Tumours of the jugular foramen are rare and much less common than the acoustic neuromas. Glomus jugulare is the commonest type of tumour at this site while other tumours include Schwannomas, meningiomas, epidermoids, metastatic carcinomas, chordomas and osteoclastomas. A case of ectopic glioma was reported in 1976. Neumannomas have a marked predilection for young females and occur most frequently on the left side, in a ratio of 9:1. Hakuba et al reviewed 42 previously reported cases of jugular foramen tumours including 12 personal cases of Pluchino and added 3 of their own. Kay et al reported 13 cases treated at the Cleveland Clinic Foundation, Ohio. We present 3 cases of jugular foramen tumours, treated at the Neurosurgery Department of Civil Hospital, Karachi, between 1988 and 1993.

Case Reports

Case 1
A 60 years old male presented with 2 years history of occipital headache, left earache without discharge, vertigo and poor balance. His examination revealed bilateral papilloedema, horizontal nystagmus, a complete sensorineural deafness on the left side, an ipsilateral LMN facial weakness and hemiparesis. Both his plantars were extensors. CT showed a high density lesion in the left jugular foramen with obstructive hydrocephalus, suggestive of a meningioma. He had total excision of the tumour through a posterior fossa craniectomy with temporary ventricular drainage. The tumour was histologically confirmed to be a Schwannoma. Patient had transient difficulty with sputum retention which resolved along with his other symptoms. The deafness, however, persisted.

Case 2
A 43 years old male complained of pain in the neck and right ear with deafness for 5 years. He had horizontal nystagmus, slurred speech, cerebellar ataxia, a right nerve deafness and an ipsilateral reduced corneal reflex. CT showed a tumour in the right jugular foramen. He had total tumour excision through a retromastoid craniectomy.
The tumour was histologically proven to be a neurinoma. Postoperatively the patient had lower cranial nerve palsies which took 2 months to recover. He had persistent deafness.

Case 3
A 35 years old female presented with 18 months history of progressive weakness of all the 4 limbs, suboccipital headache, swallowing difficulty and right sided deafness. On examination, she had spastic tetraparesis, LMN right facial palsy with cerebellar ataxia, coarse nystagmus, reduced corneal reflex on the right and ipsilateral sensorineural hearing loss. CT showed an enlarged jugular foramen on the right. MRI showed a large dumb-bell shaped tumour (Figure) in the posterior fossa, arising from the right jugular foramen with considerable medullary compression. Carotid angiography ruled out the
possibility of a glomus tumour. She had a retromastoid craniectomy and it was discovered that the
constriction in the middle of the tumour giving it a dumb-bell appearance was caused by an
overstretched spinal root of right accessory nerve. The neurinoma was totally excised and its histology
confirmed. She had a total recovery except for the deafness.

Discussion

Three cases of jugular foramen tumour are presented. Deafness was the commonest mode of
presentation and often antedated other symptoms such as headache and difficulty with swallowing,
phonation and balance. All 3 patients had sensorineural type of hearing loss on admission. In the review
of Hakuba et al\cite{3}. 36 out of 45 cases cited were deaf. Nine out of thirteen cases presented by Kaye et al\cite{5}
had deafness.

Only one of our patients had hydrocephalus with signs of raised intracranial pressure. Facial nerve
weakness of LMN type was present in two patients, which is an unusual pre-operative finding with an
acoustic nerve tumour. Hakuba et al\cite{3} found that almost half the patients they reviewed had facial nerve
involvement before surgery. All our cases had nystagmus, diminished corneal reflex and cerebellar
ataxia and two had long tract signs. All patients had a positive CT and only one patient had in addition
an MIRI and a carotid angiography. Total excision was achieved in all the three patients and there was
no mortality. Temporary difficulty with swallowing and sputum retention in the post-operative period
resolved. All patients recovered completely except for deafness which did not resolve. Over a followup
period of 3 months to 4-1/2 years there has been no recurrence. The differential diagnosis is between an
acoustic and a jugular foramen tumour. Unilaterally enlarged jugular foramen and a normal internal
auditory meatus on tomography in the presence of a jugular foramen syndrome is highly suggestive.
Wide anatomical variations exist in the size of jugular foramen, attributed to the size of lateral sinuses
as has been noted by Rhoton and Buza\cite{6} and by DiChiro et al\cite{7}. The right foramen is larger than the left.
CT is usually diagnostic although an MRI provides better resolution of the tumour. Complete excision
of these tumours is achieved through a suboccipital retromastoid craniectomy.

References

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