



Giant cell tumors in temporomandibular joint region involving lateral skull base: A literature review

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Abstract

Purpose: To review the literature associated with giant cell tumors in temporomandibular joint and lateral skull base region.

Materials and Methods: A review of the literature was performed to record the tumor location, patient's personal data, and extent of resection, radiotherapy, recurrence, clinical features, and Follow-up.

Results: Nine articles were identified and reviewed in the presented study. The most common clinical symptoms in GCTs in the TMJ-SB region were malocclusion and restriction of mouth opening, other symptoms such as persistent headache and hypoacusis also were not rare because of skeletal invasion of the skull base and ears. Skull base GCTs occur most commonly in young adults with an equal sex distribution. The total removal of giant cell tumors in TMJ without adjuvant radiotherapy is the most effective treatment for localized disease.

Conclusion: GCTs of the skull commonly affect young adults, with an equal sex distribution, and are most often centered in temporal bone. Total removal is associated with the lowest recurrence rate and should be the goal of treatment. If total removal cannot be achieved, the combination of subtotal removal and radiation results in a similar recurrence rate.

Keywords: giant cell tumor, lateral skull base, temporomandibular joint, temporal bone, radiotherapy

1. Introduction

Giant cell tumors (GCTs) are a group of rare benign neoplasms that are most commonly found in the epiphysis of long bones. Around 1–2% of these lesions present in the head and neck, with the skull base being a commonly reported site (temporal, sphenoid and ethmoid bones) and other sites including the mandible, maxilla, nasal cavity, thyroid, larynx, hyoid, tongue and the soft tissues of the neck. In the skull base the temporal bone is a common site of occurrence of GCTs. Although benign, these tumors have a locally destructive character which can be potentially dangerous in the presence of the intricate neurovasculature of the temporal bone and skull base [1].

To date, only a few reports of GCTs arising in the mandible, larynx, sphenoid, and hyoid bone can be found in the literature. Most of them occurred in the temporomandibular

joint and skull base (TMJ-SB) region or involved the lateral skull base [2].

The Purpose of this study is review the literature associated with giant cell tumors in temporomandibular joint and lateral skull base region.

2. Materials and methods

Using a PubMed literature search, we identified and reviewed papers using these key words: giant cell tumor, lateral Skull base, temporomandibular joint, temporal bone, and radiotherapy.

Papers were retrieved from 1987 to 2016 and we recorded the tumor location, patient's personal data, extent of resection, radiotherapy, recurrence, clinical features, and Follow-up (Table 1).

Table 1: Summary of GCT Cases. 1987 – 2016

Study reference	Age/sex	Clinical features	Location	Extent of excision	Adjuvant therapy	Recurrence	Follow-up
Findlay <i>et al.</i> , (1987) [3]	23/M	Hearing loss, otalgia, partial FN paralysis, V never hypesthesia	Temporal	Total removal	RT	No	8 months
Pai <i>et al.</i> , (2005) [4]	26/M	Hearing loss, tinnitus, swelling, FN paralysis, CHL	Temporal	total removal	None	No	1 year
Elder <i>et al.</i> , (2007) [5]	2/F	Mass behind ear, aural discomfort	Temporal	Total removal	None	No	13 months
	7 weeks /F	mass in the EAC	Temporal	Total removal	None	No	11 months
Matsushige <i>et al.</i> , (2008) [6]	77/F	Headache, vomiting dizziness, temporal hemorrhage	Temporal	Total removal	None	No	1 year
Isaacson <i>et al.</i> , (2009) [7]	42/M	Total hearing loss, EAC bulge.	Temporal	Total removal	None	No	3 years
	47/M	Recurrence with otalgia and aural fullness, prior surgery with MCF and subtotal clearance, MHL	Temporal	Total removal	None	No	10 years

Zizuka <i>et al</i> , (2012) ^[8]	32/M	Left aural hearing loss, tinnitus, fullness, EAC bulge, CHL	Temporal	Total removal	None	No	4 years
Zhang <i>et al</i> , (2013) ^[9]	34/ M	Preauricular pain, temporomandibular symptoms,	Temporal	subtotal removal	None	Yes	18 months
	27/ F	Headache, dizziness, temporal hemorrhage	Temporal	subtotal removal	RT	No	24 months
	52/ M	Headache, vomiting dizziness, temporal hemorrhage	Temporal	subtotal removal	RT	No	33 months
	44/ M	Total hearing loss, EAC bulge.	Temporal	Total removal	RT	No	21 months
	54/ F	Preauricular pain, temporomandibular symptoms,	Temporal	Total removal	None	No	7 months
	19/ M	Preauricular pain, temporomandibular symptoms	Temporal	Total removal	RT	No	31 months
	33/ M	Preauricular pain, temporomandibular symptoms	Temporal	Total removal	None	No	12 months
Prasad <i>et al</i> , (2014) ^[1]	35/ F	Total hearing loss, EAC bulge.	Temporal	Total removal	None	No	11 months
	36/M	preauricular mass, hearing loss, tinnitus, temporoparietal, pain	Temporal	Total removal	None	No	120 months
	31/F	hearing loss, tinnitus, HTN, stroke with hemiparesis, SLE	Temporal	Total removal	None	No	96 months
	46/M	temporal swelling, hearing loss, tinnitus, temporoparietal pain	Temporal	Total removal	None	No	48 months
	67/M	hearing loss, tinnitus, vertigo, HTN, vocal cord palsy due to previous thyroid surgery	Temporal	subtotal removal	RT	Yes	24 months
Yi Sben <i>et al</i> , (2016) ^[2]	57/M	hearing loss, tinnitus	Temporal	Total removal	None	No	15 months
	29/ M	swelling in the left temporomandibular joint	TMG	Total removal	None	No	5 Years
	42/ M	left-sided headache and temporal numbness presented to the outpatient clinic for treatment	Temporal	Total removal	None	No	5 Years
	71/ F	trismus, malocclusion, hearing loss, and facial paralysis.	Temporal	Total removal	None	No	12 months

FN = Facial nerve; V nerve = trigeminal nerve; RT = radiotherapy; GCT= giant cell tumor; TMJ= temporomandibular joint. (EAC)= external auditory canal. F= Female. M= male. CHL= conductive hearing loss. SLE = systemic lupus erythematosus; HTN = hypertension; MHL = Mixed hearing loss; MCF= Middle cranial fossa

3. Results

Of 9 papers published on this topic between 1987 and 2016, we were able to retrieve and interpret all of them (Table 1). The most common clinical symptoms in GCTs in the TMJ-SB region were malocclusion and restriction of mouth opening because of the temporomandibular joints, zygomatic arches, and mandibles were the anatomic structures affected by these GCTs in the TMJ-SB region. Other symptoms such as persistent headache and hypoacusis also were not rare because of skeletal invasion of the skull base and ears.

Skull base GCTs occur most commonly in young adults with an equal sex distribution. The total removal of giant cell tumors in TMJ without adjuvant radiotherapy is the most effective treatment for localized disease and avoids the risks associated with radiation exposure, including radiation damage to normal surrounding tissues. If total removal is not feasible, then subtotal removal with adjuvant radiotherapy provides acceptable tumor control rates.

4. Discussion

To GCTs of bone arise from differentiated mesenchymal cells of the bone marrow. The majority of GCTs are found in the epiphysis of long bones, with <2% involving the skull ^[10, 11]. When the cranium is involved, GCTs most commonly arise from the floor of the middle fossa, specifically the temporal and sphenoid bones ^[3, 12]. Males and females are usually affected equally, and although most are considered benign, GCTs of the skull base may cause extensive local destruction,

as seen in our patient. Primary tumors arising from the skull base vary widely based on their cell of origin and include osteoma and osteoblastoma, chondroma, chondroblastoma, chondrosarcoma, chordoma, hemangioma, meningioma, and cholesteatoma ^[3]; however, bone replacement and mastoid tegmen invasion, also seen in our patient, are less common in these other tumors. Patients with temporal GCTs commonly present with conductive hearing loss, aural fullness, pain behind the ears, and facial weakness ^[8]. In contrast, sphenoid GCTs present with headaches, facial hypoesthesia, vision failure, and ophthalmoparesis ^[11]. Although GCTs commonly invade the infratemporal fossa, interfering with middle ear structures and resulting in conductive hearing loss ^[11, 13].

CT and MRI are excellent tools for characterizing and diagnosing GCTs. On CT, GCTs often appear as soft tissue density masses with destructive expansion into the bone, leaving the cortex intact. However, CT alone is insufficient for differentiating GCTs from other similar appearing masses, such as giant cell reparative granulomas and brown tumors. MRI provides better characterization and delineation of the tumor through its superior contrast resolution and different sequences ^[14].

The skull bones are reported to be more frequently involved than other parts of the face like the mandible or the maxilla. The difference between dealing with GCTs in other parts of the body, especially the long bones, and in the skull base is manifold. Firstly, the surgical anatomy of the skull base is extremely complicated and hence requires great expertise and

skill in the excision of these tumors. Secondly, total tumor removal is imperative in the first sitting because a recurrence in this complex area would be particularly difficult to treat. Thirdly, it is difficult to be radical in tumor removal of the skull base as resection or injury to vital neurovascular structures leaves an unacceptable degree of morbidity. Finally, the GCTs of the skull base are dealt with by the otology/skull base surgeons, ENT/head and neck surgeons or maxillofacial surgeons who bring with them their expertise and familiarity of the use of the microscope and the microdrill that facilitates precise tumor identification and removal ^[1].

In Yi Sben *et al*, (2016) study concluded that Craniomaxillofacial surgery for GCTs in the TMJ region invading the skull base is feasible in selected patients. A meticulous plan via a multidisciplinary approach is mandatory for the success of such treatment ^[2].

In Prasad *et al*, (2014) study concluded that GCTs are benign and locally aggressive and this makes surgical management difficult in the skull base due to the complex anatomy. A thorough knowledge of the anatomy of the skull base and the various skull base approaches is necessary to treat this subset of tumors ^[1].

Based on the results of our literature review of skull base GCTs, we found that total removal without adjuvant radiotherapy for the treatment of cranial GCTs is effective for local treatment of disease. Furthermore, it avoids the risks associated with radiation exposure, including radiation damage to normal surrounding tissues, and precludes the risk of sarcomatous degeneration. If total removal is not feasible, then subtotal removal plus radiotherapy appears to result in acceptable tumor control rates. For many years, chemotherapy had no role in the treatment of GCTs; however, denosumab, a receptor activator of nuclear factor kappa-b ligand (RANKL) inhibitor, is increasingly used in the treatment of GCTs with positive results.

5. Conclusions

GCTs of the skull commonly affect young adults, with an equal sex distribution, and are most often centered in temporal bone. Total removal is associated with the lowest recurrence rate and should be the goal of treatment. If total removal cannot be achieved, the combination of subtotal removal and radiation results in a similar recurrence rate.

6. References

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