PRIMARY EXTRACRANIAL MENINGIOMA OF THE SPHENOID SINUS: A HIDDEN MENACE

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Abstract:
Extracranial meningioma are infrequent tumours. Here we are discussing a case of 40 years old male patient who came with chief complain of gradually increasing nasal obstruction for 5 years. Right sided facial swelling, anosmia and post nasal drip for 2 years. MRI and CT scan showed soft tissue mass occupying sphenoid sinus, maxilla and infratemporal fossa. Preoprative biopsy was consistent with meningioma. Mass was removed via combine (external and endoscopic) approach. On follow up 6 month postoperatively, patient was disease free.

Key Words: Meningioma, Extracranial, Sphenoid sinus

INTRODUCTION:

Tumors of the central nervous system (CNS) are infrequent, but meningioma is the one tumor which is encountered most frequently. Arising from the arachnoid cells of the meninges, they make upto 13-18% of all intracranial tumors.[1] Ectopic meningiomas have been found at various anatomic sites. In the head and neck, they have been reported in the floor of the mouth, nose and paranasal sinuses, as well as in the lungs, retro-peritoneum and thigh.[2] It is believed that the arachnoid cells along the peripheral nerves are responsible for the origin in these cases.[3] In the following section, we describe the hospital course of a patient who presented with an extracranial meningioma involving the sphenoid sinus.

CASE REPORT:

A 40 years old male presented in our out-patient department, with a history of persistent nasal obstruction for a period of 5 years, right sided facial swelling, gradually increasing in size for the past 2 years. He, further on, complained of anosmia and post-nasal drip for the past 2 years. A previous biopsy performed 2 years back was inconclusive. On clinical examination, the individual had no lesion or mass visible on anterior rhinoscopy or on a rigid nasal endoscopic examination. There was right sided non-tender facial swelling. Vision and extra-ocular movements were intact. Rest of the ear, nose and throat examination was un-remarkable. He had a magnetic resonance image performed of his paranasal sinuses which showed a mass involving the sphenoid sinus (Fig 1). The patient underwent an endoscopic biopsy from the sphenoid sinus under general anesthesia. Frozen section favored a diagnosis of neoplasm and the final histopathology came out to be Grade I meningioma. The patient was advised surgery in collaboration with neurosurgery. He was lost to follow up for 6 months. After 6 months he returned with increase in facial swelling and a computed tomographic scan showed extension of the disease in the maxilla as well as in infra-temporal fossa region. He underwent surgical excision via a Weber-Fergusson approach. Intra-operatively right total maxillectomy was performed, along with removed of the tissue from the right infra-temporal fossa. Sphenoid disease was removed using rigid nasal endoscopes. The residual disease in and around the sinus was removed under microscope to ensure adequate and safe excision. The whole cavity was then packed with a BIPP (Bismuth iodoform paraffin paste) soaked ribbon gauze. Post-operatively vision was intact. He remained vitally stable, the BIPP pack was removed 48hours after the surgery and patient was discharged the next day. On his follow up a week later, he was stable and 2 months down the road, an MRI scan of the paranasal sinuses was carried out, which did showed only mild soft tissue thickening (Fig II). On his recent follow up 6 months later, patient is free of disease.

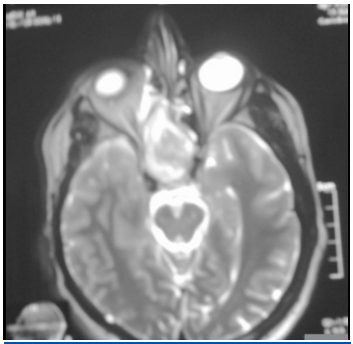

Fig. I: Disease involving primarily the sphenoid sinus

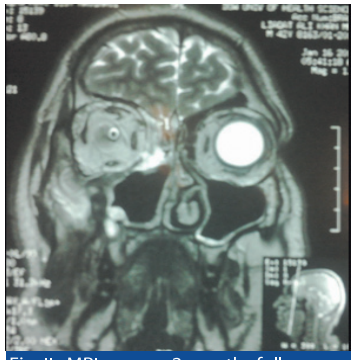

Fig. II : MRI scan on 2 months follow up with no evidence of disease

DISCUSSION:

Meningiomas are benign tumors representing 13 to 18% of all primary intracranial neoplasia.[1] Primary extracranial meningiomas are histologically identical to intracranial meningiomas. The aetio-pathogenesis is considered to be due to the migration of arachnoid cells, derived from the neural crest cells. Apart from that different mechanisms have also been proposed such as; originating from arachnoid cells of nerve sheaths emerging from skull foramina, from pacchionian bodies possibly displaced or entrapped in an extracranial location during embryologic development, by trauma or cerebral hypertension displacing arachnoid islets or,deriving from undifferentiated mesenchymal cells.[4]

They usually occur in 40 to 60-year-old patients and are rare in the pediatric age group. Primary extracranial meningiomas represent 1 to 2% of all meningiomas.[5] Meningioma of the nose and paranasal sinuses may occur as a secondary extension of a primary tumor in the cranial cavity or primarily in the nose and paranasal sinuses de novo.

The symptoms for involvement of paranasal sinuses are non-specific and are similar to the symptoms of sinusitis and the involvement of adjacent structures. The most common complaints are nasal discharge, nasal mass and epistaxis. Nasal obstruction, anosmia, headaches and proptosis are also frequent complaints.[6] This creates a problem to diagnose based on clinical examination. The differential diagnosis would then include epithelial neoplasms (carcinoma), melanoma, olfactory neuroblastoma and nasopharyngeal angiofibroma.[7]

The roentgenographic findings are usually nonspecific and include clouding or opacification of the sinuses, bony sclerosis, and focal destruction of the surrounding sinusoidal or nasal cavity bony tissues.[8] Despite that, computed tomography helps in demonstrating intra-tumoral calcification and homogenous contrast enhancement. Magnetic resonance imaging compliments the findings on CT scan imaging by providing relevant information regarding involvement of surrounding structures and invasion into the orbit or intracranially.[9] For follow up imaging, either modality can be adopted. On biopsies taken, histologic features can easily help in differentiating these entities from other lesions common in the paranasal sinuses. Where queries regarding differentiating between carcinomas and melanomas arise, immune-histochemical stains prove beneficial.

Owing to the proclivity for local permeation through suture lines and skull foramina, it is easy for meningiomas to spread from one sinus to another as well as into the brain. It is still worth mentioning that clinical and radiological features cannot predict the nature of these lesions.

Treatment of primary extracranial meningioma is via a surgical excision as meningiomas are known to be radio resistant. Recurrence is very rare following adequate surgical resection.[10] Most meningiomas are benign with no tendency to metastasize and malignant change is rare. In general, the prognosis of primary meningioma of the sinonasal tract appears to be excellent.[11] Recurrence of disease is usually at the same anatomical site as the site previously operated for the disease. This, therefore, denotes the presence of residual disease rather than recurrence of the same. Studies have reported little difference in the 5 year and 10year disease free survivals. Rushing et al reported the 5 year and 10 year disease free survivals to be 91.2% and 90.1% respectively, thus indicating that once the patients survive disease free for 5 years, they are unlikely to die secondary to it.[12]

CONCLUSION:

Primary extracranial meningioma originating from the sphenoid sinus is a rare entity which requires an adequate diagnosis. This relies almost completely on tissue biopsy. Once identified, complete surgical excision is the curative treatment with a good prognosis and an acceptable disease free survival. It is, therefore, essential to have basic knowledge regarding this pathology.

REFERENCES:

1. Bayar MA, Iplikçioglu C, Kökes F, Gökçek C. Intra-Extracranial Meningioma. Turkish Neurosurgery 1993; 4: 170 – 172
2. Gökduman CA, Iplikcioglu AC, Kuzdere M, Bek S, Cosar M. Primary meningioma of the paranasal sinus. J Clin Neurosci 2005; 12: 832-834
3. Deshmukh SD, Rokade VV, Pathak GS, Nemade SV, Ashturkar AV. Primary extra-cranial meningioma in the rightsubmandibular region of an 18-year-old woman:a case report. Journal of Medical Case Reports 2011; 5:271
4. Serry P, Rombaux PH, Ledeghen S, et al. Extracranial sinonasal tract meningioma: a case report. Acta Otorhinolaryngol Bel 2004;58: 151-5
5. Kainuma K, Takumi Y, Uehara T, Usami S. Meningioma of the paranasal sinus: a case report. Auris Nasus Larynx2007; 34: 397-400
6. Thompson LDR, Gyure KA. Extracranial sinonasal tract meningiomas. Am J Surg Pathol 2000; 24: 640-650
7. Haber DM, Maniglia MP, Schiavetto RR, Molina FD et al. Primary Meningioma of the Paranasal Sinuses: Case Report. Intl. Arch. Otorhinolaryngol, 2007;11: 70-73
8. Moulin G, Coatrieux A, Gillot JC, et al. Plaque-like meningioma involving the temporal bone, sinonasal cavities and both parapharyngeal spaces: CT and MRI. Neuroradiology 1994; 36: 629–31
9. Filho JR, Floriano VH, Felipe LF et al. Clinical-Radiological Aspects Of Primary Extracranial Meningioma Of The Ethmoid Sinus In A Child. Arq Neuropsiquiatr 2008; 66:274-275
10. Mair R, Morris K, Scott I, Carroll TA. Radiotherapy for atypical meningiomas. Neurosurg 2011; 115: 811-819
11. Marzin FA, Carmona GL, Florez MA, Garcia PF. Extracranial meningioma of the paranasal sinuses. Acta Otorrinolaringol Esp. 2010;61:238-240
12. Rushing EJ, Bouffard JP etal. Primary Extracranial Meningiomas: An Analysis of 146 Cases. Head and Neck Pathol2009; 3 :116–130

