INTRODUCTION

Intracranial neurinomas account for 8% of all primary intracranial neoplasms. Lunardi P et al (1991) reported that in the cerebello pontine angle these are the most common lesions (59% of all SOL's and 80-90% of all tumours). These are predominantly solid tumours but cystic degeneration in these tumours can give rise to problem of differentiation. They reported that such small cystic changes in acoustic Schwanoma are not uncommon and have been seen in 9.6-20.5% of cases. Such cysts are small and contiguous with neurinoma and are easily identifiable after infusion of contrast. However, predominantly cystic tumour without any solid component on MRI are rare. Authors report two cases of cystic acoustic neuroma.

Case 1

A 50 years old female patient presented with 6 months history of progressive left sided hearing loss and complaints of facial asymmetry and unsteadiness while walking. There was no history of headache. Neurological examination showed absent corneal reflex on left side and 7th nerve paresis of the lower motor neuron type. There was sensorineural deafness and signs of cerebellar involvement on left side. Rest of the examination was normal. Pure tone audiometry revealed high pitched hearing loss on left side. MRI with contrast showed an extraaxial cystic lesion in left cerebellopontine angle with no evidence of solid component. There was no obstructive hydrocephalus. Retromastoid craniectomy exposed a large cyst which contained light yellow coloured fluid with a small solid component near the internal acoustic meatus. Postoperatively there was no neurological deterioration. Histopathological diagnosis was acoustic neuroma.

Case 2

A 40 years old male patient presented with history of right sided deafness and tinnitus of 4 months duration. Patient had unsteady gait and right sided involuntary movements for 3 months. He had difficulty in swallowing liquids for 3 days. On examination there was papilloedema, absent right sided corneal reflex, mild weakness of right 7th nerve, right sided hearing loss, and loss of right sided palatal and uveal reflex. There was dysmetria and
dysdiadochokinesia and ataxia suggestive of right cerebellar involvement. MRI showed a large cystic extraaxial tumor in right cerebello-pontine angle with obstructive hydrocephalus (Fig. I and II). A suboccipital crainectomy revealed a cystic tumor containing clear fluid with whitish granular solid component at internal acoustic meatus. The nodule was completely removed. Patient had postoperative 7th nerve palsy. Histopathological diagnosis was acoustic neuroma.

DISCUSSION

Acoustic neuroma with cystic changes are common but it is rare for them to be composed mainly of cysts. There have been reports of large cystic tumours with small solid components. Such tumours are characteristically, large and different from solid counterparts in clinical and radiological features.

Kameyama et al (1996) reported that they have short clinical history and atypical initial symptoms in form of facial pain, dysgeusia, early 7th nerve palsy, unsteadiness and vertigo. Hearing loss is less severe. There may be sudden headache and hearing loss because of intratumoral haemorrhage (Asano K et al, 1995). These tumours are large in size, and, in upto 50% of cases, despite of big size there is no widening of internal acoustic meatus (Kameyama et al, 1996). Tali ET et al (1993) reported that cysts in these tumours are usually single but may be multiple in 30% of the cases. There is circumferential enhancement after contrast administration and on MRI the intramural cysts commonly exhibit higher signal intensity than that of CSF (less commonly being isointense), on both T1 and T2 weighted images. This increased intensity is particularly because of necrotic material, blood and colloid rich fluid. The difference in the MR characteristic of these extramural / arachnoid cysts associated with acoustic schwannommas and those of typical arachnoid cysts not associated with neoplasia may be related to higher protein and/or colloid contents secreted by the tumour. Most extramural cysts have epicenters between the tumour and brain, suggesting that the most likely mechanism of formation is peritumoral adhesions. It creates a pseudoduplication caused by trapping of fluid between the leptomeninges and the mass, resulting in an acquired type of arachnoid cyst (Tali ET et al, 1993).

Kameyama et al (1996) reported that on the basis of CT and MR images these cystic acoustic neuromas can be divided into three types:

Type A: Large single cyst with thin tumourous wall.
Type B: Single cyst with thick tumourous wall.
Type C: Multicystic.

Differential diagnosis is from other cystic lesions of the CP angle i.e. arachnoid cysts and epidermoid cysts, less commonly cerebellar astrocytoma, metastasis, or hemangioblastoma with large cystic components and rarely a cystic medulloblastoma or meningioma, and partially thrombosed giant aneurysms of the basilar origin (Lunardi P et al, 1991).

These tumours have lobular growth pattern with high nuclear atypia and numerous macrophages. Lunardi P et al (1991) and Charabi S et al (1993) reported that all patients have a Antoni type-B tumour. Asano reported intratumoral haemmorrhage with niveau formations. Histologically these cases revealed a lots of abnormal sinusoid or telangiectasis vessels. Small haemorrhages, haemosiderin deposits and haemosiderin containing phagocytes are also more abundant as compared to solid tumours. On immunohistochemical study by Charabi S et al (1994) the cyst wall was found to contain numerous S-100 positive fibrils distributed throughout the whole extent of the wall. Laminin, fibronectin and vimentin are dominantly located around the vessel walls and factor VIII is found inside the vessels. GFAP positive lamellae occur in 50% of the cases near the surface of the cyst walls. The density of ki-67 positive cells is 36 times lower in non-cystic controls indicating that the increase in tumour size in cystic acoustic neuromas may be due to expansion of the cysts and not by an actual increase in the growth rate of the tumour cells.

Over all, the prognosis and outcome, though dependant on size of tumour, appears favourable (Lunardi et al). Aspiration of cyst facilitates recognition and preservation of facial nerve and removal of small solid component ensures good prognosis. In study by Charabi S et al (1994) at one year follow up, the outcome was found to be poorer as compared to the solid tumours of matching size.