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Multiple Cranial Nerve Palsies

A case of brain tumour

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HISTORY

A twenty two year old young businessman was admitted at Shaikh Zayed Hospital Lahore on 22-10-1986 with history of:–

Hoarseness of Voice	1 year
Morning headache	1 year
Heaviness right side tongue	1 year
Difficulty in swallowing	11 months
Regurgitation of food	11 months
Vertigo & unsteady gait	10 months
Facial weakness with inability to close right eye:	4 months
Deafness & ringing of ears	2 months
Double vision	1 month

Patient was alright one year ago when he developed hoarseness of voice which was gradual in onset and initially occured when he spoke lund, but later it became more pronounced and steady and lasted since then. He developed dull, morning headache which used to diminsih in intensity toward evening and increased on straining or coughing. Around the same time he developed a feeling of heaviness of right side of tongue. A month later he developed difficulty in swallowing and started regurgitation of food through nose although this was not a constant feature of his illness. He was admitted at another hospital and was discharged after a week with certain medication which did not improve his illness. After one month he was again hospitalized at a teaching hospital at Lahore and remained there for more than two months. During his stay he was subjected to two brain scans, a CT scan and a postnasal biospy in addition to routine studies but no diagnosis was made. After that he had treatment from many doctors for four months but in vain. By this time he developed right sided facial palsy and was put on steriods by a physician and brain scan was repeated but was reported normal. He received 30–60 mg of Prednisolone/day for a number of weeks and developed cushingoid features. Two months before admission to this hospital he developed deafness and ringing of right ear and a month ago developed double vision which, was marked on looking towards right. There was no history of significant vomiting, chronic ear discharge, convulsions. No history of streptomycin or salicylate use.

Past history, family history and personal history, were non contributory.

GENERAL PHYSICAL EXAMINATION

A young man of good physique, cushingoid face, staring right eye Temp: 98.6F^{O}, Pulse: 78/m, B.P. 135/90 mm Hg. Oedema, clubbing, goitre pallor and tremers were absent. There were no palpable lymph nodes.

Systemic Examination C.N.S.

Higher mental function intact

Cranial Nerves

Right 6th, 7th, 8th, 9th, 10th, 11th, 12th, affected. No sensory or motor deficit at the periphery. Reflexes and cerebellar functions intact. Fundi showed bilateral papilledema. Other systemic examination was within normal limit.

Diagnosis = Space Occupying Lesion Brain

INVESTIGATIONS

Blood Complete Picture

Normal count with Hb of 11.6 gm/dl and ESR of 5. mm Blood Sugar (R) 108 mg/dl

X-ray skull: on 8-10-1986 showed a mass lesion on the right side in basal view involving mid cranial fossa and possibilities of a jugular angle tumour or an acoustic neuroma were suggested.

Brain scan: done on dates 22.3.1986, 1.6.1986 and 22.7.1986 before the admission to this hospital were reported to be naormal.

C.C.F.

Was reported to be normal except proteins 69 mg % (N=20-45) cells 5/cmm lymphocytes.

C.T. Scan of brain at Sheikh Zayed Hospital on 11.10.86 shows a large solid space occupying lesion originating from base of brain over riding the jugular foramen and extending to the occipital area up to midline posteriorly indenting the posterior horn of lateral ventricle. The ventricles are moderately dilated. Tumour shows specks of calcification.

DISCUSSION

When one sees any patient like this who presents with multiple cranial nerve palsies without demonstrable sensory or motor deficit at the periphery one usually thinks of basal menigitis which typically may involve many cranial nerves, but in the absence of fever and neck rigidity one should be very careful in giving this diagnosis. Tumours usually don't care for anatomical boundaries and it is unusual for the tumour to involve the cranial nerves in series without producing other sensory or motor deficit.

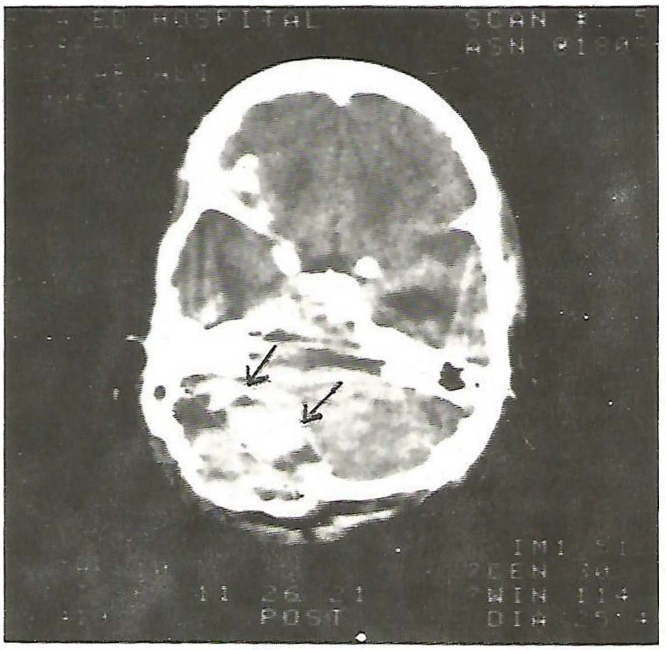
The interesting feature in this patient is how gradually the cranial nerves are involved in series and inspite of repeated brain scans and a CT scan the lesion remained undetected. Whether there was failure to recognise the lesion, on clinical grounds or in interpretation of brain scans, is hard to say.

When we saw the case frankly speaking we did not encounter any difficulty in making the diagnosis. In fact we picked up the lesion on plain radiographs of skull and it was confirmed on CT scan. The question whether the lesion has advanced recently is difficult to agree in view of history of patient. To evaluate the case we have got to know something about the behaviour of brain tumours and only then we can explain how this patient presented. For this I will briefly discuss the features of brain tumours and effects they produce by increase in intracranial pressure.

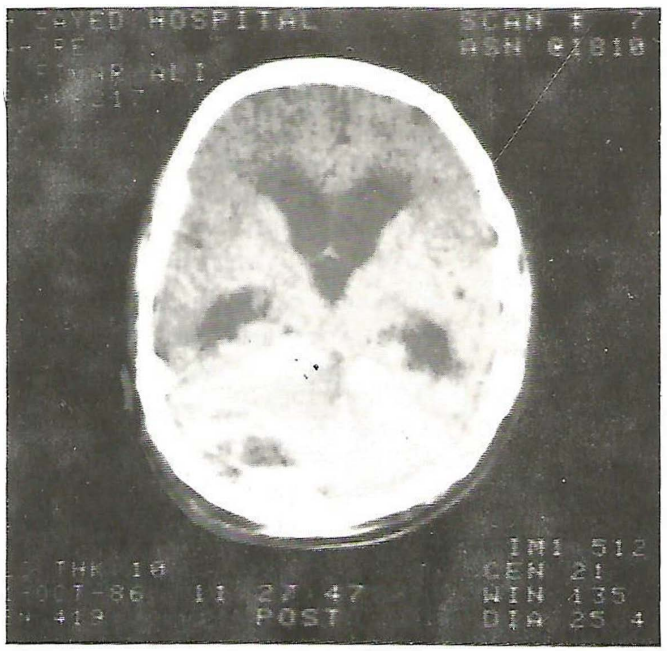
BRAIN TUMOURS

When we suspect that there is possibility of brain tumour in a clinical situation we must decide what is type of tissue, it is arising from

Meningioma	Glioma-40-50%
Neuroma	Astrocytoma
Vascular (Angioma)	Oligodendroglioma
Pituitary	Ependymoma
Pineal	Medulloblastomas



C.T. Scan with enhancement showing space occupying lesion in the posterior cranial fossa and right of mid line.



C.T. Scan without enhancement showing space occupying lesion in the posterior cranial fossa (marked with arrow). Mass is obstructing the 4th ventricle and causing dilatation of 3rd ventricle.

Sarcomas Neuroastrocytomas
Epidermoid Cyst
Secondary Tumour

II Compression of Mid Brain & Upper Pons.

Mechine like hyperventilation, moderately dilated pupils and sluggish to light, absent ciliospinal reflex and decreberate rigidity can develop.

III. Damage to Pons and Upper Medulla.

Quiet respiration, patient deeply comatosed and flaccid. Blood pressure rises, pulse falls.

IV. Medullary Damage

Respiration, irregular gasping slow, pupils widely dilated. Low BP leads to death.

V. Progressive Temporal Lobe or Uncal Herniation

Third nerve palsy present in stage II, III, IV of medullay dysfunction.

VI. Herniation of Cerbellar Tonsils

Severe headache, neck rigidity, tonsillar tilt if one tonsil herniates.

VII. Cerebellar Herniation Upward Over the Free Edge *of* of Tentorium.

Paralysis of upward gaze, stupor, brain stem compression.

VIII Compression of Arteries and Veins

Kinking and obstrucion of arteries can cause infraction of brain, remote from tumour.

IX. Erosion of Bone

X. Hyperostos

e.g. meningioma can stimulate osteoblastic activity.

Symptoms: Headache, vomitting, seizure, altered consciousness, mental changes, abnormal sensation

Incidence

Gliomas	40-50%
Astrocytomas Grade I	5-10
Astrocytomas Grade II	2-5
Astrocytomas Grade III & IV	20-30
Medulloblastoma 3-5	3-5
Qligodendroglioma 1-4	1-4
Ependymoma	1-3
Meningioma	12-20
Pituitary tumour	5-15
Neurolemmomas	3-10%
Metastatic	5-10
Blood vessel tumours (Endotheliomas)	05-1
A.V. malformation, Haemangioblastoma	
Tumours of developmental defects	2-3
(Dermoid, Epidermoid teratomas	
chrodomas Craniopharyngioma)	3-8
Pinealomas	5-8
Sarcomas, Papillomas of choroid plexus	1-3
Lipomas, unclassified etc.	1-3

Defects due to raised Intracranial Pressure:

Skull is a rigid structure and increase in intracranial contents produced by tumours, soon leads to an increase in intra-cranial pressure in addition to tumour mass itself. Infarction/haemorrhage produce increae in size of tumour. Oedema contributes to this process. Tumour may cause rapid rise in tension by obstructing the outflow of C.S.F.

I. Early Brains Stem Compression.

Impairment of consciousness, Cheyne stokes respiration, pupils are small, but responsive to light, bilateral signs of motor weakness, extensor plantar responses, increased tone in neck muscles, painful stimulation may produce decorticate posture.

Signs:- Papilledema, hydrocephalus, slow pulse, raised BP, false localising singns, respiratory changes, CSF rhinorrhea cranical nerve palsies, endorcrine changes.

Gliomas:

50% tumours of Brain and 25% of spinal cord are of glial origin.

Astrocytomas:

Most common and most frequent tumour causing aquiductal stenosis, hydrocephalus and early death. Astrocytoma of cerebellum is common in children and have low grade malignancy. In adults cerebellar astrocytomas are usually seen in childhood & adolesence. Pontine location is most common.

Ependymomas:

are second commonest after glioma. They are Infrequent as compared to astrocytomas.

Oligodendrogliomas:

Relatively uncommon and constitute less than 10% of gliomas. Usually found in 4th and 5th decades of life and predeominantly supratentorial with more than 50% occuring in frontal lobe.

Grow slowly and arise within white matter of CNS. As they grow larger they produce symptoms which are produced by compression, often sharply demarcated calcification is common which can be demonstrated in 70% cases on plain films of skull. As the tumours grow calcium deposition increases.

Medulloblastoma:

Most frequent tumour of posterior fossa in children. Accounts for less than 30% gliomas of children, almost exclusively seen in children. As tumour is soft, it may attain considerable size before producing symptoms. Spread through subarachnoid route is uncommon.

Neuroastrocytomas:

Usually occur in children and young adults most common site is medial side of temporal lobe or floor of 3rd ventricle, usually firm tumour, may contain areas of calcification.

Vascular Tumours

Capilary Telangiectasia:

A.V. Malformation

3% cause of S/A haemmorhage and epilepsy.

Venous Malformation

Hemangiolblastoma

Glomus Tumour

Red or blue mass, most pathologists believe that these tumours are derived from receptor nerve cells of ganglions nodosum of vagus nerve. Visible as pulsating mass through tympanic membrane. Extension of growth through petrous temporal bone casues 3rd-6th nerve palsy. Invasion of posterior fossa causes paralysis of lower cranial, nerves. Patiemt frequently complains of vascular murmur in ear.

Meningiomas:

These are second most common tumours arising both in brain and spinal cord and account for 17% tumours of CNS. Peak incidence is in 5th-6th decade. They arise from arachnoid cap cells in arachnoid villi.

Supratentorial meningiomas are most common in parasagital region. Other common locations are plague like tumours compressing the cenvexity of brain, sphenoid ridge parasellar region, basofrontal area. Most common site for infratentorial meningiomas is inferior aspect of petrous, temporal bone at cerebello pontine angle, arachnoid sheath at optic nerve.

Usually sphenoid or oval, some develop as a plate or sheet.

Neurilemmomas:

Commonest tumour of spinal cord (30%) and 3rd in frequency of intracranial tumours, rare in children, common in 4th decade or later.

Almost all are attached to 8th nerve. In other nerves, also associated with neurofibromatosis.

Sarcoma

Arise from connective tissue and bloos vessels of brain. Soft tumour

Developmental

Paraphyseal cyst
Chordoma
Dermoid Cysts
Epidermoid Cysts
Tumours of pineal gland.

Considering the site of tumour, multiple cranial nerve palsies and calcification, I think that it is either a meningioma or oligodendroglioma, Features which go against oligodendroglioma are that phyramidal tracts are spared and there is not other medullary dysfunction as this tumour arises from brain tissue. To grow so big it has to involve brain tissue. Meningioma is most likely. Next probability is Glomus tumour, but calcification is not a usual feature of glomus tumour.

DISCUSSION SUMMARY

DR. JAVAID AKHTAR *(Cons, E.N.T)* There are a couple of interesting features in this case. When E.N.T. consult was sought for this patient. He had an interesting feature of right blue ear drum and we considered that the patient has a glomus tumour and a jugluvenogram will be a helpful procedure to differentiate glomus tumour. The second possibility of blue ear drum which I don't consider in this case is high jugular bulb. Only thing that goes against it is calcification that Dr. Pervaiz has mentioned. Most of the time meningiomas do calcify. For a glomus tumour to get some calcification may have to be present for 10 years or so.

DR. IFTEKHAR *(Cons. Ophthal)* When the 7th nerve is involved, exposure keratitis is of concern, but there was no exposure keratitis in this patient. The right lateral rectus was paralysed. On looking at fundus I found bilateral papilledema. It is due to obstruction in the flow of exoplasm in the axons of optic nerve. To develop papilledema we must have a viable nerve and it can not develop in an atrophic nerve.

LT. GEN. M.A.Z. MOHYDIN *(Prof. Med.)* First of all I have got to appreciate that the case was very well presented and it was not a usual case which I as a physician has been seeing. If you see the chief complaints of the patient you will appreciate that symptoms developed gradually in an orderly fashion. If you see hoarseness in any patient you have to examine the larynx and can't call it psycho-

gical unless proven otherwise Similarly if a patient complains of morning headache which lasts for more than three months it is usually organic in nature. Same thing goes for heaviness of tounge as complaints of tounge are not too often. He is a patient with involvement of multiple nerves and seeing the bilateral papilledema, he is a sure case of space occupying lesion. The diagnosis should have been made much earlier if careful clinical examination was performed and not putting much reliance on scans. Now the question comes up whether it is a meningioma or a vascular tumour? I don't know whether any body has heard. any bruit on his nead. In a case like this with bilateral papilledema lumber puncture is contraindicated and that is why it was not done in this case.