PRIMARY EXTRADURAL SINONASAL MENINGIOMA WITH PROPTOSIS AS PRESENTATION : A RARE CASE REPORT WITH REVIEW OF LITERATURE

Neurosurgery

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ABSTRACT

Meningiomas are one of the common brain tumours which have origin for arachnoid cap cells. Extradural meningiomas are rarer variants classified into primary and secondary in nature based on their source of origin. The common location of primary extradural meningiomas is head and neck with paranasal sinuses being a common site in head and neck. The treatment of choice lies with complete excision of tumour which usually follows a good prognosis and low rates of recurrences. Current literature is scarce about primary extradural meningiomas with majority being case reports or short case series only.

We here present a case report of a primary extradural meningioma of right frontal and ethmoidal sinus treated by surgical excision. This case report adds on to the current knowledge of literature about primary extradural meningiomas.

Summary

A 28 year old male presented with chief complaints of right eye proptosis and right frontal headache with no neurological deficit reported to our centre with a trans nasal biopsy report of his lesion done elsewhere to be psammomatous meningioma Grade I. On reviewing his MRI Brain and paranasal sinuses, a totally extradural space occupying lesion in right frontal and ethmoidal sinuses communicating into intracranial cavity pushing right frontal lobe up and extending into roof of right orbit was found. Patient underwent right frontal craniotomy with complete excision of tumour with subsidence of proptosis in immediate post operative period.

Primary extradural meningiomas are rare entities with scarce literature available. We write this case report to add on to current literature available and also briefly discuss the literature review known currently.

KEYWORDS

Meningioma, Extradural, Primary, Paranasal Sinuses

INTRODUCTION :

Meningioma is a common intracranial neoplasm with a variety of histomorphologic growth patterns which are usually easily recognized. Meningioma constitutes 18-20% of total brain tumours reported in live population out of this about 1% meningiomas are extradural^(1,3) which comprises of primary extradural and those extending from a primary intradural meningioma, namely secondary extradural meningiomas. The common locations for extradural meningiomas include intradiploic, orbit, paranasal sinuses, nasopharynx, adrenal gland, mediastinum, skin and lungs. The first reported case of primary extradural meningioma of sinonasal region dates back to 1730 when it was first reported then by Johann Salzmänn.⁽²⁾ In India, There have been case reports reported with a series of three cases being the largest series by Ranjan G. Aiyer et al⁽⁴⁾ but not much of the case reports and literature available in post CT era. Only few of these reported cases presented as proptosis. Here we present a rare case of primary extradural meningioma of frontal sinus with extension to ethmoid sinus and reaching upto roof of the orbit leading to presentation of patient as proptosis of involved side. It's a rare case reported presented here with relevant literature review.

Case report :

A 28 year old gentleman formally educated upto 12th standard and farmer by occupation presented with chief complaints of protrusion of right eye for last two years and headache over right frontal region for 2 months. The protrusion was insidious in onset, gradually progressive in nature, painless, not associated with any diminution of vision or any restriction of eye movements, discharge from eyes, conjunctival congestion, corneal erosion, preceding trauma, pulsations in eye ball. Patient also started having headache localised in right frontal region insidious in onset, gradually progressive, constant dull aching, relieved with medication with no aggravating factor no diurnal variation, not associated with any aura, conjunctival congestion, nausea, vomiting, loss of consciousness or any focal neurological deficit. No history of any seizures, weakness in any part of body or change in behaviour was there.

Patient reported with above presentation to some other hospital where patient was investigated with MRI brain with paranasal sinuses (Fig 1) which suggested presence of an extradural space occupying solid

cystic lesion iso to hypo intense solid part and CSF intensity cystic lesion pushing right frontal lobe upwards extending into right frontal sinus and right ethmoid sinus reaching upto roof of right orbit and pressing on superior and medial rectus muscle. A transnasal biopsy was done elsewhere which reported it to be psammomatous meningioma grade I and patient was referred to our centre.

Fig 1. MRI Brain and paranasal sinuses showing extradural solid cystic mass occupying right frontal sinus, right ethmoidal sinus, pushing the right frontal lobe upward and extending upto roof of right orbit compressing superior and medial rectus causing eye to prop out and down.

On examination patient was conscious with MMSE 27/30 and intact vision and other cranial nerves and no sensory motor deficits. There was an extraaxial proptosis of right eye ball with no loss of vision or extrocular movement. (Fig 2) On CT evaluation lesion was causing widening of right frontal sinus with no breach of bone limits and thinning but no erosion of right orbit. Rest of the routine investigations of the patient were essentially normal.

Fig 2. Pre operative picture of patient with right eye proptosis

Patient underwent right frontal craniotomy and excision of lesion. Tumour was extradural, solid cystic in nature, cystic part contained serous fluid, solid part was greyish, firm, non suckable, mildly vascular with well defined plane of cleavage. Tumor was expanding the walls of right frontal sinus, complete excision of tumour was done with adequate decompression and visualisation of right optic nerve as it exits from right orbit and continues as intracranial segment. Post operative recovery of patient was uneventful with subsidence of proptosis (Fig 3) in immediate post operated period and intact vision and extraocular movements. MRI brain done in post operated period suggested of complete excision of tumour (Fig 4).

Fig 3. Post operated day 1 clinical picture of patient with subsidence of right eye proptosis following tumour excision.

Histopathological examination reported presence of tumour cells arranged in intersecting fascicles and whorls comprising of meningothelial cells having round to uniform nuclei with abundant eosinophilic cytoplasm and indistinct cell borders with numerous psammoma bodies with MIB-1 index < 2%. Presence of meningeal lining was also reported. IHC reported to be positive for expression of EMA.

DISCUSSION :

Primary extradural meningiomas (PEM) represent 1% to 2% of all meningiomas^(1,3) while PEMs in head constitute 80% to 90% of all PEMs^(5,6). It can be calculated that the percentage of PEMs in head as a proportion of all meningiomas is between 0.8% and 1.8%. The origin of PEMs is still unclear. Various hypothesis for origin of PEM includes : 1. Ectopic meningocytes or arachnoid cap cells in the cranial, peripheral nerve sheaths and subcutaneous tissue. Meningiomas in paranasal sinuses and orbit were suggested to come from arachnoid cells located along the cranial and peripheral nerve sheaths.⁽⁷⁾ 2. misplaced arachnoid cells in an extracranial location during embryologic development; 3. Stemming from the arachnoid cap cells trapped in the cranial suture during birth and molding of the head; 4. dural and/or arachnoid cells entrapped in the sites of traumatic cranial fractures ; 5. multipotential mesenchymal cells, including fibroblasts and Schwann cells or a combination of these located extradurally. PEM are slightly more common in males with bimodal peak of distribution in second and fifth decade of life.⁽⁸⁾ The most common clinical feature with painless swelling in affected region or pressure effect of growing lesion as was present as proptosis in our case. Neurological deficits or effects of raised ICP features are uncommon. In 2000, Lang et al proposed a simple and useful classification of PEMs in skull according to their location relative to the cranium⁽⁹⁾ which was later modified by Bassiouni et al in 2006⁽⁹⁾ with emphasis on dural attachment as it has significant surgical implication. Type 1 purely extracalvarial, Type 2 purely calvarial, Type 3 calvarial tumours with extracalvarial extensions. A round lesion associated with local skull changes is one of the common radiographic features in PEMs in head. Usually, the skull changes present as hyperostosis (59%), osteolysis (35%) or mixed (6%), more obvious particularly on bone-window CT scans^(6,10). This was the radiological finding present in our case also where blastic changes present in bone cortices of frontal sinus and thinning of roof of right orbit was also present, Presence of the osteolytic skull lesion, scalp swelling and painful extracranial soft

tissue mass indicates presence of malignant PEM⁽¹¹⁾. The most common histopathological subtype of PEMs in head is meningothelial meningioma, others includes transitional, psammomatous and fibrous⁽¹⁾. In our case it was psammomatous meningioma. The choice of treatment for PEM is surgical removal with dual goal of the confirming diagnosis and excision of mass, possibly all the involved tissue should be excised with reconstruction wherever required⁽⁶⁾. The prognosis after surgical excision of PEM is usually good with less chances of recurrences.

In our case also we achieved complete excision of lesion with uneventful recovery in post op period achieving subsidence of proptosis in immediate post surgery period only. The patient is in regular follow up at present. Our case adds on the literature for the rare entity of primary extradural meningiomas with more common sites as paranasal sinuses which responds well to complete surgical excision.

CONCLUSION:

Primary extradural meningioma is an uncommon location for meningioma with paranasal sinuses and orbit being one of the common location of these meningiomas which should be kept in consideration with clinical presentation of painless swelling or mass effect over affected region with typical radiological findings and should be treated primarily by surgical excision. The pathology has a good prognosis with less propensity of recurrences.

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