RARE CASE OF INTRACRANIAL-EXTRACRANIAL MENINGIOMA PRESENTING AS CERVICAL MASS IN YOUNG PATIENT

Ayesha[1], Jaya Latha[2], G. Shaul Hameed[3], Durga Prasad[4], Uday[5]

[1]Assistant Professor, Department of Radiodiagnosis, MNJIO & RCC, Hyderabad.
[2]Professor, Department of Radiodiagnosis, MNJIO & RCC, Hyderabad.
[3]Assistant Professor, Department of ENT, SMC, Vijayawada.
[4]Assistant Professor, Department of ENT, SR Medical College, Nandyal.
[5]Resident, Department of Oncosurgery, MNJIO & RCC, Hyderabad.

ABSTRACT

INTRODUCTION

Intracranial meningiomas with extracranial extension are rare tumors with few cases reported in literature. We here with report a case of 15 year female patient presenting with a progressive swelling on right side of neck and proptosis of right eye. Suture lines in the base of skull and the optic canal could have provided a way for the extracranial and intraorbital spread respectively.

KEYWORDS

Meningiomas, Intracranial–Extracranial, Young Age.

HOW TO CITE THIS ARTICLE: Ayesha, Jaya Latha, G. Shaul Hameed, Durga Prasad, Uday. "Rare Case of Intracranial-Extracranial Meningioma Presenting As Cervical Mass In Young Patient". Journal of Evidence based Medicine and Healthcare; Volume 2, Issue 52, November 30, 2015; Page: 8708-8712, DOI: 10.18410/jebmh/2015/1210

INTRODUCTION: Meningiomas are one of the most common primary intracranial tumors, accounting for upto 30% of all intracranial tumors. They arise from arachnoid meningothelial (cap) cells. They are typically seen in middle aged-older patients. Meningiomas in children and young patients are uncommon. In this article, we present a rare case of intracranial-extracranial[1] (cervical and orbital) meningiomas, especially in young patient.

CASE REPORT: A 15 year old female patient presented with progressive swelling on right side of neck, proptosis of right eye, of 2 year duration. She had mild blurring and occasional pain in the eye. There were no CNS deficits or cranial nerve palsies.

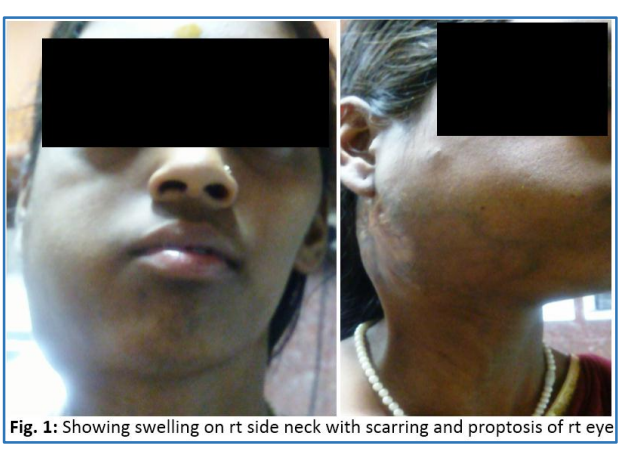


Fig. 1: Showing swelling on rt side neck with scarring and proptosis of rt eye

Local examination revealed a non tender, non- pulsatile hard mass on right side of neck[2] and overlying skin showed scarring due to previous H\O local burns

CECT of head and neck showed a large tumor with intracranial, cervical and intraorbital components. A large enhancing soft tissue mass (8x5.9x8.4cm) with amorphous calcifications was noted on right side of neck involving carotid, parapharyngeal spaces extending upto base of skull. It was encasing the distal CCA, ICA ECA and IJV. Extrinsic compression of the airway was noted. Tumor infiltration into sternomastoid was seen.[3] Enhancing intracranial mass[4] was seen in the right parasellar region extending posteriorly into the right parapontine and CP angle cisterns. Anteriorly it was seen in the presellar region. Enhancing fusiform shaped retro bulbar mass was seen encasing the optic nerve extending from the posterior aspect of the globe to the orbital apex with a small intracanalicular component.

Hyperostosis of the base of skull (greater wing of sphenoid, basisphenoid, clivus, petrous temporal[5,6] and occipital bone along the tumor) was noted.

Submission 23-11-2015, Peer Review 24-11-2015,
Acceptance 26-11-2015, Published 30-11-2015.
Corresponding Author:
Dr. Ayesha, H. No. 8-3-214/50 B,
Srinivasa Colony West, Ameerpet, Hyderabad.
E-mail: ayesharadio@gmail.com
DOI: 10.18410/jebmh/2015/1210

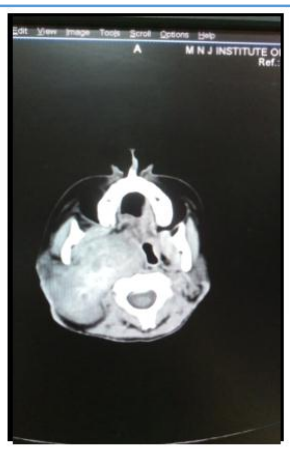


Fig. 2A: Plain CT Head & Neck showing Mass with calcification

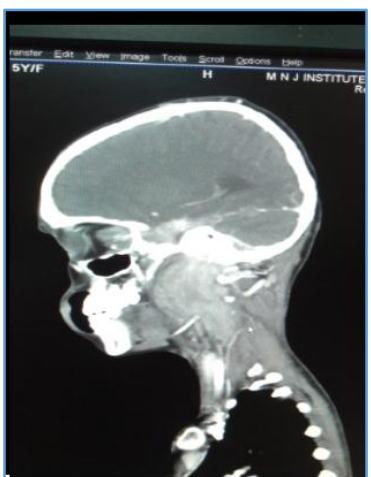


Fig. 2B: CECT Showing tumor extent

DISCUSSION: Intracranial–extracranial meningiomas are rare tumors.[7] Many theories have been postulated to explain the extracranial spread of tumor.[8,9] This may occur by direct extension of meningioma through a natural, traumatic or iatrogenic skull defect. Extension can be through the foramina, suture lines or there can be transdiploic extension.[10] A parasellar meningioma can extend into orbit through the superior orbital fissure or optic canal.

Meningiomas occurring ectopically in the neck are exceptional. Typical patients present with parotid[2] or parapharyngeal masses and cranial nerve palsies. Meningioma is not considered in differential diagnosis. The tumor is an extension of an unsuspected intracranial mass, occasionally with associated multiple meningiomas or other neurogenic tumors. Ectopic meningiomas should be considered in patients with parotid and parapharyngeal masses, particularly those with cranial nerve deficits; jugular foramen syndrome is most characteristic. Associated occult intracranial and temporal bone[10] tumors and the cranial form of neurofibromatosis should be suspected. Treatment of cervical meningiomas is excision. The most important aspect of intracranial-extracranial lesions is recognition and treatment of the intracranial portion.

Extracranial/ectopic/extraduralmeningiomas. [11,9,12] are rare subsets that arise outside dural covering of the brain and spinal cord and have no connection with dura or other intracranial structure.Lang et al[13] classified extracranial mengiomas as:

Type 1: purely extracalvarial.
Type 2: purely calvarial:
Type 3: calvarial and extracalvarial.

Intracranial meningiomas with extracranial extension are rare lesions with only few reported cases in literature.[8,1]

Louis DN, Ohgaki H, Wiestler OD, Cavenee WK. World Health Organization classification of tumors of the central nervous system[11,14,15]

The radiographic features are similar to intracranial meningiomas.

In our case, the tumor was an intracranial meningioma (parasellar with extension into posterior fossa) with extracranial spread into base of skull/neck through the suture lines and intraorbital spread[1,7,16] (through the optic canal).

Both CT and MRI can show the extent of the lesion. FNAC, biopsy of the lesion confirmed the diagnosis of a meningioma. Whenever possible, a complete excision of tumor is the treatment of choice with upto 80% chance of local recurrence.

Histopathalogically, the tumor showed sheets and islands of meningothelial cells, -round to oval with hyperchromatic nucleus and moderate amount of cytoplasm, few psammoma bodies also seen–s/o meningioma–psammomatous type grade 1.

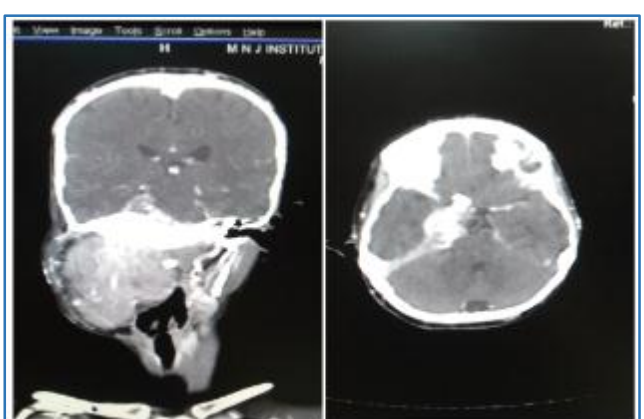


Fig. 3A, 3B: CECT showing extension of tumor

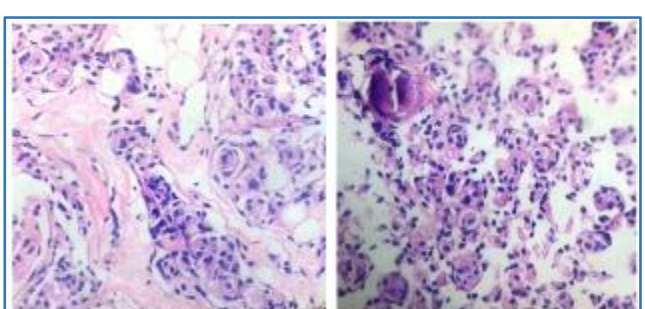


Fig. 4: Histopathology images s/o meningioma–Psammomatous type grade 1

TUMOURS OF NEUROEPITHELIAL TISSUE

Astrocytic tumours	
Pilocytic astrocytoma	9421/1[1]
Pilomyxoid astrocytoma	*9425/3**
Subependymal giant cell astrocytoma	9384/1
Pleomorphic xanthoastrocytoma	9424/3
Diffuse astrocytoma	9400/3
Fibrillary astrocytoma	9420/3
Gemistocytic astrocytoma	9411/3
Protoplasmic astrocytoma	9410/3
Anaplastic astrocytoma	9401/3
Glioblastoma	9440/3
Giant cell glioblastoma	9441/3
Gliosarcoma	9442/3
Gliomatosis cerebri	9381/3
Oligodendroglial tumours	
Oligodendroglioma	9450/3
Anaplastic oligodendroglioma	9451/3
Oligoastrocytic tumours	
Oligoastrocytoma	9382/3
Anaplastic oligoastrocytoma	9382/3
Ependymal tumours	
Subependymoma	9383/1
Myxopapillary ependymoma	9394/1
Ependymoma	9391/3
Cellular	9391/3
Papillary	9393/3
Clear cell	9391/3
Tanycytic	9391/3
Anaplastic ependymoma	9392/3
Choroid plexus tumours	
Choroid plexus papilloma	9390/0
Atypical choroid plexus papilloma	*9390/1**
Choroid plexus carcinoma	9390/3
Other neuroepithelial tumours	
Astroblastoma	9430/3
Chordoid glioma of the third ventricle	9444/1
Angiocentric glioma	*9431/1**
Neuronal and mixed neuronal-glial tumours	
Dysplastic gangliocytoma of cerebellum (Lhermitte-Duclos)	9493/0
Desmoplastic infantile astrocytoma/ ganglioglioma	9412/1
Dysembryoplastic neuroepithelial tumour	9413/0
Gangliocytoma	9492/0
Ganglioglioma	9505/1
Anaplastic ganglioglioma	9505/3
Central neurocytoma	9506/1
Extraventricular neurocytoma	*9506/1**
Cerebellar liponeurocytoma	*9506/1**
Papillary glioneuronal tumour	*9509/1**
Rosette-forming glioneuronal tumour of the fourth ventricle	*9509/1**
Paraganglioma	8680/1
Tumours of the pineal region	
Pineocytoma	9361/1
Pineal parenchymal tumour of intermediate differentiation	9362/3
Pineoblastoma	9362/3
Papillary tumour of the pineal region	*9395/3**
Embryonal tumours	
Medulloblastoma	9470/3
Desmoplastic/nodular medulloblastoma	9471/3
Medulloblastoma with extensive nodularity	*9471/3**
Anaplastic medulloblastoma	*9474/3**
Large cell medulloblastoma	9474/3
CNS primitive neuroectodermal tumour	9473/3
CNS Neuroblastoma	9500/3
CNS Ganglioneuroblastoma	9490/3
Medulloepithelioma	9501/3
Ependymoblastoma	9392/3
Atypical teratoid / rhabdoid tumour	9508/3

TUMOURS OF CRANIAL AND PARASPINAL NERVES

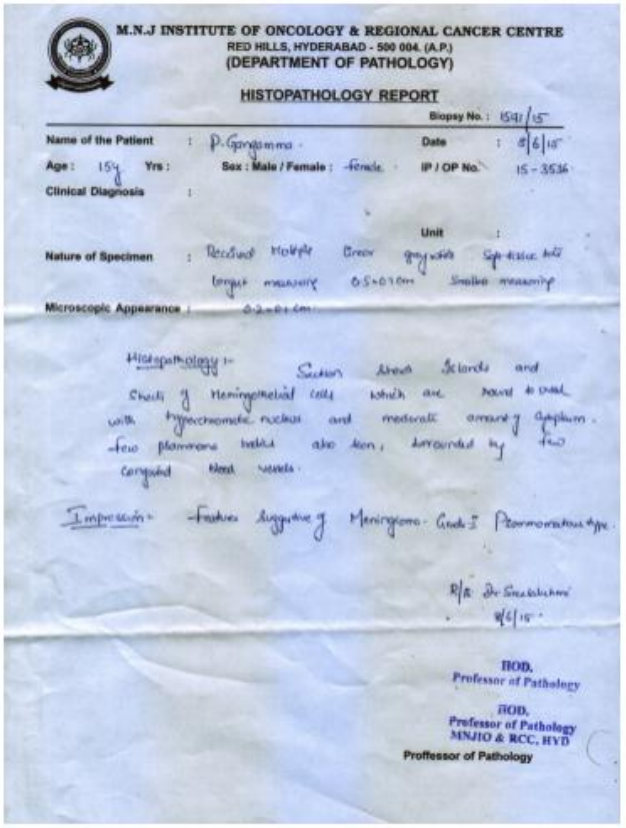
Schwannoma (neurilemoma, neurinoma)	9560/0
Cellular	9560/0
Plexiform	9560/0
Melanotic	9560/0
Neurofibroma	9540/0
Plexiform	9550/0

[1] Morphology code of the International Classification of Diseases for Oncology (ICD-O) (WHO) and the Systematized Nomenclature of Medicine (http://snomed.org). Behaviour is coded /0 for benign tumours, /3 for malignant tumours and /1 for borderline or uncertain behaviour.

* The italicised numbers are provisional codes proposed for the 4th edition of ICD-O. While they are expected to be incorporated into the next ICD-O edition, they currently remain subject to change.

Perineurioma	
Perineurioma, NOS	9571/0
Malignant perineurioma	9571/3
Malignant peripheral nerve sheath tumour (MPNST)	
Epithelioid MPNST	9540/3
MPNST with mesenchymal differentiation	9540/3
Melanotic MPNST	9540/3
MPNST with glandular differentiation	9540/3
TUMOURS OF THE MENINGES	
Tumours of meningothelial cells	
Meningioma	9530/0
Meningothelial	9531/0
Fibrous (fibroblastic)	9532/0
Transitional (mixed)	9537/0
Psammomatous	9533/0
Angiomatous	9534/0
Microcystic	9530/0
Secretory	9530/0
Lymphoplasmacyte-rich	9530/0
Metaplastic	9530/0
Chordoid	9538/1
Clear cell	9538/1
Atypical	9539/1
Papillary	9538/3
Rhabdoid	9538/3
Anaplastic (malignant)	9530/3
Mesenchymal tumours	
Lipoma	8850/0
Angiolipoma	8861/0
Hibernoma	8880/0
Liposarcoma	8850/3
Solitary fibrous tumour	8815/0
Fibrosarcoma	8810/3
Malignant fibrous histiocytoma	8830/3
Leiomyoma	8890/0
Leiomyosarcoma	8890/3
Rhabdomyoma	8900/0
Rhabdomyosarcoma	8900/3
Chondroma	9220/0
Chondrosarcoma	9220/3
Osteoma	9180/0
Osteosarcoma	9180/3
Osteochondroma	9210/0
Haemangioma	9120/0
Epithelioid haemangioendothelioma	9133/1
Haemangiopericytoma	9150/1
Anaplastic haemangiopericytoma	9150/3
Angiosarcoma	9120/3
Kaposi sarcoma	9140/3
Ewing sarcoma - PNET	9364/3
Primary melanocytic lesions	
Diffuse melanocytosis	8728/0
Melanocytoma	8728/1
Malignant melanoma	8720/3
Meningeal melanomatosis	8728/3
Other neoplasms related to the meninges	
Haemangioblastoma	9161/1
LYMPHOMAS AND HAEMATOPOIETIC NEOPLASMS	
Malignant lymphomas	9590/3
Plasmacytoma	9731/3
Granulocytic sarcoma	9930/3
GERM CELL TUMOURS	
Germinoma	9064/3
Embryonal carcinoma	9070/3
Yolk sac tumour	9071/3
Choriocarcinoma	9100/3
Teratoma	9080/1
Mature	9080/0
Immature	9080/3
Teratoma with malignant transformation	9084/3
Mixed germ cell tumour	9085/3
TUMOURS OF THE SELLAR REGION	
Craniopharyngioma	9350/1
Adamantinomatous	9351/1
Papillary	9352/1
Granular cell tumour	9582/0
Pituicytoma	*9432/1**
Spindle cell oncocytoma of the adenohypophysis	*8291/0**
METASTATIC TUMOURS	

Table 1: The 2007 WHO Classification of Tumours of the Central Nervous System


M.N.J INSTITUTE OF ONCOLOGY & REGIONAL CANCER CENTRE
RED HILLS, HYDERABAD - 500 004. (A.P.)
(DEPARTMENT OF PATHOLOGY)

HISTOPATHOLOGY REPORT

Biopsy No. : 1541/15

Name of the Patient : P. Goraamma Date : 9/6/15

Age : 15y Yrs : Sex : Male / Female : Female IP / OP No. 15-3536

Clinical Diagnosis :

Unit :

Nature of Specimen : Received Multiple Grey greyish Soft tissue bits longest measuring 0.5×0.1cm Smallest measuring

Microscopic Appearance :

Histopathology :- Sections showed Islands and Sheets of Meningothelial cells which are found to oval with hyperchromatic nuclei and moderate amount of eosinophilic few psammoma bodies also seen, surrounded by few congested blood vessels.

Impression :- Features suggestive of Meningioma - Grade I Psammomatous type.

D/R Dr. Sreelakshmi 9/6/15

HOD, Professor of Pathology

HOD, Professor of Pathology MNJIO & RCC, HYD

Professor of Pathology

Fig. 5: Pathology report

CONCLUSION: We present a rare case of intracranial meningioma in a young lady with extracranial and intraorbital extension. CECT showed the extent of the tumor with hyperostosis and Histopathological examination confirmed the diagnosis.

REFERENCES:

1. Cech DA, Leavens ME, Larson DL. Giant intracranial and extracranial meningioma: case report and review of the literature. Neurosurgery 1982; 11: 694-697.
2. Nichols RD, Knighton RS, Chason JL, Strong DD. Meningioma in the parotid region. Laryngoscope. 1987; 97: 693–696. doi: 10.1288/00005537-198706000-00008. [Pub Med] [Cross Ref].
3. Galm T, Macgregor F, Sinclair AM, Hunter J, McPhaden A. Extracranial meningioma presenting as a neck mass in a patient with underlying ankylosing spondylitis. J Rheumatol. 2006; 33: 1883–1885. [Pub Med].
4. Gilbert ME, Shelton C, McDonald A, Salzman KL, Harnsberger HR, Sharma PK, et al. Intraventricular meninigiomas: a report of 25 cases. Neurosurg Rev. 2006; 29:36–40. doi: 10.1007/s10143-005-0418-1. [Pub Med] [Cross Ref].
5. Agrawal A, Rao KS, Makannavar JH, Shetty L, Patel N. Extracranial meningioma in the vicinity of the temporal bone: a difficult preoperative diagnosis. Surg Neurol. 2007; 67: 102–105. doi: 10.1016/j.surneu.2006.02.043. [Pub Med] [Cross Ref].
6. Ferlito A, Devaney KO, Rinaldo A. Primary extracranial meningioma in the vicinity of the temporal bone: a benign lesion which is rarely recognized clinically. Acta Otolaryngol. 2004; 124: 5–7. doi: 10.1080/00016480310014903. [Pub Med] [Cross Ref].
7. Friedman CD, Costantino PD, Teitelbaum B, Berktold RE, Sisson GA., Sr Primary extracranial meningiomas of the head and neck. Laryngoscope. 1990;100:41–48. [Pub Med] 7. Nakagawa H, Lusins JO. Biplane computed tomography of intracranial meningiomas with extracranial extension. Journal of computer assisted Tomography 1980; 4: 478-483.
8. Neeff M, Baysal E, Homer J, Gillespie J, Ramsden R. Intracranial/Extracranial meningioma arising in the hypoglossal canal: case report. Skull base: official Journal of North American Skull Base Society [et al] 2007; 17: 325-330.
9. Batsakis JG. Pathology consultation. Extracranial meningiomas. Ann Otol Rhinol Laryngol. 1984;93:282–283. [Pub Med].
10. Chang CY, Cheung SW, Jackler RK. Meningiomas presenting in the temporal bone: the pathways of spread from an intracranial site of origin. Otolaryngol Head Neck Surg. 1998; 119: 658–664. doi: 10.1016/S0194-5998(98)70030-0. [Pub Med] [Cross Ref].
11. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK. World Health Organization classification of tumors of the central nervous system. 4. Lyon: IARC Press; 2007.
12. Friedman CD, Costantino PD, Teitelbaum B, Berktold RE, Sisson GA. Primary extracranial meningiomas of the head and neck. The Laryngoscope 1990; 100: 41-48.
13. Primary extradural meningiomas: a report on nine cases and review of the CT-era literature Frederick F. Lang, M.D., Department of Neurosurgery, M. D. Anderson Cancer Center, The University of Texas, Houston, Texas.
14. Longstreth WT, Dennis LK, McGuire VM, Drangsholt MT, Koepsell TD. Epidemiology of intracranial meningioma. Cancer 1993; 72: 639-648.
15. Louis D, Ohgaki H, Wiestler O, Cavenee W, Fuller C. World Health Organization classification of tumours of the central nervous system. JOURNAL OF NEUROPATHOLOGY AND EXPERIMENTAL NEUROLOGY 2008; 67: 260.
16. Akif M. Intra-Extracranial Meningioma. Turkish Neurosurgery 1993; 4: 170-172.