for taking biopsy from suspicious areas. Acute form of invasive fungus has tendency to extend beyond the confines of sinonasal region via natural foramina and perivascular channels and by erosion of surrounding bones making the access difficult with endoscopes, especially in narrow corridors. Involvement of the anterior wall of the maxillary sinus is quite frequently encountered in our practice, which is not accessible with standard endoscopic procedures.

The extended endoscopic approaches (Denker's [4], medial orbital wall removal and extended skull base approach) allow removal of lateral nasal wall, a major part of the anterior wall of the maxillary sinus, medial orbital wall and tissue, skull base and beyond. The procedure, however, requires expertise, good assistance and specialized instruments. The angle of surgical instrumentation via the nostrils in Denker's approach narrows down the working area during the removal of lateral part of anterior wall of maxilla and soft tissues of cheek. The exposure of the superior part of the anterior wall of maxilla, zygomatic arch, and anterior orbital contents is difficult even with experienced surgeons [5, 6]. The simultaneous presence of skin discoloration/blackening of the anterior face is a contraindication for endoscopic approach. The surgical volumes of ROCM cases encountered during CAM epidemic also created feasibility challenges. Operating in multiple unfriendly suites, COVID-positive and medically unstable patients mandated procedures to be completed fast and

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efficient. All these scenarios mandated open surgical procedures (OSP) in the management of ROCM.

Most of the cases of ROCM can be managed with endoscopic approach. However, endoscopic procedures take much longer, are equipment intensive and need assistants. In a tertiary hospital like ours that happened to be one of the largest COVID facility also, there was an acute shortage of manpower, and everyone was expected to act fast. There was a heavy load of ROCM patients needing immediate attention. Therefore, patients were managed with OSP. OSP allows excellent visualization of all those sinonasal areas that may be hidden in the endoscopic approaches. The surgical procedures have a short learning curve and can be performed with a limited number of instruments. A proper tissue handling limits the visibility of surgical scar.

Patients of ROCM with the following symptoms were managed using OSP:

1. Numbness of cheek, medial canthus and supraorbital region.
2. Significant facial fullness indicating anterior cheek spread (Fig. 11.1a).
3. Facial discoloration/blackening and involvement of subcutaneous tissue (Fig. 11.1b).
4. Palatal discoloration, palatal perforation, and palatal perforation and loosening of teeth (Fig. 11.1c).

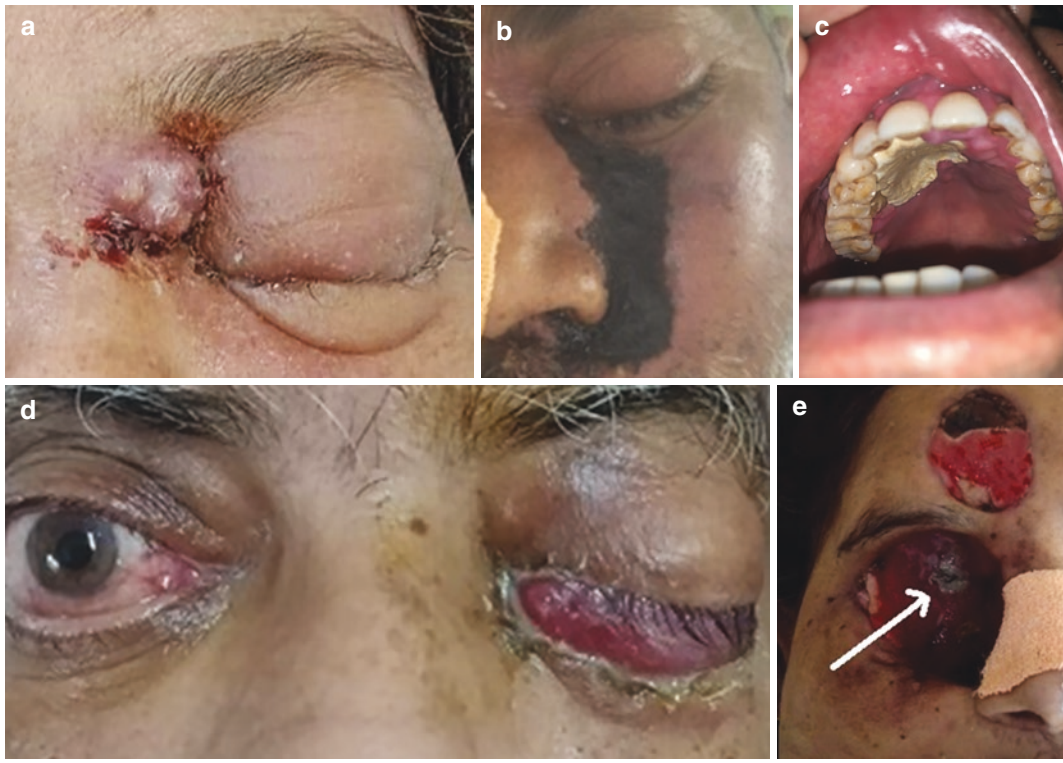

Fig. 11.1 (**a**) Clinical photograph showing extension of disease to the lacrimal drainage system with cutaneous invasion and facial fullness. (**b**) Clinical photograph showing facial discoloration indicating gross bone invasion by the disease leading to the fungal involvement of the skin after erosion of anterior wall of maxilla. (**c**) Bogginess of the palate and ulceration and loosening of teeth. (**d**) Ptosis, proptosis and conjunctival chemosis on the left. (**e**) A case of post orbital exenteration showing intracranial progression of residual/impending disease in ROCM

5. Proptosis/discoloration of orbital contents with loss of vision (Fig. 11.1b).
6. Failed extended endoscopic procedure.
7. Intracranial extension of disease (Fig. 11.1e).

11.1 Role of Radiology in Surgical Planning

Contrast CT and MRI [1] are the advised radiological investigations. In view of sudden surge in ROCM cases needing immediate surgical debridement and imitations of MRI facilities across all surgical units of the hospital, contrast-enhanced computed tomography (CECT) was taken as the acceptable preoperative radiological investigation. CECT is easy to perform, cost effective and quicker and provides details of bony anatomy. CECT also provides worthwhile anatomical information of orbital and cranial tissue involvement. Contrast-enhanced MRI was reserved for cases with orbital involvement and suspected intracranial spread and for follow-up assessment. MR angiography was used to assess superior ophthalmic artery and central retinal artery in selected cases. The radiological indications for OSP are:

1. Obliteration of pre-antral fat plan (Fig. 11.2a).
2. Simultaneous obliteration of pre-antral and retro-antral fat plans (Fig. 11.2a, b).
3. Collection of fluid anterior to the anterior wall of the maxilla.
4. Erosion/destruction of anterior/inferior wall of maxilla.
5. Residual disease in anterior wall/inferior wall of maxilla after extended endoscopic procedures (Fig. 11.2b).
6. Gross intra-orbital disease (Fig. 11.2c).
7. Lateral orbital wall erosion.
8. Central retinal artery occlusion.
9. Intracranial extension of disease (Fig. 11.2d). Though minimal intracranial extension can be accessed endoscopically, large abscesses and necrosis mandates neurosurgical assistance and craniotomy.

11.2 Diagnosis

The diagnosis is established by a specific clinical profile of the patient. The risk factors in the current surge are the history of COVID-19 infection, uncontrolled diabetes, uncontrolled kidney disease, transplanted severely immunocompromised patients, prolonged use of immunity-lowering drugs (steroids, chemotherapy, etc.). A good clinical assessment (discoloration and crusting of sinonasal region) is required to confirm the diagnosis. Nasal endoscopic assessment is performed to obtain certain information like tissue appearance and disease extent. Tissue biopsy is done for fungal smear and culture, and histopathology. Simultaneous assessment of blood and serum parameters is done to prepare the patient for surgery, to control the comorbidities and to obtain effectiveness of antifungal therapy.

11.3 Surgical Procedures

11.3.1 Sublabial Approach [4, 5, 7, 8]

The indications for sublabial approach are infrastructural maxillectomy, removal of subcutaneous tissue and anterior wall of maxilla, and combined removal of anterior and medial wall of the maxilla (Denker's procedure). The approach allows good exposure of the anterior wall of the maxilla. The incision is made 3–5 mm above the upper gingivo-buccal sulcus (Fig. 11.3a). The incision is deepened till the anterior wall of the maxilla. The periosteum elevator is used to elevate the tissue from the anterior wall of the maxilla. Often pus can be found collecting subperiosteally via infraorbital foramina [2, 3]. Canine fossa is used to enter the maxillary sinus in Caldwell Luc approach by chisel, osteotome or drill. The widening of the anterior wall window is done in all directions with Kerrison rongeur, bone nibbler or drill. The procedure allows simultaneous removal of unhealthy subcutaneous tissue, medial, superior and posterior wall of maxilla. The wound is closed with 3-0 vicryl suture in layers.

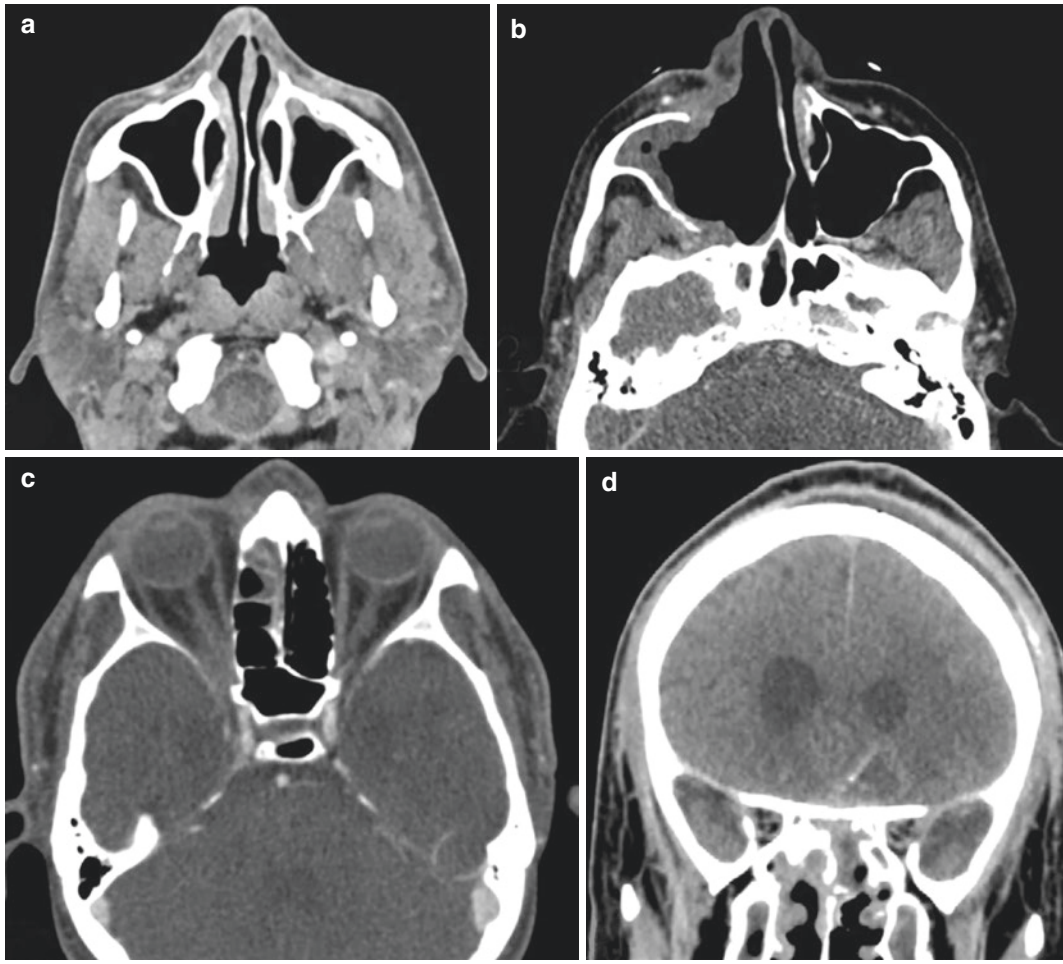

Fig. 11.2 (**a**) Radiological findings in cases treated with open approaches showing obliteration of pre-antral fat on left. Simultaneous obliteration of pre-antral and retro-antral fat plans. (**b**) Obliteration & infilteration of anterior wall of maxillary in post endoscopic maxillectomy case. (**c**) Gross intra-orbital disease. (**d**) Left anterior cranial cavity showing rim enhancement with hypointense mass

Infrastructural maxillectomy is done by incising the palatal mucosa medial and posterior to the diseased tissue with adequate margin (Fig. 11.3b). The osteotomy is made below the inferior orbital fissure from pyriform aperture till the posterior limit of the anterior wall of the maxilla. Curved osteotome is used to separate the posterior maxilla from the pterygoid plates. The heavy scissors are used to cut soft tissue around specimen. Internal maxillary artery bleed is secured after removal of the anterior specimen. Involved pterygoid plates, pterygoid muscles, involved infratemporal fossa contents and involved nasopharyngeal tissue can be removed after the removal of the anterior specimen. Pterygoid plexus ooze is managed by cautery, digital pressure and surgery. The conventional pack is placed after creating a bed with suture. The artificial palate can be applied in the same sitting if available. The pack removal is generally done after 2–3 days. Ryles tube feeding can be given in non-rehabilitated patients till rehabilitation with artificial palate.

The limitations for sublabial approach are the exposure of the supero-lateral part of anterior wall of maxilla, superior part of the ethmoid

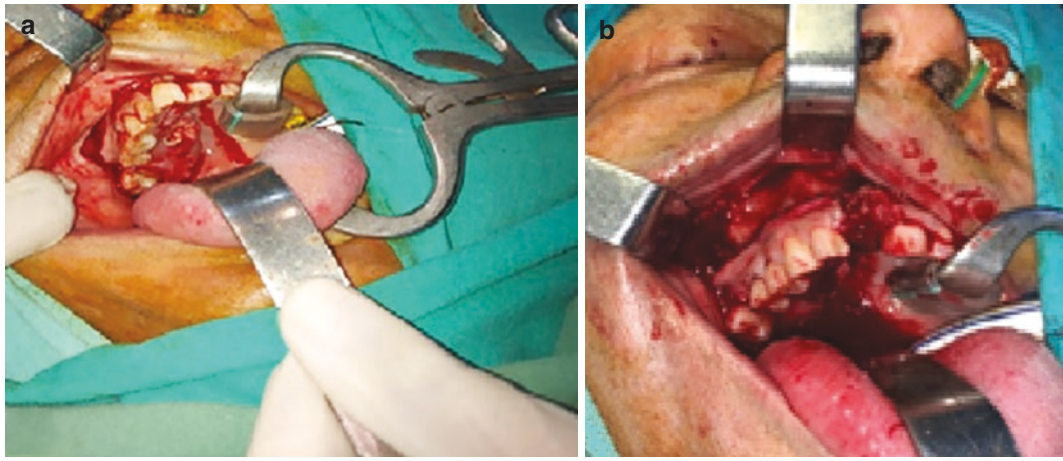

Fig. 11.3 (**a**) Intraoperative photographs showing palatal excision by sublabial approach. (**b**) Palatal incision and separation of right inferior maxilla from the rest of the maxilla

sinus, frontal sinus, sphenoid sinus and anterior-superior orbital tissue. Certain limitations of procedure can be overcome by using endoscopic assistance.

11.3.2 Lateral Rhinotomy Approach [5, 9, 10]

It is the best open approach to deal with the sino-nasal pathology. The procedure allows exposure of complete maxilla, anterior maxillary contents, retro-maxillary space (infratemporal fossa, pterygomaxillary fossa), nasal cavity, and ethmoid and sphenoid sinus. The incision is made along the naso-facial groove from medial canthus. The incision is curved around the nasal ala (Fig. 11.4). The incision is deepened till the frontal process of maxilla and pyriform aperture. The nasal cavity is being entered along the pyriform aperture. The medial flap is secured with stay suture anteriorly for easy handling of nasal tissue. The lacrimal sac is being lifted from the lacrimal fossa. The nasolacrimal duct is cut with a sharp instrument. The flap is elevated subperiosteally to expose the anterior wall of the maxilla.

After lip splitting incision the lateral flap is reflected laterally. This facilitates easy manoeuvring of the soft tissue contents present anterior, lateral, inferior and posterior to maxillary bone. Hence the lateral rhinotomy incision can be combined with lip splitting incision to deal with these para-maxillary areas if required (Fig. 11.5). The debridement of tissue is performed till clinical necrosis is found. The presence of bleeding margin is suggestive of healthy tissue.

For total maxillectomy, the osteotomy is made at the level of the frontal process of the maxilla, zygomatic arch, hard palate and retro-maxillary areas to separate the maxillary bone from surrounding tissues [5].

Combining Lynch Howarth incision with lateral rhinotomy incision allows clearance of disease from frontal sinus, medial orbit and anterior skull base (Fig. 11.6). The flap is raised by elevating the periorbita from lamina papyracea. Lamina papyracea with ethmoid air cells is removed to access the nasal cavity. The upper limit of lamina papyracea removal is the fronto-ethmoid suture line. Kerrison rongeur is used to remove the frontal sinus anterior wall for access in the frontal sinus. The skull base is identified at the ethmoid roof and followed posteriorly to clear the disease along the skull base and sphenoid sinus. Involved medial orbital fat, periorbita and medial muscles can be removed by the same approach. The posterior septum can be removed to access the contralateral side of the nasal cavity if required. Endoscopic assistance provides superior quality view when disease extends close to vital

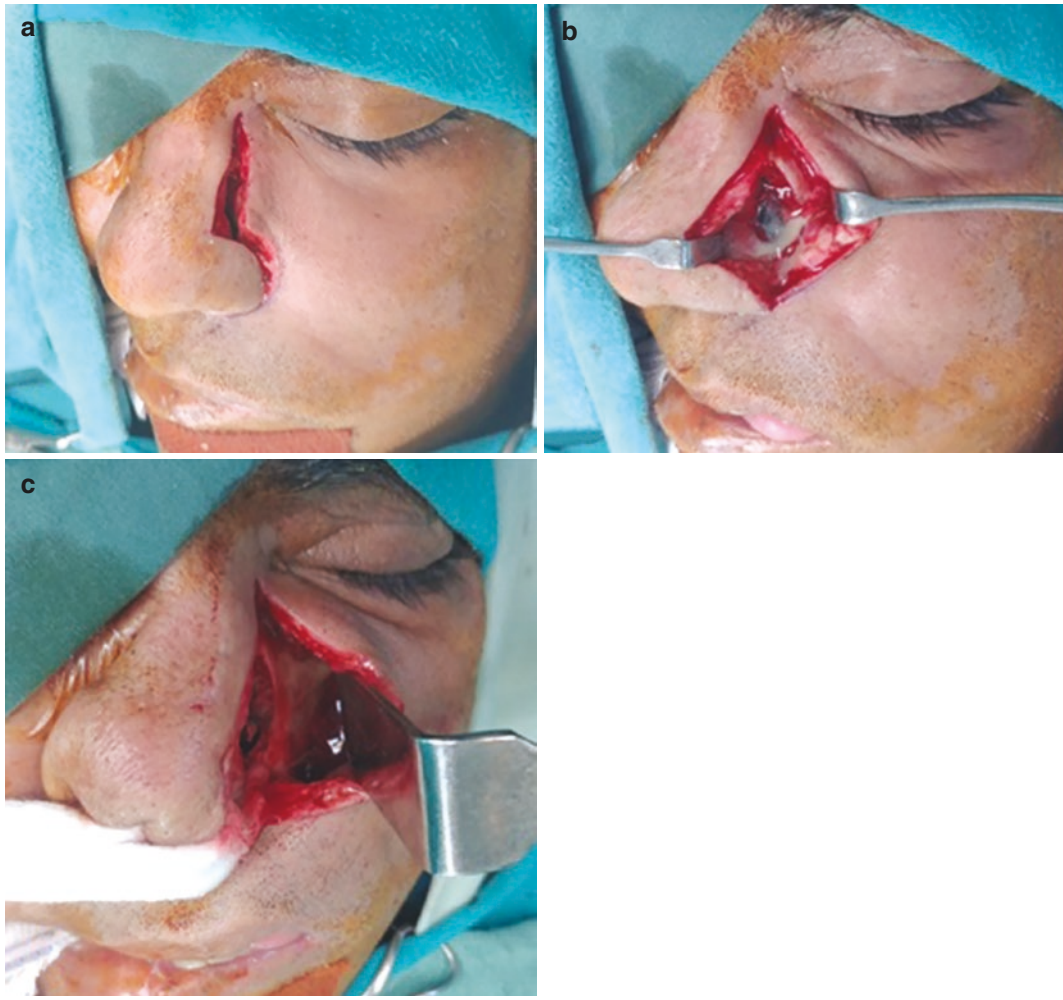

Fig. 11.4 (**a**) Lateral rhinotomy incision. (**b**) Mucopurulent discharge can be seen pouring out as soon as the flap is raised. (**c**) Exposure of anterior wall of maxilla and the nasal cavity

structures like skull base and orbital tissue. The procedure has certain limitations in dealing with lateral orbital contents, zygomatic bone and temporal fossa, and in the case of need of simultaneous orbital exenteration.

11.3.3 Weber Ferguson Approach [5]

It allows complete exposure of maxillary bone, para-maxillary spaces, orbital tissue, infratemporal fossa, zygomatic bone, lateral orbital wall and temporal fossa (Fig. 11.7). It can be combined with bi-coronal incision to handle the anterior skull-base contents. Orbital exenteration is mostly done with this incision. Sub-ciliary incision is combined with lateral rhinotomy incision. Lateral extension of incision is based on the lateral limit of the disease. Sub-ciliary incision runs 3–5 mm inferior to the inferior lid margin. Sub-ciliary incision is combined with supra-ciliary incision to remove lid edges. The flaps are raised as mentioned in the lateral rhinotomy approach. The orbital tissue is released from the anterior orbital rim by incising periorbita. Medial and lateral canthal ligaments are

incised. The plane is created subperiosteally around the orbital contents till the orbital apex. The curved artery clap is applied at the level of the orbital apex. Heavy scissor is used to cut the tissue from the orbital apex anterior to clamp. Further slicing of orbital apex tissue can be done after removal of the anterior orbital contents. The left-out orbital apex tissue is sutured or cauterized to limit the chances of ophthalmic artery bleeding. The cut edges of lids are sutured to prevent contamination from external environment. The cavity is filled with medicated packs. The dental rehabilitation is done with palatal prosthesis by placing it intraoperatively or in an early postoperative period.

Fig. 11.5 Lip splitting incision allows easy lateral access

Fig. 11.6 Combination of lateral rhinotomy with Lynch Howarth incision

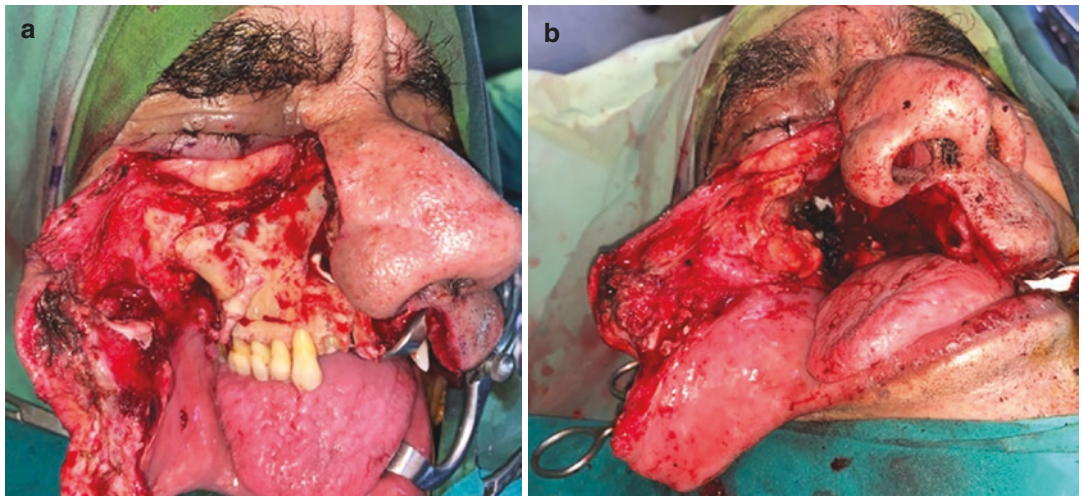

Fig. 11.7 (**a**) Weber Ferguson approach allows complete exposure of maxillary bone and peri-antral tissue. (**b**) Surgicel is visible after maxillectomy in intra-temporal fossa

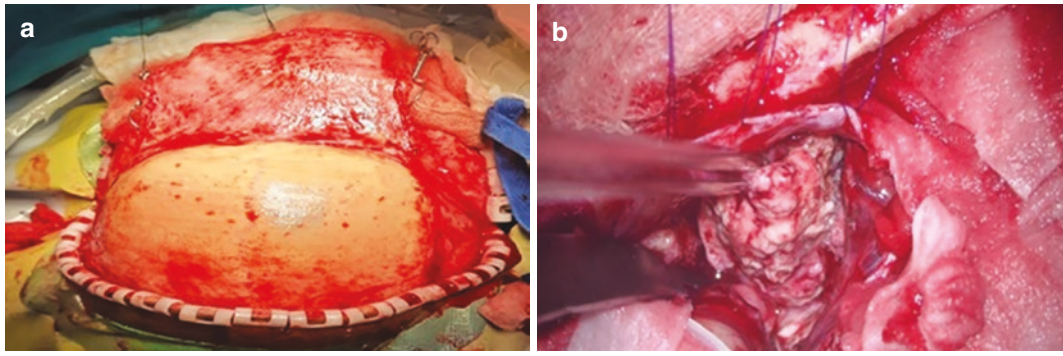

Fig. 11.8 (**a**) Bi-coronal incision and flap. (**b**) Microscopic picture of fungal mass

11.3.4 Transcranial Approach [5–13]

Anterior and middle cranial fossa is mostly invaded by the CAM. The routes of extension are cribriform plate, orbital apex, roof and sphenoid body. The cerebral involvement is considered life threatening as surgical excision is difficult and amphotericin has limitations in crossing blood brain barrier. The surgical excision may have survival advantage in the early stage of cerebral extension. Bi-coronal approach or extended endoscopic approach is the most preferred approach as it allows exposure of the entire anterior skull base.

Bi-coronal incision is made a few centimetres behind the hairline (Fig. 11.8a). The flap is elevated over pericranium till the orbital rim. The wide pericranial flap is made for the postoperative skull base reconstruction. The osteotomy is made, and brain tissue is elevated from the skull base. The diseased tissue is removed (Fig. 11.8b).

11.4 Postoperative Management

Surgical cavity pack is removed on the 2–3 postoperative days. The nasal douche is advised after pack removal to prevent mucosal dryness. The cavity cleaning is done regularly for early epithelialization. Facial suture removal is done between seventh and tenth postoperative days. Palatal prosthesis is placed intraoperatively or after a certain interval from surgery for oral rehabilitation. Ryle's tube feeding is removed after placement of prosthesis. Dressing is applied over orbital exenteration sites to prevent crusting. The open surgical cavity can be obliterated by flaps or free tissue transfer after a certain interval from complete treatment. The patient can wear dark goggles for cosmetic issues till an artificial eye is applied.

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