Case Presentation

A 45-year-old female presented with a history of right-side nasal obstruction and epistaxis over a three month period. Anterior rhinoscopy revealed a fleshy, non tender red mass in the right nostril which was completely blocking the nasal passage. There was no palpable regional lymph node. A CT scan showed a space-occupying lesion filling the right nasal cavity totally obstructing the maxillary ostium. Resection of the mass and an anterior ethmoidectomy were done under general anesthesia. Histopathology study revealed melanocytes in the sub mucosa. The stroma contained a large number of spindle-shaped cells with hyperchromatic nuclei. The histopathological findings led to the diagnosis of malignant melanoma. A metastatic workup including CT chest, abdomen, and LFT was done and was negative for tumor.

One month later, a follow up CT of the sinus was done and revealed ethmoidal air cells opacification (Figure 1). A biopsy was taken and the pathology was in favor of malignant melanoma. The following week, the patient underwent resection of the right cribiform plate with right medial maxillectomy through a right lateral rhinotomy and coronal bifrontal approach (Figures 2 and 3). The histopathological findings revealed malignant melanoma with margins free of disease. The patient received chemotherapy and remained free of disease till 4 month following the surgery when she was found to have liver metastasis with no local recurrence. The patient passed away two month later.

Discussion

Melanomas are tumors arising from melanocytes which are neuroectodermally-derived cells located in the basal layers of skin, skin adnexas and some of the mucosal membrane. Common sites for melanomas are the head, neck and lower extremities as they are exposed to sunlight, which is one of the predisposing factors. Less commonly, they occur in the oral and genital mucosa, nail beds, conjunctiva, orbit, esophagus, nasal mucosa or nasopharynx, vagina and leptomeninges. The cause of melanoma in solar-hidden mucosa is unclear, although smoking and exposure to formaldehyde may have a role in activation of pre-existing melanocytes leading to melanogenic metaplasia.

Malignant melanoma of the nose was first described by Lucke in 1869 (Cove). The incidence of malignant melanoma in the nose and paranasal sinuses ranges between 0.5% and 1%. There is no predilection for gender, and the most commonly affected age group is 5th or 6th decade. There is no apparent correlation with chronic irritation, infection or allergy. It is primarily a tumor of the nasal cavity arising from the nasal septum, lateral wall, inferior turbinates and rarely the floor and roof of the nose. Its presence in the paranasal sinuses is due to extension.

In the nose and paranasal sinuses, malignant melanoma generally does not present dramatically. The majority of patients with nasal cavity primaries present with nasal obstruction and epistaxis. Proptosis, diplopia, pain and facial deformity are less common and are indicative of advanced disease. Malignant melanomas of the nasal cavity and sinuses are characterized by early and repeated recurrences. Nasal and sinus melanomas usually are advanced at
the time of discovery. Metastases are often found at initial presentation for both cutaneous and mucosal melanomas. Due to lack of any characteristic clinical features, the diagnosis is often based on the histopathological examination. One has to differentiate malignant melanoma clinically from polypi and other tumors. On light microscopy, sheets of closely arranged sphenoidal cells or spindle cells and multinucleated giant cells are seen. Pigmentation is variable. An amelanotic melanoma has to be differentiated from anaplastic carcinoma.

The mainstay of treatment remains surgical excision. The preferred treatment for sinonasal mucosal melanoma is wide excision with tumor free margins. Different surgical procedures like lateral rhinotomy, craniofacial resection, maxillectomy and total rhinectomy are done depending on the extent of the disease. Gilligan et al reported absolute local control by radiotherapy alone in 61% of cases. Melanoma is a relatively chemo resistant tumor; thus, the main role of chemotherapy remains as palliative treatment in the setting of disseminated disease. Dacarbazine is currently the only chemotherapeutic agent approved for the treatment of advanced disease. Recent studies showed that immunotherapy might have a role in the treatment of malignant melanomas.

The overall prognosis and survival rate in patients with these tumors is very poor, with the 5-year survival rate ranging between 10% and 40%; the median survival is 21-24 months. The single most powerful predictor of survival is the absence of regional lymph nodes. Some of the contributing reasons for the poor prognosis of sinonasal mucosal melanoma are the delay in detection and inaccurate histological diagnosis.

References