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CASE REPORT

Diagnostic and Surgical Challenge in the Management of a Rare Tumor of Skull Base and Face

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ABSTRACT

Aim: We report a rare case of lateral skull base hamartoma. The diagnostic dilemma, the therapeutic challenge and the importance of interdisciplinary approach to decrease the morbidity has been discussed.

Method: We report a rare case of a 48-year-old male with swelling over the left side of face and cheek for a duration of 12 years, who presented to us only for his cosmetic deformity and no functional impairment, after a thorough clinical examination and series of invasive and noninvasive investigations, was diagnosed to have a rare tumor hamartoma of skull base and face. A complete surgical excision without morbidity was made possible only by a multidisciplinary team approach. The investigation and surgical approach to reduce per operative morbidity has been discussed. Microscopic examination of the excised specimen with special staining techniques was conclusive for hamartoma. Patient has been on a regular follow-up for the last 2 years with no evidence of recurrence and functional compromise.

Discussion: The rarity of the tumor, the difficulty in diagnosis, management and prognosis of hamartoma have been discussed.

Conclusion: Hamartoma of the infratemporal fossa is a rare tumor and those originating in this surgically challenging site are even rarer. Further their close proximity to vital structures in head and neck poses a formidable challenge in their diagnosis and surgical management to achieve a good cosmetic and functional result.

Keywords: Hamartoma, Infratemporal fossa, Embolization.

INTRODUCTION

Hamartoma as defined in the Webster's medical dictionary as 'a mass resembling a tumor that represents anomalous development of tissue natural to part or an organ rather than a true tumor'.[1] They grow along with and at the same rate as the parent tissue unlike a cancerous tumor. The hamartoma word is originated from hamartia in Greek alphabet and means 'scribal error or mistake'. Hamartomas may be seen anywhere in the body and are often seen in infancy and childhood. Therefore, it has been believed that these lesions are developmental aberrations. They rarely invade or compress adjacent structures significantly. The most common hamartomas occur in lung, accounting for 75% of all the benign lung tumors. It remains a rare finding in the head and neck region, particularly those in the infratemporal region are clinically silent,[2] hence, it may lead to a misdiagnosis of a tumor in the presence of epithelial proliferation. Inspite of not being a neoplasm in a true sense, the clinicopathological behavior of a hamartoma is like that of a neoplasm and it is known to recur if excision is incomplete.

CASE REPORT

A 48-year-old male was referred to us by the interventional radiologist with complaints of swelling over left side of the face and the cheek for 12 years. Onset was insidious and progressively increased in size with a rapid increase in the last 2 years. Swelling was associated with mild discomfort on swallowing. There were no complaints of facial paresthesia, restriction of ocular movements or double vision. Local examination revealed a 5 × 8 cm bilobed swelling in the temporal and the infratemporal region which was firm in consistency, nontender, compressible, nonpulsatile and nonreducible (Figs 1 and 2).

CT scan and MR angiogram showed well-encapsulated soft tissue mass involving the left zygomatic, buccal, masseteric and the infratemporal spaces with no evidence of invasion of neighboring structures and bony erosion except for the thinning of the cortex of the zygomatic bone (Fig. 3). Four vessel angiograms showed tumor neovascularization by internal maxillary and facial artery (Fig. 4).

With these investigations he was diagnosed to have a vascular tumor of the face and skull base. A direct puncture tumor embolization with trucut biopsy of the lesion was done which was inconclusive. Surgical excision of the tumor was planned 72 hours postembolization jointly by ENT and oral maxillofacial surgeons. Tumor was excised *in toto* by a dual approach through a Weber Fergusson and modified transtemporal. The excised specimen was bilobed (Fig. 5) and measured about 4×5 cm and 3×5 cm. Postoperative period was uneventful. Histopathological examination of

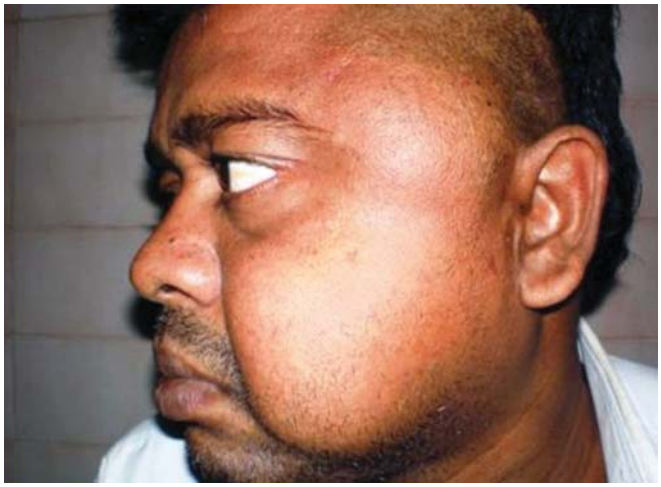

Fig. 1: Patient profile lateral view at presentation

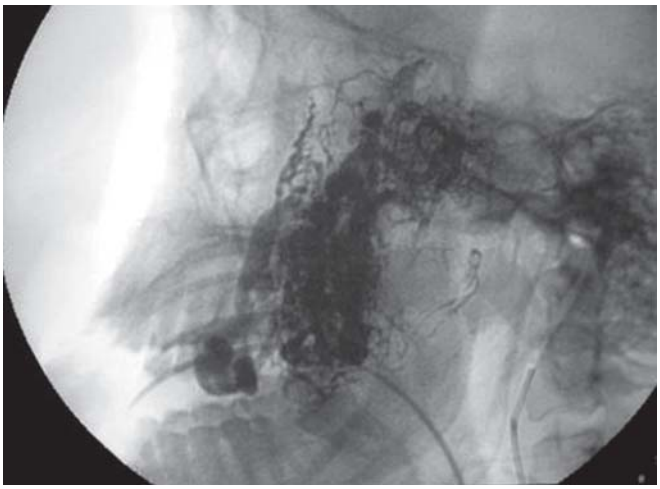

Fig. 4: Four vessel angiogram

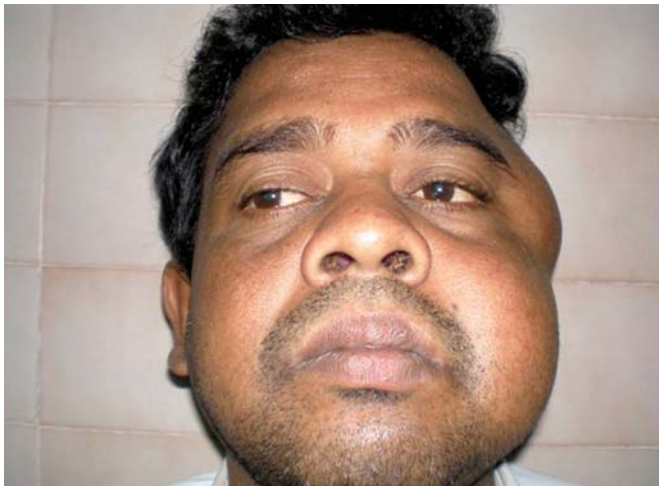

Fig. 2: At presentation showing the frontal profile view

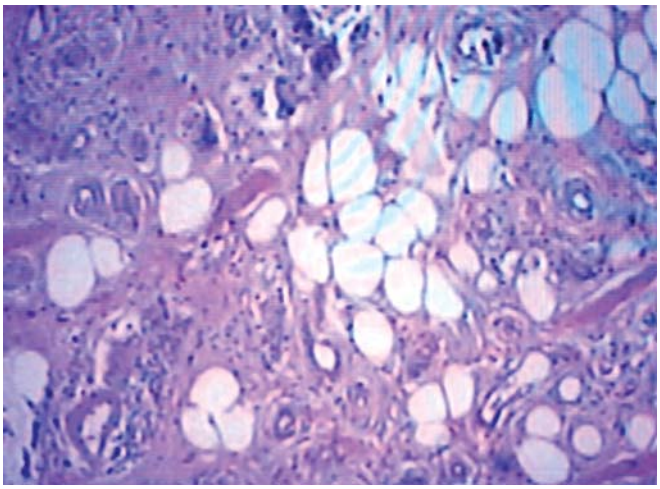

Fig. 5: Postoperative specimen

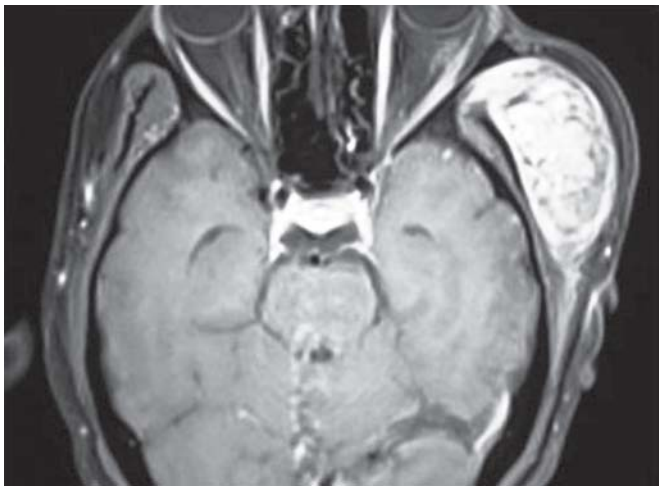

Fig. 3: MR angiogram

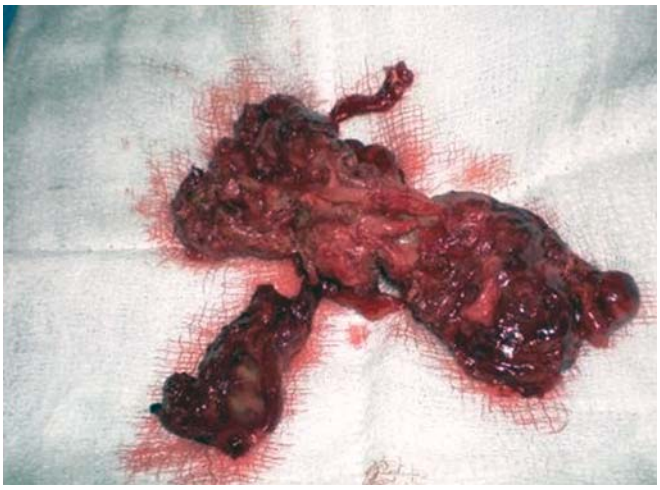

Fig. 6: Histopathological picture of the excised tissue

the tissue showed thick-walled arteries with proliferating capillaries, adipose tissue and skeletal muscle bundle which clinched the diagnosis of hamartoma (Fig. 6). Patient was discharged on the 7th postoperative day and has been doing well and is on regular follow-up for the last 2 years (Fig. 7).

DISCUSSION

Hamartoma may occur in any region as it designates a focal overgrowth of mature normal cells and tissues at site of identical cellular composition. Hamartomas of the head and neck region are uncommon those occurring in the

Fig. 7: Postoperative follow-up after 2 years

infratemporal region are even rarer.[3] The infratemporal fossa region by virtue of its relatively concealed location, is inaccessible for clinical and endoscopic examination.

They usually are asymptomatic or dormant as in our case, but when symptoms appear only when there is some functional impairment or cosmetic disfigurement.[2] Fortunately, there has been no functional compromise in our patient, the only symptom was cosmetic deformity. The epithelial and mesenchymal hamartomas are uncommon in the head and neck region. Hamartomas have been described in the nasal cavity, nasopharnx,[10] oropharynx,[5] retropharyngeal,[6] hypopharynx,[7] larynx,[2] ear,[8] Eustachian tube[9] and the deep neck spaces. Intracranial lesions like the extracerebral glioneural hamartoma extending into the parapharyngeal space have also been described. They have no tendency to regress spontaneously. Nevertheless they are prone for recurrences especially when excised incompletely. This mixed bag of characteristics has disputed the neoplastic origin of this uncommon lesion.[11]

Hamartomas can be classified into (a) mesodermal component containing type that is more frequent and (b) the epithelial or glandular element containing type that is less frequent.[12] Microscopically the vascular hamartomas contain fibrocollagenous tissue, adipose tissue, skeletal muscle fibers, few nerve bundles, immature or primitive mesenchyme along with blood vessels interspersed between them, as was seen in our case. This picture could masquerade an angiofibroma, hemangioma, lymphangioma or a teratoma.

CT scan with contrast is an indispensable diagnostic tool in these cases to know the nature of the mass, consistency, extensions and attachments of the mass. MR angiography may additionally reveal the vascularity of the tumor. Preoperative embolization of the tumor is essential as in our case the four vessel angiogram which showed a highly vascular tumor with a A-V malformation between the maxillary artery and the facial vein. Angiography identifies the vascularity of the tumor and its relationship to the ICA and demonstrates the cerebral circulation and its collateral vasculature. Neither of these investigations predicts the adequacy of the intracranial collateral blood supply after sacrifice of the internal carotid artery.

Management is by complete surgical excision through a wide open surgical approach to prevent recurrence. The ideal time advocated postembolization of such a vascular tumor is within 72 hours. Various surgical approaches to ITF have been described.[13] The commonly used approaches to the ITF are Caldwell-Luc, lateral and transcranial. Other approaches are inferior, extradural zygomatic middle fossa approach, subtemporal-infratemporal approach and orbitozygomatic-extradural approach. The Caldwell-Luc approach provides limited exposure and can be modified by incorporating a Weber-Fergusson incision. Given the radiographic and clinical findings, the tumor of present case was believed to be benign. In our case, we approached the skull base through a combined modified transtemporal Alkyte-Bramhley incision and Weber-Fergusson approach. This not only gave a good surgical exposure but also helped in limiting the postoperative morbidity and functional loss in the patient.

CONCLUSION

Hamartomas of the face, skull base and infratemporal regions are rare. Surgical excision is the best treatment modality when symptomatic. Excision should be complete to avoid recurrence. Careful planning of the surgical procedure with preoperative work-up along with preoperative embolization limits the amount of blood loss to a great extent. In the present case, team work and multimodal approach resulted in a successful outcome without significant morbidity.

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