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Article

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Guru Dutta Satyarthee[1], Luis Rafael Moscote-Salazar[2]
[1]INDIA, [2]COLOMBIA

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Primary giant myxoma of the temporal bone with major intracranial extension: presenting with hearing impairment and ear polyp

Guru Dutta Satyarthee[1], Luis Rafael Moscote-Salazar[2]

[1]Associate Professor, Department of neurosurgery, All India Institute of Medical Sciences, New Delhi, INDIA
[2]Neurosurgeon-Critical Care, RED LATINO, Latin American Trauma & Intensive Neuro-Care Organization, Bogota, COLOMBIA

Abstract: Myxomas are mesenchymal origin, benign tumor, constituting approximately half of the benign cardiac tumors. Occasionally, it may also occurs at other locations, though the intracranial location of a myxoma is considered exceptionally rare. Only isolated few cases of intracranial myxoma are reported in the literature, almost all were locally confined within the originating bone. The extensive Pubmed and Medline search yielded only eight cases of primary myxoma arising in the temporal bone with extension into intracranial compartment. However intracranial extension is limited as early detection, however, Osterdock et al reported a case also arising from temporal bone with extensive intracranial extension. Author report an interesting case of intracranial myxoma in 27- year- old- male, involving the temporal bone associated with extensive bony erosion and also extending into infratemporal fossa, mastoid, and frontoparietal region and a polypoidal mass protruding into external ear. To the best of knowledge of authors, temporal myxoma presenting with external ear polypoidal mass, which underwent successful surgical excision is not reported and represent first case in the world literature.

Key words: intracranial primary myxoma, temporal bone, surgery, outcome

Introduction

Primary intracranial myxoma is an extremely uncommon lesion, however, majority of reported cases represent metastatic intracranial myxoma. [1] It originates from mesenchymal tissue with predilection in the diminishing frequency i e. heart, skin, bone or genitalia. [2, 5-8] Primary myxoma is a benign but locally invasive tumour [2,7]. Surgical en-block excision of the lesion along with surrounding soft tissue is considered as the treatment of choice. [9-13] Incomplete excision may lead to early recurrence and further myxoma is not responsive to radiotherapy. Although radical resection is goal but always may not be possible especially in cases of intracranial

myxoma and further challenges due to gelatinous consistency, local invasiveness of lesion and nature of neoplasm.

Case illustration

A-27-year- old male presented with 5- year history of progressive hearing loss, protruding soft mass through right external acoustic meatus associated with sero-sanguinous discharge . He developed progressive bulging over the right temporal region for last four years, causing cosmetic facial distortion. He also developed slowly progressive worsening headache, which was not associated with vomiting, visual decline or double vision. He consulted otolaryngologist, who advised computed tomography scan of head with bone window view and coronal reconstruction and referred to our neurosurgery outpatient services. On examination, he was of average built and height. The swelling over right temporal and infratemporal region was hard and bony in consistency. A soft fleshy mass with sero-sanguinous discharge was protruding through external auditory meatus with intact surfaces except raw area at one place. His visual acuity was normal. He had no field defect on confrontation test. The pupils were normal with brisk light reflex. The optic fundi were normal. Right sided conductive deafness was also present. Other cranial nerves were unremarkable. Muscle strength of extremities was good and there were no pathological reflexes. The haematological and biochemical profiles were within normal limits.

The cranial computed tomography scan revealed a large heterogeneous calcified mass located in the right middle cranial fossa extending and invading into the petrous and squamus portion of right temporal bone, causing the erosion of middle cranial fossa base with extension into right infratemporal fossa causing swelling in the temporal and infratemporal fossa of, its size was 6x4.5x 5 cm. (Figure 1).

Large inhomogeneous mass lesion of size 6x4.5 x 5cm, with rim of calcification extending and invading the petrous and squamus portion of temporal bone, also causing the erosion of middle cranial fossa base with extension into infratemporal fossa.

A large hypodense area with calcified rim was noted that was extending into frontoparietal region and infratemporal region in the bone window view. (Figure 2) HRTCT of temporal bone also confirmed destruction of squamus and petrous temporal bone (Figure 3) along with middle and inner ear and associated extension into the Eustachian tube. (Figure 4)

Radiologically, the possible diagnosis of chondrosarcoma was entertained and possibility of other primary skull base tumour was also considered as differentials. He was planned for surgical management.

He underwent right-sided frontotemporal craniotomy. The tumour was causing remoulding and expansion of squamus and petrous temporal bone with erosion through outer table and extension into middle ear. It was primarily located extra-durally. It was firmly attached to dura. However, no dural infiltrated was observed during intraoperative period. It had variable consistency, areas of gelatinous consistency corresponding to hypodense areas in the CT scan were present. The rim was solid and more fibrous. Gelatinous part was relatively lesser vascular unlike solid part. Tumour was also eroded through middle fossa to extend into sub-

temporal region. Piecemeal dissection of tumour along with surrounding infiltrated muscle was carried out. External auditory meatus was closed after excision of polypoidal extension. The histopathological examination of resected specimen revealed presence of sparsely cellular tumour consisting of stellate to spindle shaped cells. These cells did not exhibit hyperchromasia, pleomorphism or mitotic activity. No mitosis or necrosis was observed. The bony specimen shows bony trabecullae, no evidence of tumor infiltration was observed. The tumour histopathology was interpreted as benign myxoma. In the post-operative period, cranial CT scan showed excision with subsidence of mass effect and no hematoma in the surgical resected cavity. He was discharged from the hospital on eighth post-operative day.

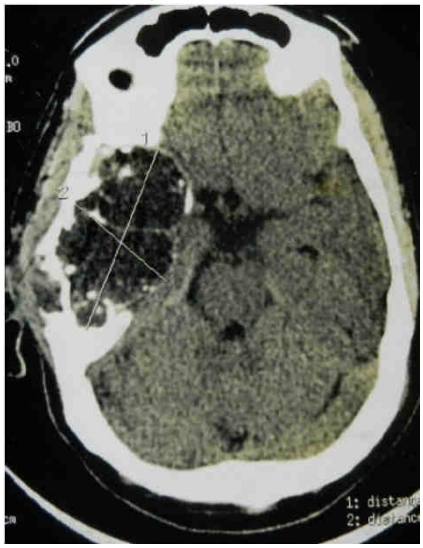

Figure 1 - Non-contrast enhanced computed tomography of head, axial section image showing large inhomogeneous mass lesion of size 6x4.5 x5cm, with rim of calcification extending and invading the petrous and squamus portion of temporal bone, also causing the erosion of middle cranial fossa base with extension into infratemporal fossa

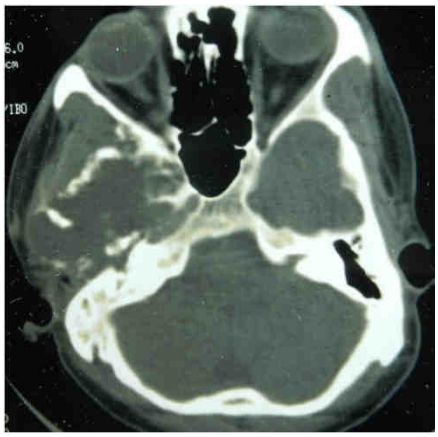

Figure 2 – Computed tomography head, bone window view image showing extra-large mass lesion with irregular rim of calcification causing erosion of middle cranial fossa and extending upto the right internal ear

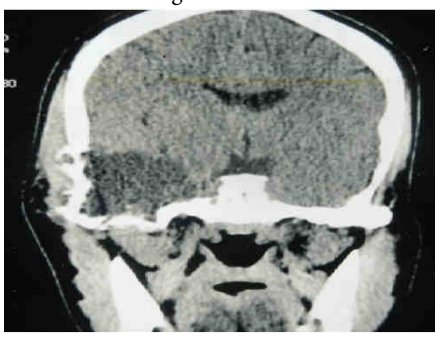

Figure 3 - Non-contrast enhanced computed tomography of head, coronal section image demonstrating large area of hypodense with calcified rim causing destruction of squamous part of the right temporal bone with extension into the infratemporal fossa

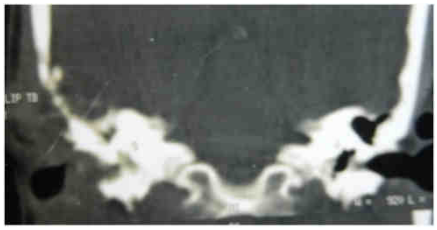

Figure 4 - Computed tomography head, bone window view, coronal section re-constructed image of 27-year male showing destruction of middle and inner ear with complete loss of mastoid air cells

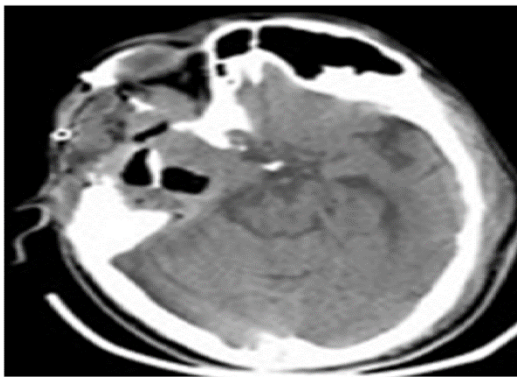

Figure 5 - Post-operative computed tomography scan of head showing excision of temporal myxoma

Discussion

A myxoma is a benign neoplasm, however, biologically it behaves like locally invasive tumour. Myxoma originates from tissues of mesenchymal origin. However, most of primary myxoma is commonly located in the heart, skin, genitalia and aponeurotic tissue. Although myxoma arising in bone, development begins in bone- marrow, subsequently causes expansion of dipole and ultimately expansion, destruction and remoulding of involved bone caused by aseptic pressure necrosis.

Primary myxoma of head and neck usually involve maxilla and mandible. [16-22] Primary myxoma involving skull base and vault is very rare. Primary intracerebral myxoma is not reported; even secondary intracranial myxoma is also uncommon. Intracerebral monastic myxoma, which usually originates as a result of metastatic deposit or remobilization from primary cardiac myxoma. Only few isolated case report of primary intracranial myxoma is reported. [1- 7, 11-13, 21, 23] The etiology of myxomas occurring in the head and neck region still remains unclear, although postulated about head and neck myxomas, these may arise from the tooth germ cells, because of presence in the maxillary and mandibular region. [22] Kleinsasser postulated primitive mesodermal tissue filling the middle ear space in the embryo and in the newborn may give rise to temporal bone myxomas. [17, 18, 23]

Myxoma can occur at any age and both sexes are at equal risk. Radiologically CT scan shows hypodense to isodense mass lesion with variation in pattern of enhancement following contrast administration. Even there is variation in degree of bony involvement, extensive bony involvement is considered extremely uncommon and our case was unique having huge intracranial extension, although only one such case was reported by Osterdock et al [3].

The management of myxoma is mainly surgical excision with wide margins of surrounding healthy soft tissue. Myxomas possess a strong tendency to recurrence, especially in the bones. [24] In cases of temporal bone myxomas, where healthy margins cannot be achieved, drilling and piecemeal removal as much as tissue are possible, can be seen as needs to be adequate. The radical surgery requirement must be evaluated in light of safety and preservation of facial nerve and inner ear function. Windfuhr and Schwerdtfeger [25] advocated en bloc excision with wide margins is not possible in view of ill-defined margins of tumor and complex anatomy of temporal bone in cases harbouring temporal bone myxomas. Therefore, it is wise to operate thoroughly but

without sacrificing vital structures. Regular follow-up is required, both clinically and radiologically, to detect recurrence early. The treatment of myxoma is surgery, although en bloc resection of intracranial myxoma is considered extremely difficult due to presence of important neurovascular structures difficult. [table 1] practically impossible to maintain its intactness during surgical dissection of these tumours, if cannot be excised with surplus surrounding tissue. But it is impossible in cranial skull-base and temporal bone [2, 4]. Due to associated danger of en-block resection, in relation to intracranial myxoma, piecemeal removal is preffered. Despite operative difficulties, the treatment of myxoma is surgical as myxoid tumours are generally insensitive to radiation therapy. Due to its locally invasiveness, complete surgical excision is difficult. Charabi et al reported left temporal myxoma which was involving mastoid, antrum and epitympenum but without any intracranial extension [2]. Klein et al reported a primary intracranial myxoma located in the posterior fossa without extensive bony involvement and intraoperatively tumour was relatively peeled from cerebellum easily [4]. However, in few cases myxoma grows beyond confines of dipole and extends into adjoining spaces. In these cases with significant intracranial extension and associated with extensive bone destruction, where attempt of radical excision may not be possible. Nagatani et al reported a case of primary myxoma of pituitary fossa, which had extensive bone erosion with spread into suprasellar cistern and inferiorly into sphenoid sinus after eroding sellar floor [16].

TABLE I

Published report of primary intracranial myxoma originating in the Temporal bone

S. No.	Series/ (Ref. no.)	Year	Site	Intracranial extension
1.	Richarth & Terrache [6]	1969	Temporal bone	Not mentioned
2.	Bulghov et al [7.]	1980	Temporal bone	Not mentioned
3.	Charabi et al. [2]	1989	Temporal bone	No
4.	Osterdock et al [3]	2001	Temporal bone	Extensive
5	Hsieh et al. [11]	2006	Temporal bone	small
6	Oruckaptan et al [1]	2010	Temporal bone	small
7	Sareen et al[21]	2010	Temporal bone external auditory canal, middle ear, mastoid antrum	nil
8	Zhang [15]	2006	Lateral skull	Details not available
	Current case	2016	Temporal bone	extensive

Osterdock et al [3] reported a 17- year -old male with left temporal bone invading petrous bone myxoma associated with a large intracranial extradural component containing central gelatinous part surrounded by hyperdense rim extending up to frontoparietal region and inferiorly up to posterior fossa. Our case also had extensive spread extending to frontoparietal region and posteriorly invading mastoid, middle ear, Eustachian tube and polyploidy like extension through middle year after perforating tympanic membrane into

external auditory canal. To best of our knowledge this is first case of primary intracranial myxoma with extensive bony erosion, which was also further extending to infratemporal fossa, mastoid, and frontoparietal region with mass protruding into external ear.

Recurrences are common, up to 25 % case may show recurrence if radical excision was not carried out [9]. The recurrence after surgery can occur as early as months and can delay up to 10 years.

Mueller et al reported a 12-year- girl, a case of surgically resected medulloblastoma, received adjuvant radiotherapy, presented with a mass lesion at the left transverse sinus during the follow-up [13]. Further added close relation to the radiation field of the posterior fossa medulloblastoma treatment, developed myxoma as a secondary radiation induced neoplasm.

Conclusion

The primary intracranial myxoma although care but important and its possibility of must be kept as one of differentials in all cases, who present with destructive skull - base lesions. The primary aim of the treatment is gross total excision, as en-block resection may not be always possible in cases with extensive intracranial extension. Further these cases need close monitoring with regular follow-up and screening cranial MRI to pick up early recurrences and provide tailored made appropriate treatment to each cases.

Correspondence
Guru Dutta Satyarthee, Associate Professor, Department of neurosurgery, All India Institute of Medical Sciences
Email: drguruduttaaims @gmail.com

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