Primary Extracranial Meningioma: The Royal Pearl Experience

Trichy Narayanan Janakiram¹ · Palak Parekh¹ · Vijayalakshmi Subramaniam^{2,3}

Received: 25 March 2015 / Accepted: 27 April 2015 / Published online: 5 May 2015
© Association of Otolaryngologists of India 2015

Abstract The extracranial occurrence of meningioma, which is a tumour of the central nervous system, is rare. Two cases of primary extracranial meningioma of the psammomatous variant are reported here. Both cases were managed successfully by endoscopic endonasal approach. Literature has been reviewed and their clinicopathological features are described.

Keywords Extracranial meningioma · Psammoma bodies · Epithelial membrane antigen

Introduction

Meningiomas are benign tumours which originate from the arachnoid cap cells. They constitute 24–30 % of all intracranial neoplasms and have an incidence of 1.5–4.5/100,000 [1]. Extracranial meningiomas account for 1–2 % of these tumours [2, 3]. Histologically, the psammomatous subtype is seen only in 2 % of meningiomas [4]. Two cases of psammomatous extracranial meningioma which were successfully managed by endoscopic endonasal approach are reported.

Case Reports

Case: 1

A 40 year old male was referred to the Department of Otorhinolaryngology with complaints of bilateral nasal obstruction and recurrent headache of 4 years duration. He also gave a history of intermittent epistaxis and diminished vision on the right side. He gave a history of 3 prior surgeries for the same complaints and external rhinotomy had been performed once. Examination showed a huge reddish mass with smooth surface occupying the whole of the nasal cavity. Gadolinium enhanced magnetic resonance imaging (MRI) with fat suppression showed heterogeneously enhancing lesion centred into the cribriform plate with extradural extension into the base of the brain in T1 weighted images (Fig. 1). The patient was posted for endoscopic excision of the mass lesion. Under General anesthesia (GA), patient was put in reverse Trendelenberg position with head in neutral position slightly tilted towards right. A 0°, 4 mm, 18 cm wide angled telescope (Karl Storz) was used and reddish multilobulated mass was visualized. It had a firm capsule and bled on touch. Needle was introduced and yellowish fluid aspirated (Fig. 2). Using a microdebrider, the mass was debulked. Biopsy was taken from various parts of the mass. A bony shell was found adherent to the dura adjoining the sellar floor, tuberculum sellae, optic canal, planum sphenoidale, fovea ethmoidalis and cribriform plate. Using Cottle's elevator, gentle dissection was carried out between the bony layer and dura. Care was taken not to breach the dura at any point. Bony shell over optic nerve on both sides was removed and optic canal was found to be dehiscant. Dissection was then carried out anteriorly over the region of the fovea and cribriform plate. While removing the bony shell abutting cribriform plate, leakage of cerebrospinal fluid

✉ Vijayalakshmi Subramaniam
vijisubbu@gmail.com

¹ Department of Otorhinolaryngology, Royal Pearl Hospital, Tiruchirappalli, Tamilnadu, India

² Department of ENT-Head and Neck Surgery, Yenepoya Medical College, Deralakatte, Mangalore 575018, Karnataka, India

³ Department of Otorhinolaryngology, Yenepoya Medical College, Yenepoya University Campus, Deralakatte, Mangalore 575018, Karnataka, India

Fig. 1 Contrast MRI with fat suppression showing heterogeneously enhancing lesion centred into the cribriform plate with extradural extension into the base of the brain in T1 weighted images

Fig. 2 Endoscopic image showing multilobulated mass with yellowish fluid draining on aspiration

(CSF) was encountered on the right side. It was repaired using dumb bell fat technique and overlay fascia lata graft. Nose was packed with nasopore, surgicel and Foley's catheter as a tamponade. MRI was done after 36 h after surgery which revealed total clearance. Foley's catheter was removed on the third postoperative day. Patient was discharged on the fifth postoperative day. Post operative period was uneventful. Nasal crusting was noted on the first follow up after 1 week. The crusts were not removed as patient had developed a CSF rhinorrhoea during surgery. Patient was advised nasal douching with normal saline. Crusting lasted for 3 months post surgery. The patient has been on regular follow up for last 2 years without any recurrence.

Case: 2

A 36 year old male presented with intermittent headache of throbbing type over fronto parietal region of 3–4 years

Fig. 3 CECT (coronal and sagittal) of the nose and paranasal sinuses showing expansile mass in the frontal region abutting the dura of the frontal lobe extending up to the right cribriform plate with sclerosis of the roof of the orbit and compression of the superior rectus muscle

duration. He did not complain of nasal obstruction or nasal discharge. There was proptosis of the right eye. However, patient had no visual complaints. Contrast enhanced computerised tomography (CECT) of the nose and paranasal sinuses showed an expansile mass in the frontal region abutting the dura of the frontal lobe. The mass had bony partition margins and extended up to the right cribriform plate. The roof of the orbit was sclerosed and compression of the superior rectus muscle was noted (Fig. 3). Endoscopic excision of the lesion was carried out under GA. Nose was prepared using 1:1000 adrenaline soaked cottonoids. Patient was put in reverse Trendelenburg position with head slightly tilted towards right. The nose was visualised using a 0°, 4 mm, 18 cm wide angled telescope (Karl Storz). The right middle meatus was visualized and uncinectomy was done. Bulla was removed. The fovea was defined and a bulge was seen in the right fovea. It was opened using a probe and yellowish fluid brought out. A 70° telescope was used to visualise the interior of the tumour. Bony rim was found which was dissected away from the dura. A very nice plane of dissection was achieved and complete bony wall removed. Dura was breached at few places and hence a CSF leak repair was mandatory. This was accomplished using a dumbbell fat technique with onlay fascia lata. Right middle meatus was packed with nasopore. Proptosis was relieved after surgery and patient was discharged on 3rd post-operative day. Histopathological examination (Haematoxylin & Eosin stain) of excised specimen in both cases showed tumour composed of spindle cells arranged in sheets/bundles, forming meningothelial pattern. Psammoma bodies were seen in between. There was no nuclear atypia or mitotic activity. Immunohistochemistry was positive for epithelial membrane antigen (EMA) (Fig. 4).

Fig. 4 Histopathological examination of excised specimen showing tumour composed of spindle cells arranged in sheets/bundles, forming meningotheelial pattern with Psammoma bodies (H & E stain) with immunohistochemical staining positive for Epithelial membrane antigen

Discussion

Meningiomas are non-glial tumours of the central nervous system, representing 24–30 % of all intracranial neoplasms. They arise from the arachnoid cap cells (meningocytes) which are derived from the neural crest. These tumours have a predilection for middle aged and elderly females [1]. They have been reported to occur extracranially in only 1–2 % of cases. Twenty percent of extracranial meningiomas are secondary extensions of intracranial tumours [2, 3]. Primary extracranial meningiomas with no direct communication with the intracranial region are extremely rare. They have been reported to occur from the sinonasal tract, cranial bones, middle ear, scalp and soft tissues of the face and neck and parotid gland. An analysis of 146 cases of primary extracranial meningiomas showed that majority originated from the skin and scalp ($n = 59$) followed by middle ear ($n = 26$) and sinonasal tract ($n = 25$). Other rare locations in the head and neck included the temporal bone, mandible, nasopharynx, parotid gland, orbit and neck [5]. Different mechanisms have been proposed to explain the etiopathogenesis of these lesions. They are believed to be arising from 1. Arachnoidal cells of nerve sheaths emerging from foramina in the skull 2. Displaced or entrapped Pacchionian bodies from an extracranial location during embryological development 3. Arachnoid islets displaced by trauma or cerebral hypertension 4. Undifferentiated mesenchymal cells [5].

Symptoms of these lesions depend on the anatomic site of involvement. Meningiomas involving the sinonasal tract may mimic sinusitis with patients presenting with nasal obstruction, anosmia, facial pain, nasal discharge and epistaxis. Nasal endoscopy usually shows a firm reddish pink to grey mass in the nasal cavity which could be globular or lobulated, but well-circumscribed with displacement and without infiltration into the surrounding tissues [6].

The tumours are rubbery in consistency. Depending on cellular morphology, 4 histological subtypes have been described namely meningotheliomatous type which constitutes 63 %, followed by transitional (19 %), fibrous

(13 %), and psammomatous (2 %) [4]. The differential diagnosis of sinonasal tracts meningiomas includes mucocoele, olfactory neuroblastoma, carcinoma, hemangioma, sarcoma and angiofibroma [7, 8]. Histopathology and immunohistochemistry are confirmatory. Meningiomas are strongly immunoreactive to EMA, pancytokeratin and Vimentin [9].

An aggressive psammomatoid variant of ossifying fibroma may be confused for psammomatous meningioma. Both lesions occur in middle age and patients can present with proptosis. Both lesions have abundant psammoma bodies but, meningiomas lack associated osteoblasts and osteoclasts rimming the psammoma bodies and the distribution of psammoma bodies in meningioma is haphazard. The back ground stroma in ossifying fibroma is storiform and more compact [5].

This series comprised of 2 cases of psammomatous meningioma occurring extracranially. While one case was completely extradural, the other was intradural. Our patients were relatively young and both were males in contrast to prior studies which report occurrence of these tumours predominantly among middle-aged and elderly women. No intracranial component was demonstrable in either of the 2 cases. Surgery is reported to be the only curative modality for these tumours [5, 6]. Complete extirpation of these masses may be accomplished by endoscopic, open or a combination of both approaches [5, 10]. However, meticulous surgery aided by thorough pre-operative planning based on CT or MRI images is essential to minimize recurrence. Recurrences have been observed in the same location and hence believed to represent residual tumours rather than recurrences [7]. Both the patients reported here were managed successfully by endoscopic endonasal approach with complete tumour removal and resolution of symptoms with no recurrence on long term follow up.

Conclusion

Primary extracranial meningiomas are rare. Their clinical features depend on the site of involvement. Imaging modalities such as CT and MRI establish the extent of

lesion but confirmation of diagnosis is based on histopathological examination. Complete excision of these tumours can be accomplished by endoscopic endonasal approaches when planned well and done meticulously.

References

1. Louis DN, Ohgaki H, Wiestler OD, Cavanee WK (eds) (2007) WHO classification of tumours of the central nervous system. International Agency for Research on Cancer, Lyon, pp 164–172
2. Friedman CD, Costantino PD, Teitelbaum B, Berkold RE, Sisson GA (1990) Primary extracranial meningiomas of the head and neck. *Laryngoscope* 100:41–48
3. Fernandez LR, Rivares EJ, Perez OJ, Ros ML, Gambo GP, De Miguel HD (1991) Primary meningioma of the nose and the paranasal sinus. *Ann Otolaryngol Chir Cervicofac* 108:34–37
4. Lee JH, Sade B, Choi E et al (2006) Midline skull base and spinal meningiomas are predominantly of the meningothelial histological subtype. *J Neurosurg* 105:60–64
5. Rushing EJ, Bouffard JP, McCall S et al (2009) Primary extracranial meningiomas: an analysis of 146 cases. *Head Neck Pathol* 3:116–130
6. Swain RE, Kingdom TT, DelGaudio JM, Muller S, Grist WJ (2001) Meningiomas of the paranasal sinuses. *Am J Rhinol* 15:27–30
7. Thompson LDR, Gyure KA (2000) Extracranial sinonasal tract meningiomas. *Am J Surg Pathol* 24:640–650
8. Gökduman CA, Iplikcioglu AC, Kuzdere M, Bek S, Cosar M (2005) Primary meningioma of the paranasal sinus. *J Clin Neurosci* 12:832–834
9. Ocque R, Khalbuss WE, Monaco SE, Michelow PM, Pantanowitz L (2014) Cytopathology of extracranial ectopic and metastatic meningiomas. *Acta Cytol* 58(1):1–8
10. Jiang RS, Chang KM, Hsu CY (2000) Endoscopic sinus surgery for ethmoid sinus meningioma. *Zhonghua Yi Xue Za Zhi* 63:158–161